

Abstract booklet

Hematopathology Summer Meeting, Gdańsk 8-9th July 2024

Atheneum Gedanense Novum

Local institutions:

Copernicus PL Hospitals Gdańsk Medical University of Gdańsk (MUG)



Scientific Programme

<u>Day 1</u>

Key Lecture: New WHO classification of lymphomas. Part 1- Prof. German Ott (Institut für Klinische Pathologie, Stuttgart, Germany)

Novel therapies in lymphomas - prof. Maciej Zaucha (MUG)

Lymphoma mimickers. Post- transplant disorders – dr Monika Klimkowska (Karolinska Institutet, Stockholm, Sweden)

Molecular diagnostics in lymphomas – prof. Ewa Chmielik, dr Karolina Gajda, dr Wojciech Fidyk (MSCNRIO Gliwice)

Slide seminars 1,2

Day 2

Lymphomas of the CNS – E. Iżycka-Świeszewska (MUG, Copernicus)

Key Lecture: New WHO classification of lymphomas. Part 2- Prof. German Ott

High grade lymphomas in my practice - prof. Grzegorz Rymkiewicz (MSCNRIO Warsaw)

Pediatric lymphomas – dr Jagoda Małdyk, prof. Wiesława Grajkowska (Medical University of Warsaw, Children's Memorial Health Institute, Warsaw)

Al in hematopathology – dr Krzysztof Borkowski (Evident Scientific)

Slide seminars 1, 2

Organizing committee:

prof. Ewa Iżycka-Świeszewska (MUG, Copernicus PL), dr Monika Klimkowska (Karolinska Institutet, Stockholm), prof. Ewa Chmielik (Maria Sklodowska-Curie National Research Institute of Oncology, Gliwice), prof. Wiesława Grajkowska (Children's Memorial Health Institute, Warsaw), dr Jacek Gulczyński (MUG, Copernicus), dr Monika Nowaczyk (Copernicus PL), lek. Mateusz Michalski (MUG, Copernicus PL)

LECTURES:

1. "New WHO classification of lymphomas".

Prof. German Ott Institut für Klinische Pathologie, Stuttgart, Germany

General remarks

The 5th edition of the World Health Organization (WHO) Classification of Haematolymphoid Tumours (WHO-HAEM5) builds on the revised 4th edition of 2017 (WHO-HAEM4R) and, therefore, represents a systematic evolution of concepts laid down in this and former editions of the classification. Changes to existing nomenclature were made sparsely and only when deemed necessary or for clarity or consistency. The WHO-HAEM5, like all 5th Edition WHO Tumour volumes, has adopted a (Linnean) hierarchical classification system ordering diseases in increasing levels of specificity (https://tumourclassification.iarc.who.int/home): category (e.g., mature B-cell), family/class (e.g., large B-cell lymphomas), entity/type (e.g., diffuse large B-cell lymphoma, not otherwise specified [DLBCL, NOS]) and subtype (e.g., DLBCL, NOS, germinal center B-cell-like). Within a family, the entities are generally arranged from indolent to increasingly aggressive. For the first time, non-neoplastic conditions mimicking lymphoma or representing an important differential diagnosis have been included in the WHO classification, as well as a chapter on genetic tumor syndromes associated with haematolymphoid tumours. The WHO-HAEM5 recognizes the ever-growing impact of molecular data in the diagnostic work-up, but also appreciates that the required diagnostic resources are not universally available. Therefore, to enable a pragmatic approach to diagnosis, "essential" and "desirable" diagnostic criteria for each entity are defined in a hierarchical way, a principle followed throughout the 5th edition of the classification.

Indolent (small) B-cell lymphomas

Major changes have been made in the follicular lymphoma (FL) and the splenic B-cell lymphomas/leukaemias sections. Of major importance, the grading of FL is now optional in a diagnostic pathology report. A new term, classic FL (cFL), incorporating FL grades 1, 2 and 3A was proposed. This adjustment takes into consideration that most of these FL have a follicular growth pattern, are composed of centroblasts and centrocytes, show a homogeneous immunophenotype with expression of germinal center (GC) markers, and also harbor the t(14;18)(q32;q21) in most (60-85%) cases, implying a common biological background. FL with a predominantly diffuse growth pattern and FL with unusual cytological features, e.g., "blastoid FL" are set apart from cFL. The term "follicular large B-cell lymphoma (FLBCL)", formerly FL grade 3B, has been introduced to emphasize its close relationship with DLBCL, while still regarded as belonging to the FL family. The family/class of splenic B-cell lymphomas/leukemias has been reorganized in WHO-HAEM5. It includes hairy cell leukemia (HCL), splenic marginal zone lymphoma/leukemia (SMZL), splenic diffuse red pulp small Bcell lymphoma/leukemia (SDRPL) and a new entity, splenic B-cell lymphoma/leukemia with prominent nucleoli (SBLPN). B-cell prolymphocytic leukemia (B-PLL) has been removed because of its recognized heterogeneous nature and continuing debate on its validity as an entity. Likewise, the provisional category of WHO-HAEM4R "hairy-cell leukemia variant" has been withdrawn given its unrelatedness to HCL. Instead, a new "placeholder term" of splenic B-cell lymphoma/leukemia with prominent nucleoli (SBLPN) has been adopted for those cases of primary splenic lymphomas that at present defy reliable assignment to a biologically meaningful entity. Only minor changes have occurred in the other chapters on indolent/small Bcell lymphomas. Prolymphocytic progression of CLL/SLL is a new term used for cases with >15%

prolymphocytes in peripheral blood with a CLL/SLL immunophenotype. The term histologically aggressive CLL/SLL is reserved for cases with enlarged or confluent proliferation centers (PCs) according to traditional definitions of spanning the diameter of a visual field using a 20x objective lens and 10x ocular lens or with high proliferation indices (>2.4 mitoses/PC of traditionally defined size or >40% Ki67+ nuclei). The name 'Richter transformation' is recommended instead of 'Richter syndrome'. In-situ mantle cell neoplasm and leukemic non-nodal MCL have been promoted to definitive entities.

Aggressive B-cell lymphomas

Updates from WHO-HAEM4R to WHO-HAEM5 in the aggressive lymphomas are only minor. The WHO-HAEM5 still recommends COO-classification of DLBCL NOS in diagnostic pathology reports. In contrast, the more complex, genetically-based classifiers of DLBCL NOS that have recently been developed need to be further tested and validated in larger multicenter DLBCL cohorts before these or other new classifiers can be recommended for implementation in routine diagnostics. For primary diffuse large B-cell lymphomas of the central nervous system (CNS), primary large B-cell lymphoma of the vitreoretina and primary diffuse large Bcell lymphoma of the testis represent aggressive LBCL that arise in immune-privileged sites, the WHO-HAEM5 has created an umbrella category termed 'primary large B-cell lymphoma of immune-privileged sites' to encompass these three entities. With time, DLBCLs of other anatomic sites may be added to this group as their underlying biology is better understood. Another group of aggressive LBCL that has been upgraded to a definitive entity is the high-grade B-cell lymphoma (HGBCL) with 11q aberration (WHO-HAEM4R: Burkitt-like lymphoma with 11q aberration). The definition of the so-called 'double hit lymphomas' (WHO-HAEM4R: HGBCL with MYC and BCL2 and/or BCL6 rearrangements) has changed to accommodate new biological data. Recent studies suggest that aggressive BCL with MYC and BCL2 rearrangements constitute a relatively homogeneous biological and clinical entity. These lymphomas constitute a separate entity in the WHO-HAEM5. Since they morphologically present as either DLBCL (the majority) or HGBL, and since morphological diagnosis usually precedes results of genetic testing, these cases should be diagnosed as "DLBCL with" or "HGBL with rearrangements of MYC and BCL2" even though they belong to the same entity. In contrast, aggressive lymphomas with MYC and BCL6 rearrangements are molecularly heterogeneous and distinct from those with dual MYC and BCL2 rearrangements. Consequently, aggressive LBCL with MYC and BCL6 rearrangements are grouped among DLBCL NOS (if the morphology fulfills the criteria of DLBCL) or among HGBCL, NOS (if the morphology is 'high-grade').

Fluid overload-associated large B-cell lymphoma (FO-LBCL), previously known in the literature as "HHV8negative effusion-based lymphoma", is now considered a definitive entity. In contrast to primary effusion lymphoma, which is by definition KSHV/HHV8+ and typically affects young to middle-aged HIV positive men and generally involves a single body cavity, FO-LBCL affects older adults and frequently involves more than one body cavity. A broad array of immunologic settings predisposes to lymphoid proliferations and lymphomas (LPDs). This understanding is reflected in the WHO terminology of "immune deficiency and dysregulation" (IDD). The WHO-HAEM5 introduces a standardized three-part modular nomenclature consisting of the pathologic entity, oncogenic virus, and IDD setting: for example, diffuse large B-cell lymphoma (DLBCL), EBV+, post solid organ transplant. Especially, recognition and diagnosis of IDD-related hyperplasias and lymphoproliferative disorders of varied malignant potential are crucial to allow patientcentered care and avoid overtreatment as lymphoma. Both the pathologic diagnosis and the IDD setting impact the clinical behavior. The modular nomenclature and the expansion and more granular understanding of underlying IDD categories positions the WHO-HAEM5 classification to support standardized clinical diagnostics, exploration of novel biology-based treatment across IDD-setting and, crucially, collaborative research that is truly global in scope.

2. Lymphoma mimickers. Post- transplant disorders.

dr Monika Klimkowska Karolinska Institutet, Stockholm, Sweden

Knowledge of both inborn and acquired aberrations of the immune system is constantly increasing. On the other hand, the growing repertoire of therapeutic modalities requires as much attention since these may inadvertently affect the immune system, and thus require as much attention. Lymphoproliferative disorders developing in the setting of impaired immune response share some morphological features, such as occurrence of hyperplastic lesions, polymorphic proliferations or lesions that fulfil the diagnostic criteria of malignancy. However, the clinical course in these cases is variable and often deviates from scenarios known in immunocompetent subjects. Overview of the major categories of inborn immune defects and the resulting lymphoproliferations is presented, followed by current view on classification of disorders secondary to specific treatments, including

3. Molecular diagnostics in lymphomas.

dr Wojciech Fidyk, dr Karolina Gajda, prof. Ewa Chmielik Maria Sklodowska-Curie National Research Institute of Oncology , Gliwice

Molecular diagnostics in hematological diagnostics encompasses a variety of research methods. Among these, tests utilizing the FISH technique play a significant role. Most of the diagnosed diseases have a characteristic set of chromosomal abnormalities that need to be examined to establish an accurate diagnosis. These tests also serve prognostic purposes and allow for better predictions regarding disease progression and aggressiveness. For the proper execution of FISH diagnostics, it is essential to be familiar with the appropriate diagnostic protocols and recommendations from national and international clinical societies. These schemes outline the necessary molecular probe panel to be performed, distinguishing between the time of initial diagnosis and disease monitoring. During the diagnostic procedures, the ability to appropriately analyze the obtained results and a deep understanding of the method are crucial to avoid numerous difficulties related to the interpretation of the final data.

The family of large B-cell lymphomas comprises a wide spectrum of tumours. Algorithm for classification of aggressive B-cell lymphomas in WHO-HAEM5 includes MYC, BCL2 and BCL6 rearrangement and complex 11q gain/loss patterns. Fluorescence in situ hybridisation (FISH) is a method for identification of genetic aberrations e.g. translocations, deletions, amplifications in formalin-fixed, paraffin-embedded (FFPE) tissue sections. EBV infection plays an essential role early in pathogenesis causing B cells thats why qualitative detection of human Epstein-Barr virus (EBV) EBER RNA plays an important role in diagnosis. There are many genetic fndings and biomarkers that have potential relevance in aggressive / large B-cell lymphomas.

4. Lymphomas of the Central Nervous System.

prof. Ewa Iżycka-Świeszewska MUG, Copernicus PL

CNS lymphomas include B- cell mainly and rarely T- cell neoplasms. In diagnostics of CNS lymphomas, the clinical context is critical. The most common entity is high grade primary diffuse large B-cell lymphoma of the CNS, which belongs to the category of primary large B-cell lymphomas (LBCLs) of immune-privileged sites (IP-LBCLs). PCNS -LBCL can relapse also outside the CNS, typically within testes or vitreo-retina. Its essential

features: large B-cell lymphoma primarily confined to the CNS at presentation; exclusion of secondary brain involvement, and exclusion of immune deficiency/dysregulation-related settings. Typical it has postgerminal-centre B-cell phenotype (IRF4 [MUM1]+, BCL6+, CD10-), and is EBV – negative (> 97% of cases). When histology is not definitive (corticosteroid-mitigated PCNS-LBCL), demonstration of clonality or MYD88 and/or CD79B hotspot mutations is helpful. PCNS-LBCL constitutes 2.4–3% of all brain tumours. The next frequent is immunodeficiency- associated lymphoma HGBL (8-10% of all primary CNS lymphomas) found in patients with inherited or acquired immunodeficiency, related to HIV/ AIDS and iatrogenic disease, posttransplant.

Moreover, brain can be very rarely involved by lymphomatoid granulomatosis, intravascular large B-cell lymphoma, as well as miscellaneous low grade lymphomas such as MALT lymphoma of the dura, small lymphocytic lymphoma or lymphoplasmacytic lymphoma. Lymphomatoid granulomatosis is a highly malignant disorder with angiocentric/ angiodestructive pattern, and polymorphous lymphoid infiltrates of EBV-positive atypical B cells in T cell–rich inflammatory background forming ill-defined lymphohistiocytic nodules and causing brain infarcts. Exceptional rare T-cell CNS lymphomas include anaplastic large cell lymphoma (ALK+ nad ALK-), as well as T-cell and NK/T–cell lymphomas.

CNS lymphomas have different clinical presentation including progressive neurocognitive deterioration, neurological deficits, headaches, palsies, and stroke-like incidents.

5. Story of the discovery of BLL-11q (HGBL-11q) by flow cytometry From Burkitt-like lymphoma with 11q aberrations (BLL,11q, 2016 WHO) through High grade B-cell lymphoma with 11q aberrations (HGBL-11q, 2022 WHO) to new date, not yet published (next WHO classification?)

prof. Grzegorz Rymkiewicz

Maria Sklodowska-Curie National Research Institute of Oncology, Warsaw

In 2011, we described, for the first time, a previously unrecognized form of a very rare lymphoma, similar to Burkitt lymphoma (BL) without rearrangement of the MYC gene (Pieńkowska-Grela B and Rymkiewicz G et al., Partial trisomy 11, dup (11) (q23q13), as a defect characterizing lymphomas with Burkitt pathomorphology without MYC gene rearrangement, Med Oncol 2011) but with a duplication and an inversion of a duplicated fragment of the long arm of the chromosome 11, visible in classic cytogenetics (CC) - a karyotype supplemented with fluorescent in situ hybridization (FISH) using several FISH probes. This publication was based on only four cases diagnosed at our medical centre between 1998 and 2011. The discovery of this lymphoma was directly related to flow cytometry immunophenotypic analyses by the author of this lecture and CC assessments of cellular suspension obtained by fine needle aspiration biopsies. In another publication (Salaverria I et al., A recurrent 11q aberration pattern characterizes a subset of MYCnegative high-grade B-cell lymphomas resembling Burkitt lymphoma, Blood 2014) we characterized molecularly, among others, by means of an array comparative genomic hybridization and single nucleotide polymorphism, recurring changes on the long arm of the chromosome 11, as 11q-gain/loss, on several available cases. Based on these two publications in 2017, the lymphoma similar to BL with 11q-gain/loss was included as a provisional nosological entity in the updated of 2017 World Health Organization (WHO) classification, as a MYC-negative "Burkitt-like lymphoma with 11q aberration, BLL,11q". The recent 5thedition of the new 2022 WHO Classification of Haematolymphoid Tumours (WHO-HAEM5) changed the name of the provisional BLL,11q entity to "High-grade B-cell lymphoma with 11q aberration, HGBCL-11q". Our contribution to the discovery of HGBCL-11q was appreciated and I became a co-author of the WHO-HAEM5 subsection on HGBCL-11q. During this lecture I discuss all clinical, pathomorphological and molecular aspects of HGBCL-11q. The characteristics of this entity (in WHO-HAEM5) in the major part remain the same: an aggressive mature B-cell lymphoma (B-NHL) with a morphology similar to BL or with an intermediate or blastoid appearance in most cases and coarse apoptotic debris within starry sky macrophages. HGBCL-11q

frequently affects children or young adults with strong male predominance, although cases have been reported in older patients up to 82 years of age. Compared to BL, HGBCL-11q commonly has a nodal presentation often with a single bulky tumor, along with some localized lymphadenopathy. The most commonly affected regions are head and neck and abdominal lymph nodes. Infiltration of bone marrow or cerebrospinal fluid involvement is not typically seen although the bone marrow involvement has been reported. Rare cases have also been reported in the post-transplant setting. This lymphoma has a distinctive CD20+/CD10+/BCL6+/BCL2-/ MUM1-/MYC+/ CD44-/CD43± immunophenotype and a high Ki-67 index approaching 100%. Unlike BL, HGBCL-11q is often positive for LMO2, CD56 and always EBER-negative. According to the definition, HGBCL-11q is a MYC rearrangement negative aggressive lymphoma with 11qgain/loss, representing a chromosomal aberration specific to this neoplasm. The minimal gain region covers 11q23.2–q23.3 while the minimal telomeric loss includes 11q24.3-11qter. Loss in the 11q24qter region is considered more specific to this entity than the centromeric gain. The gene expression profile of HGBCL-11q is similar to that of BL, however, molecular studies confirmed that the mutational spectrum is different from that of BL and is more similar to that of diffuse large B-cell lymphoma (DLBCL) of germinal center B-cell like (GCB) subtype. Potential driver mutations are described in BTG2, DDX3X, ETS1, EP300, GNA13, and NFRKB genes. BL-associated mutations in TCF3 and ID3 are absent in HGBCL-11q.

Of importance, the WHO-HAEM5 definition of HGBCL-11q describes this entity as MYC rearrangementnegative, suggesting that the 11q-gain/loss aberration may functionally replace the MYC rearrangement (MYCR). However, there is a growing number of reports describing cases with co-occurrence of the 11qgain/loss pattern and MYC rearrangements (HGBCL-11q,MYCR). These rare cases of HGBCL-11q,MYCR are by definition excluded in the WHO-HAEM5 and knowledge about their clinicopathologic, cytogenetic, and molecular characteristics is limited.

The aim of the last part of this lecture is to decipher the clinicopathological, chromosomal and mutational characteristics of HGBCL-11q, MYCR in relation to HGBCL-11q, and additionally to compare with other aggressive MYCR lymphomas such as BL and High-grade B-cell lymphoma not otherwise specified with MYCR. Our data show that HGBCL-11q, MYCR is distinct from BL and HGBCL-11q at the clinicopathological, cytogenetic and molecular levels; although it shares common features of both B-NHLs. The chromosomal change distinctive of HGBCL-11q, MYCR is the gain or amplification of 3q29 compared to the other B-NHLs. Our cohort of patients with HGBCL-11q, MYCR had a similar, excellent relapse-free survival as the patients with HGBCL-11q and BL, if treated with BL-targeted regimens.

In conclusion, the entire history of the discovery HGBCL-11q as well as the clinical, pathomorphological, and molecular characteristics of HGBCL-11q are described in the context of clinical, pathomorphological, and molecular similarities and differences between MYC-negative HGBCL-11q and MYC-positive HGBCL-11q.

6. Pediatric lymphomas.

dr Jagoda Małdyk, prof. Wiesława Grajkowska Medical University of Warsaw, Children's Memorial Health Institute, Warsaw

Lymphomas occuring in children are diagnosed according to the same rules as those in adults. The most commonly diagnosed include: Burkitt's lymphoma, lymphoblastic lymphoma, anaplastic large cell lymphoma (ALCL) and diffuse large B-cell lymphoma (DLBCL). In diagnosing the lymphoblastic lymphomas, precursor cell markers such Tdt are used. The exception is indolent T - lymphoblastic proliferation, which mimics lymphoma, but is an expansion of T lymphoblasts without features of clonal TCR rearrangement. The absence of immunoexpress of Tdt does not exclude lymphoblastic lymphoma but may be a cause of misdiagnosis. In the case of B-cell proliferation, it is necessary to exclude Burkitt's lymphoma, for the diagnosis of which it is necessary to demonstrate MYC translocation. Within the group of mature B-lymphocyte lymphomas, two paediatric variants have been distinguished: paediatric nodal marginal zone

lymphoma and paediatric follicular lymphoma due to the different clinical course and different therapy than in adult counterparts. In children , the wait and watch approach is recommended.

Of the mature T lymphocytes, the most commonly diagnosed lymphoma in children is ALCL with CD30 expression which needs to be differentiated from from other peripheral T lymphocyte lymphomas, Hodgkin's lymphoma and DLBCL. Most ALCLs in children are ALK1+. The immunoexpression of ALK is also detected in other paediatric small blue cell tumours such as neuroblastomas or rhabdomyosarcomas. The second major group of peripheral T-cell proliferations are proliferations associated with EBV infection/EBV positive T-cell and NK-cell lymphoid proliferations and lymphomas of childhood. In this group we can distinguish: EBV+T-cell lymphoma/systemic EBV -positive T-cell lymphoma of childhood and CAEBV/chronic active EBV disease/ - entities with a different clinical course and a different therapeutic approach.

7. Al in hematopathology.

dr Krzysztof Borkowski Evident Scientific

A modern pathology laboratory can make a diagnosis based on image data. This can be done using digital pathology systems. The diagnosis is made by a pathologist. Artificial intelligence-based diagnostic systems can facilitate and speed up this process. But these systems do not diagnose, these systems are morphology-based clinical decision support tools (CDST).

The use of virtual slides makes it possible to use them in routine diagnostics, consultations and teaching systems. Combining digital pathology with artificial intelligence systems makes it possible to accelerate diagnosis. Using the huge amount of data contained in virtual preparations, such systems can very quickly indicate places that are critical to making a diagnosis. Automatic scanning systems can search for specific regions during scanning

AI-based systems allow for the extraction of information from tissue specimens, supporting a quantitative and structured approach to morphology, and may yield morphology-based clinical decision support tools (CDST) in pathology workflows

Modeling the pathologist's thought process and actively engaging pathologists in the design and validation of deep models is essential to the successful implementation of AI models with real clinical utility. The pathologist's role is to first verify the quality of the virtual slide then to find and mark the structures and then check their recognition. Clinical validation is a big problem. This clinical validation will require wider adoption of digital workflows, particularly in hematopathology, where models will need to be very well validated across multiple datasets to ensure reproducibility and accuracy. This requires wide access to WSI imaging data and clinical data. Only the combination of image data with clinical data allows the creation of neural networks that are high-class diagnostic tools.

There is an increasing number of scientific works on the use of AI in hematopathology. However, there are numerous legal barriers, such as the validation and certification of scanning systems and software used to create neural networks.

SLIDE SESSION

Waldenstrom macroglobulinemia/ lymphoplasmacytic lymphoma – a case study.

prof. Bogna Wróblewska, Dpt. of Pathology Medical University of Warsaw

Waldenstrom macroglobulinemia is a rare indolent B-cell non-Hodgkin lymphoma with lymphoplasmacytic morphology, associated with IgM monoclonal gammopathy. The coexistence of WM and AL amyloidosis is an uncommon but well-described phenomenon. In patients suffering from IgM AL amyloidosis soft tissue involvement and neuropathy are more prevalent in comparison to non-IgM patients. We describe a case of 82-year old female with WM and intercurrent IgM AL amyloidosis, presenting with massive amyloidomas of both lower extremities, without significant cardiac and renal involvement. The patient was refractory to several lines of treatment and finally started on zanubrutinib monotherapy, with rapid and sustained very good partial hematologic response and clinical improvement.

(A case study was accepted for publication in: Current Problems in Cancer Case Reports.)

Differential diagnosis of extranodal marginal zone lymphoma.

prof. Anna Szumera- Ciećkiewicz,

Maria Sklodowska-Curie National Research Institute of Oncology, Warsaw

An extranodal marginal zone lymphoma (eMZL) is the third most common type of breast B-cell lymphoma. Localization in the breast accounts for <1% of malignant breast tumours and <1% of all lymphomas. It may be often clinically mistaken for breast cancer when a firm palpable mass has been developed and detected through mammography or ultrasound. Breast eMZL follows an indolent course, usually managed with excision/radiotherapy alone. The criteria for identifying primary breast lymphomas include (1) the lymphoma's primary localization should be in the breast; (2) lymphomatous infiltrates directly contacting breast tissue; (3) no history of lymphoma elsewhere within the last 6 months; (4) the involvement of axillary lymph nodes indicates a more advanced stage of the disease.

Here, we present a 52-year-old female with a right breast tumour (primary lesion) size 40 x 29 x 20 mm; radiologically category 5: highly suspicious of malignancy, without abnormalities in morphology or LDH level or lymphadenopathy. The eMZL was diagnosed with a wide panel of antibodies. In summary, breast eMZL is a rare malignancy. Most patients presented with stage I-II disease; however, isolated cases may be diagnosed with stage IV disease primarily due to occult bone marrow involvement. Interestingly, eMZL is frequently non-FDG avid on staging PET/CT. The overall survival is around 80%. It can be effectively treated with radiation therapy, providing long-term disease control.

Histiocytic sarcoma of the brain – diagnostic problems.

dr Aleksandra Sejda,

University of Warmia and Mazury Faculty of Medical Sciences; Olsztyn

Histiocytic sarcoma is very rare tumor occurs in lymph node and extranodal locations. Up to date less than 100 cases were reported in Centeral Nervous System (CNS). We describe a case of 44-male presented with headache and epilepsis. MRI scans reveals pathologic contrast-enhancing mass in temporal lobe with suspicion of meningioma. On H&E slides tumor was composed of non-cohesive large, pleomorphic cells with abundant eosinophilic cytoplasm and marked mitotic activity. There were foci of necrosis and inflammatory background. Tumor cells were positive for CD68, CD14, CD4 and S100. Patient was treated with radiotheraphy. The year after the primary diagnosis he experienced local recurrence. Histocytic sarcoma should be taken under consideration when hematolymphoid tumors are presented in CNS.

Diffuse large B-cell lymphoma , CD56 positive of the thyroid. -

dr Dorota Ponikiewska, prof. Ewa Chmielik

Maria Sklodowska-Curie National Research Institute of Oncology, Gliwice

Diffuse large B-cell lymphoma is the most common non-Hodgkin lymphoma. CD56 expression in this type of lymphoma is very unusual and occurs in 1.2% to 7% of cases. Lymphomas with CD56 expression often occur in extranodal locations, especially in the Waldeyer's ring and gastrointestinal tract. The assessment of the clinical significance and actual incidence is hampered by the lack of routine inclusion of CD56 in immunohistochemical panels and flow cytometry used in the diagnosis of diffuse large B-cell lymphomas. Lymphomas with CD56 expression are usually characterized by an adhesive type of growth, which can cause difficulties in differential diagnosis, especially in rarer locations. In our case, the lesion concerned the thyroid and cervical lymph nodes in a 71-year-old woman with no history of malignancies. Initially, based on the material obtained from a fine-needle aspiration biopsy of the left thyroid lobe tumor, a suspicion of a poorly differentiated neoplasm (cytological category V according to the Bethesda 2023 classification) was made, and then a cancer metastasis was diagnosed in a fine-head biopsy of the cervical lymph node due to, among other things, cohesive arrangements of neoplastic cells. In the postoperative material, the thyroid tissue was almost entirely covered by infiltration of large B-cell lymphoma, similar was visible in the cervical lymph nodes. In the immunohistochemical examination, the neoplastic cells expressed CD45 LCA, CD20, CD79a, CD19, CD56, PD-L1 22C3, while no reaction with ALK1 (CD246), BCL2, c-MYC, CD10, MUM1, CD30, CD15 and EBV LMP1 was observed. CD3 and CD5 expression was present in the accompanying T lymphocytes. Remnants of lymph nodes expressing CD23 were also visible. No MYC rearrangement was observed in the FISH study. Primary thyroid lymphomas are rare, accounting for 1 to 5% of thyroid malignancies and 1 to 2% of extranodal lymphomas. Most often, they are diffuse large B-cell lymphomas. They are 2.5 times more common in women, and the median age of occurrence is 65 years. Factors that worsen the prognosis include high clinical stage, poor general condition, advanced age of the patient, non-GCB subtype, dual MYC/BCL2 expression, and failure to

use rituximab in therapy. The role of CD 56 in diffuse large B-cell lymphoma is unclear. Few studies indicate that the presence of expression may indicate a poor prognosis.

Diagnostic problems in a marginal zone lymphoma

Lek. Kamil Buczkowski, dr Iwona Szutkowska

Copernicus PL- Dpt. of Pathomorphology, Dpt. of Hematology, MUG Gdańsk – Dpt. of Pathology & Neuropathology

We present a case of a 39-year-old male who was admitted to a hematology outpatient clinic exhibiting severe B symptoms preceded by a COVID-19 infection. Comprehensive testing ruled out EBV, CMV, HIV, HBV, HCV, and Toxoplasmosis.

Initial biopsies of lymph nodes revealed distorted architecture with preserved germinal centers and paracortical proliferation, made of small B-cell lymphocytes (CD3-, CD5- CD10-, CD23-, bcl6- and bcl2+). The trephine biopsy was inadequate, showing only a few nodules of small B-cell lymphocytes without signs of monoclonality (kappa=lambda).

The overall morphology was unlikely to indicate a lymphoproliferative process, suggesting atypical proliferation of the paracortical zone.

Due to persistent lymphadenopathy and the detection of a small monoclonal population of Bcell lymphocytes in the flow cytometry, repeated lymphadenectomy and trephine biopsy were performed. The histological picture was similar to the previous biopsies.

The samples were sent to Germany for a consultation, where the clonality of the process was confirmed by PCR, finding the presence of the amplification product of framework regions FR3A and FR2A, resulting in a change of diagnosis to the Marginal zone lymphoma with partial follicular colonization and bone marrow infiltration.

The patient commenced an R-COP immunochemotherapy regimen, reaching a clinical and radiological remission.

Histiocytic high grade neoplasm in a child - a challenging case.

prof. Ewa Iżycka-Świeszewska, dr Grażyna Kobierska-Gulida, prof. Ninela Irga-Jaworska Copernicus PL, MUG Gdańsk - Dpt. of Pediatrics, Oncology and Hematology, MUG

We present problematic diagnostic process in a 9 years old boy with mediastinal and cervical lymphadenopathy , hepatosplenomegaly, and cranial nerve VI palsy. His history was short, with rapid nodal enlargement. The supraclavicular lymph node was examined, showing total effacement of the architecture and diffuse infiltrates, in part alveolar growth pattern of a malignant cells with abundant cytoplasm, vesicular nuclei and prominent nucleoli. Neoplasm showed high mitotic activity, apoptosis, some multinucleated cells, and scattered eosinophils and plasmacytes within stroma. Moreover, angioinvasion and emboli within the perinodal fat tissue. The wide immunophenotyping was performed, showing positivity for LCA, Vim, CD56, CD4, and CD31, pan CK, Ck8/18, CD68 in part of cells S100+ and CD163+, CD43+ low. Negative lymphohematological markers included CD20, CD3,

CD8, CD2, CD1a, MPO, CD30, ALK1, MUM1, CD10, CD5, bcl2, PAX5& PAX8, CK19 & CK7, CD79a, Tdt, CD34, CD23, CD21, CD138, lambda/ kappa. Sarcoma NOS from spectrum Histiocytic/ dendritic cell sarcoma was proposed by us. The slides were sent for consultation to the reference centre in Germany, where lysozyme was found positive, and Histiocytic sarcoma with unusual expression of keratin was diagnosed. In the next centre in Warsaw, blastic plasmacytoid dendritic cell neoplasm was suggested. In NIH USA, additional IHC revealed negativity of CD123, CD14, CD117, TCF-4, but interestingly repeated S100 & CD163 were interpreted negative. Finally, Hematopoietic malignancy of uncertain lineage, with features of myeloid/ monocytic sarcoma and histiocytic sarcoma was diagnosed in USA. The oncologists continued therapy as for relapsed AML then. However, constant nodal progression, enlargement of hepatic and splenic metastases, and CNS involvement occurred. The patient died 9 months since the onset of symptoms. This case shows diagnostic differences and difficulties in several referential centers in such extremely rare neoplasm.

An Unusual Case of Intravascular Large B-cell Lymphoma- the autopsy study.

assoc. prof. Rafał Pęksa

Department of Pathomorphology, Medical University of Gdańsk,

An unusual case of Intravascular Large B-cell Lymphoma (IVL) is documented in a 59-year-old male electromechanic who presented with progressive neurological deterioration following hospitalization in Nigeria. The clinical course included cognitive decline, quadriparesis, and hepatomegaly. Laboratory investigations revealed elevated levels of C-reactive protein (CRP), D-dimers, lactate dehydrogenase (LDH), and β2-microglobulin, along with anemia and thrombocytopenia. Despite comprehensive diagnostic efforts—including neuroimaging, infectious disease screening, and consultations with multiple specialists—a definitive diagnosis was not reached during the patient's lifetime. Post-mortem examination uncovered cerebral edema with cerebellar tonsillar herniation and Duret hemorrhages. Histopathological examination identified the hallmark intravascular proliferation of large B-cell lymphoma cells, showing both cohesive and dyscohesive growth patterns within the lumina of blood vessels. This case highlights the diagnostic complexity of IVL, a rare entity initially described in 1959 and classified as a distinct lymphoid neoplasm in the 2001 WHO Classification of Haematolymphoid Tumours. It emphasizes the need to consider IVL in patients with progressive neurological symptoms and systemic laboratory abnormalities, even when initial diagnostic findings are inconclusive.

The list of discussed case studies, without abstracts:

- ALK-positive histiocytosis of the lung dr Aleksandra Sejda, University of Warmia and Mazury Faculty of Medical Science; Olsztyn
- Panniculitis- like T-cell lymphoma in a young man dr Dorota Ponikiewska, prof. Ewa Chmielik; PIB NIO Gliwice
- Adult T-cell leukemia/lymphoma prof. G. Ott, Stuttgart, Germany
- Aggressive NK cell Leukemia prof. G. Ott, Stuttgart, Germany
- Monomorphic epitheliotropic intestinal T-cell lymphoma dr M. Klimkowska, Stockholm, Sweden
- MYC/BCL6 pseudo-double hit lymphoma prof. G. Ott, Stuttgart, Germany
- Blastoid variant of mantle cell lymphoma masquerading as high grade B-cell lymphoma prof. G. Ott, Stuttgart, Germany
- Florid reactive lymphoid hyperplasia/lymphoma-like lesion of the female genital tract prof. G. Ott, Stuttgart, Germany
- Castleman's disease with indolent lymphoblastic T-cell proliferation dr. M. Klimkowska, Stockholm, Sweden
- Kikuchi-Fujimoto lymphadenitis dr. M. Klimkowska, Stockholm, Sweden
- High grade B-cell lymphoma with 11q aberration prof. G. Ott, Stuttgart, Germany
- BV+ diffuse large B-cell lymphoma prof. G. Ott, Stuttgart, Germany
- Reactive intralymphovascular blastic cell proliferation prof. G. Ott, Stuttgart,
- Indolent T -lymphoblastic proliferation in a tonsil of a girl dr. Jadwiga Małdyk, Medical University of Warsaw
- IgD- positive plasmacytoma in a tonsil of a boy dr. Jadwiga Małdyk, Medical University of Warsaw
- Enteropathy-associated T-cell lymphoma dr. M. Klimkowska, Stockholm, Sweden
- Systemic mastocytosis involvement in lymph node (or in GI tract) dr. M. Klimkowska, Stockholm, Sweden
- Diffuse large B-cell lymphoma, EBV-negative, after liver transplant, followed by plasmablastic lymphoma EBV-negative
 dr. M. Klimkowska, Stockholm, Sweden

dr. M. Klimkowska, Stockholm, Sweden

POSTER SESSION

"A case report- Co-occurence of follicural lymphoma and invasive breast carcinoma" Boroń-Wanot, D. Ponikiewska, E. Chmielik

Tumor Pathology Department, Maria Sklodowska-Curie, National Research Institute of Oncology, Gliwice Branch, Gliwice, Poland

Women with a history of breast cancer are diagnosed more frequently with other primary malignancies than the general population, but the simultaneous occurrence of breast cancer and follicular lymphoma is very rare and has been described in the literature only a few times. We would like to introduce a patient in whom a diagnosis of breast cancer was confirmed within 3 months of the diagnosis of follicular lymphoma.

A 63-years-old post-menopausal woman initially presented with symptoms of significant ascites. Laparotomy was performed and revealed massive tumor of the retroperitoneal space. Examination of the hepatoduodenal ligament lymph node showed effaced structure of the lymph node and raises suspicion of lymphoproliferative disorder. Immunohistochemistry examination was negative for CD3, CD5, IgD, CyclinD1 and positive for CD20, BCL2, BCL6, CD10, which indicated follicular lymphoma.

Imaging studies revealed not only uncountable, enlarged lymph nodes, but also showed a left breast lump. Ultrasound-guided biopsy of the left breast revealed triple negative invasive ductal carcinoma grade 3 (NG 3). In addition, small foci of lymphoid infiltration with abnormal immunophenotype and low proliferative activity Ki67 10% were presented in the breast.

Treatment of follicular lymphoma was initiated with the R-CHOP chemotherapy. The patient underwent left radical mastectomy and left axillary lymphadenectomy. Postoperative pathological examination of the left breast confirmed the presence of invasive ductal carcinoma. What is more, all axillary lymph nodes were involved by infiltration of follicular lymphoma positive for CD20, BCL2, BCL6, CD10, with Ki67 about 50%.

"Difficulties in diagnosing mediastinal lymphoma with unusual immunohistochemical profile". Anna Księżarek, Ewa Stobiecka, Sławomir Pakuło, Dorota Ponikiewska, Ewa Chmielik

Tumor Pathology Department, Maria Sklodowska-Curie, National Research Institute of Oncology, Gliwice Branch, Gliwice, Poland

The presented case shows how diagnosing neoplastic lesion from core biopsy can present a challenge. The limited tissue sample and possible aberrant immunohistochemical staining may become diagnostic pitfalls while differentiating between carcinoma and lymphoma. 63-year old female patient with history of left breast luminal carcinoma was referred to National Institute of Oncology due to enlarged upper mediastinal lymph nodes. In further radiological tests a supraclavicular lymph node was selected for core needle biopsy. The infiltrating cells were negative for GATA3, ER, PR, HER-2, synaptophysin, chromogranin, napsin A, CK7, CK AE1/AE3, SOX10, LCA, CD20 and positive for TTF1, vimentin, p53, focally EMA and CD30. The tissue sample was used entirely, with uncertain diagnosis varying between dedifferentiated carcinoma and anaplastic lymphoma.

Positive TTF-1 reaction proved to be aberrant in further testing from surgical biopsy of supraclavicular lymph nodes. The suggested diagnosis following morphological and immunohistochemical correlation was primary mediastinal large B-cell lymphoma. According to 5th ed. Of WHO Classification of Heamatolymphoid Tumours one of essential features of this entity is a

presence of large B-cell lymphoma in the anterior mediastinum. This was not valid to our patient which so far presented only nodal involvement. The disease progressed to liver and bone marrow, based on the liver biopsy it was stated that the whole image corresponds to: Diffuse large B-cell lymphoma, not otherwise specified (NOS). Positive TTF-1 reaction in large B-cell lymphomas is a rare phenomenon but some of the TTF-1 clones can present such an aberrant staining. The suggested diagnosis of PMBCL was not consistent with the clinical data as the patient was an elderly woman with only nodal presentation and with later confirmed liver and bone marrow involvement but no tumor in the mediastinum present. The literature states unusual PMBCLs presentation, such as rare cases of just nodal involvement in elderly patients but taking into consideration clinical manifestation and staging the diagnosis of DLBCL is more likely.

"Primary testicular follicular lymphoma in children."

Magdalena Łozińska, Jadwiga Małdyk, Aleksandra Sejda

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Lymphomas are the third most common pediatric malignancies. Childhood not-Hodgkin lymphomas represent a heterogenous group of tumors which rarely affects testis. We present a case report of 9-year old male admitted to the Departement of Pediatric Oncology and Heamtology due to painless enlargement of right testis. Ultrasound showed an hypoechoic, ill-defined, hypervascular mass covering approximately 2/3 of the right testis. MRI confirm this findings. Gross examination of removed testis revealed ill-defined, slightly nodular, beige tumor. H&E slides showed numerous reactive-appearing neoplastic follicles contain mainly centrocytes and a few centroblasts. Tumor cells were positive on CD20, PAX5, bcl6, CD23 and weakly MUM1 and CD10, whereas CD5, CD43, C-MYC, FOXP1, TdT, CD34, MPO, EBER, bcl2 were all negative. Finally, the diagnosis of follicular lymphoma was made. In cases when expansile, proliferative follicles are presented in tissue, follicular lymphoma has to be taken under consideration even in unusual locations.

"Pathomorphological changes of bronchopulmonary system in multiple myeloma"

Prof.Liliya Volos, Danylo Halytsky, Halyna Stoliar

Lviv National Medical University, Department of Pathological Anatomy; Lviv Regional Department of Pathology, Lviv, Ukraine

The study analyzes morphological changes in the lungs, bronchi, and pleura in patients who died from multiple myeloma (MM). Multiple myeloma is a malignant plasma cell tumor that causes secondary immunodeficiency, increasing susceptibility to infections, especially in the respiratory system. The aim of the study was to identify specific morphological changes in the lungs of patients with MM, based on a retrospective autopsy analysis of 16 patients. The immediate causes of death included chronic renal failure, pneumonia, and pulmonary heart disease.

The results revealed numerous changes in the respiratory system, including pneumonia, fibrosis, emphysema, uremic pneumonitis, enlarged lymph nodes, calcinosis, chronic bronchitis, amyloidosis, and intraalveolar hemorrhages. The findings indicate that interstitial calcium deposits without visible lung deformation posed a diagnostic challenge, despite modern diagnostic methods. The authors highlight that metastatic calcification of the lungs and myocardium was a significant factor contributing to mortality in patients with MM.

"Virchow's Node: Anatomy, History, and Clinical Significance".

Mateusz Michalski, Piotr Paluchowski, Grażyna Kobierska-Gulida

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Virchow's node, located in the left supraclavicular fossa, is a key element in oncological diagnostics. It was first described by Rudolf Virchow in 1848, linking it to gastric cancer metastasis. Later, Charles-Emile Troisier expanded these observations, connecting the node's enlargement with metastases from other cancers, such as lung cancer, prostate cancer, and lymphomas. Since then, an enlarged node has become known as "Troisier's sign."

Anatomically, Virchow's node lies deep in the left supraclavicular fossa, beneath the platysma and sternocleidomastoid muscles. It drains lymph from large areas of the body, including the left side of the head, neck, chest, abdomen, pelvis, and lower limbs. Lymph is channeled into the node through the thoracic duct, making it a site where cancer cells from various organs may accumulate. Clinically, an enlarged Virchow's node (Troisier's sign) often suggests metastases from advanced cancers, particularly those of the lungs, ovaries, or gastrointestinal tract. However, an enlarged node is not always indicative of cancer—it can also be caused by non-malignant diseases. Therefore, accurate diagnosis of Troisier's sign requires thorough clinical, imaging, and pathological examinations.

Complications related to the enlargement of Virchow's node include neurological syndromes such as unilateral phrenic nerve neuropathy, Horner's syndrome, and thoracic outlet syndrome. These complications arise from the node's proximity to vital structures like the brachial plexus and subclavian vessels, which can be compressed, leading to functional disturbances.

"Blood in the humoral theory" Piotr Paluchowski, Mateusz Michalski, Jacek Gulczyński, Dawid Ziemann

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The importance of blood has been recognized since ancient times, but detailed knowledge developed only with the invention of the microscope. Hippocrates introduced the humoral theory, which proposed that the human body was governed by four fluids: blood, yellow bile, black bile, and phlegm. Health was believed to rely on the balance of these "humors," and treatments like bloodletting and enemas were used to restore equilibrium. Ancient understandings of blood flow were flawed; for instance, Greek anatomist Erasistratus believed blood flowed through veins, while air passed through arteries, and Galen argued that blood properties varied by location in the body. The humoral theory remained influential throughout the Middle Ages and early modern period, with phlebotomy often scheduled based on astrological calendars. This theory also suggested that people could be categorized by their predominant humor, such as the sanguine type, associated with the qualities of blood and symbolizing energy and resilience.

The decline of humoral theory began with Anton van Leeuwenhoek's microscope in the 17th century and William Harvey's discovery of the circulatory system, marking the shift in medical thought that led to the eventual abandonment of humoral practices in Europe by the early 19th century.

"The *prehistory* of leukemia" Piotr Paluchowski, Mateusz Michalski, Jacek Gulczyński, Miłosz Chodyna

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One of the earliest descriptions of leukemia came from Peter Cullen in 1811, who described a patient with an enlarged spleen and milk-like blood serum. Cullen's use of mercury chloride treatment led to the patient's recovery, though limited medical knowledge at the time prevented a full understanding of the condition. In 1825, Alfred Velpeau observed "white blood" during an autopsy, noting a thick, white substance in the blood alongside an enlarged liver and spleen—marking the first autopsy description of leukemia.

John Hughes Bennett later examined the blood of a leukemia patient, publishing illustrations and coining the term "leukocytemia" in 1845. In the same publication, David Craigie detailed another spleen-related case, associating death with abnormalities in blood composition.

A breakthrough came with Rudolf Virchow, who in 1845 reported "white blood" in a woman's autopsy and coined the term "leukemia" in 1847. Additionally, Alfred Donné's 1842 discovery of platelets advanced microscopic blood analysis, revealing characteristics associated with chronic granulocytic leukemia, though he didn't link it directly to leukemia. Virchow's naming of the disease marked the beginning of a more structured approach to leukemia research, setting the stage for future discoveries.

"Risk of a Malignant Lymphoma After a Tattoo" Paulina Goździk, Marek Bucikiewicz, Miłosz Chodyna, Magdalena Górska-Ponikowska

Pathology and Neuropathology Dep., Medical University of Gdansk, Poland Dep.of Medical Chemistry, Dpt of Pathology & Neuropathology Medical University of Gdansk, Dpt. of Pathomorphology Copernicus Hospitals, Poland

Tattooing is a process of application of ink into the dermis. It has been widely performed for ages, as a form of art, self-expression, cultural or religious practice, or even for medical purposes. Nevertheless, there are still gaps in knowledge regarding the safety of tattoos and long-term consequences of their application. Notably, some research suggest their potential harmful effects. Tattoos may cause adverse reactions, for example: local infections, photodermatitis, eczema, exacerbation of psoriasis or atopic dermatitis. Lymphadenopathy in the region of new and old tattoos is not rarely encountered. Recent findings suggest that tattoos may be connected with malignant lymphoma.

A recent study conducted by Nielsen et al. brought novel insights to the topic: tattooed individuals have 21% higher adjusted risk of lymphoma, the highest lymphoma risk: <2 years between the first tattoo and the index year, the highest risk: diffuse large B-cell lymphoma and follicular lymphoma. Conclusions and further research directions: more studies are needed to identify the potential molecular mechanisms behind the increase of malignant lymphoma risk after a tattoo, getting a tattoo should always be carefully considered, especially if immunological or dermatological conditions are present.

"MRI in cutaneous lymphoma metastasizing to the brain" Agnieszka Sabisz, Mirosława Dubaniewicz, Edyta Szurowska, ***

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Introduction: Cutaneous lymphoma (CS), a rare form of non-Hodgkin lymphoma, primarily affects the skin, especially in individuals aged 55 and older. While central nervous system (CNS) involvement is uncommon, it is a critical consideration. T cell lymphoma is the most prevalent type, whereas B cell lymphoma, though less common, is more aggressive. MRI imaging of CNS lymphoma typically shows distinct features such as hypointense lesions on T1-weighted images and intense contrast enhancement on T1-weighted images with contrast. Differentiating CNS lymphoma from other conditions like metastasis, glioblastoma multiforme, and abscesses can be challenging due to overlapping clinical and radiological characteristics.

Case report background: A 64-year-old woman with cutaneous lymphoma in remission presented with headaches.

Initial MRI Findings: A focal lesion in the left thalamus (18x13x17mm) with diffusion restriction, low signal rim in SWI, and peripheral enhancement. Perfusion studies showed low rCBV and rCBF. Spectroscopy indicated NAA/Cr = 0.98, Cho/Cr = 1.9, Cho/NAA = 1.93, and a high lipid peak, suggesting an abscess.

Follow-up MRI Findings: New scattered foci up to 6 mm in both hemispheres, enhancing post-CM, mainly in the sulci and near the right ventricular triangle, with surrounding edema and lactates present.

Histology: The histopathological examination revealed a type of lymphoma different from the initial diagnosis: hgh grade B-cell lymphoma, EBV-positive, CD30+.

Conclusion: Regular neurologic follow-up is crucial for early detection and management of CNS involvement in cutaneous lymphoma, improving prognosis and quality of life.

"Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue of the dura mimicking meningioma: a case report" - Wiesława Grajkowska1, Katarzyna Czarnota2, Piotr Glinka3, Michał Sobstyl3, Grzegorz Rymkiewicz4, Teresa Wierzba-Bobrowicz1, Ewa Paszkiewicz-Kozik5

1Department of Neuropathology, Institute of Psychiatry and Neurology, Warsaw, Poland, 2Copernicus, PL, Gdansk, 3Department of Neurosurgery, Institute of Psychiatry and Neurology, Warsaw, Poland, 4Flow Cytometry Laboratory, Department of Cancer Pathomorphology, Maria Sklodowska-Curie National Research Institute of Oncology, Warsaw, Poland, 5 Department of Lymphoid Malignancies, Maria Sklodowska-Curie National Research Institute of Oncology, Warsaw, Poland.

The following case report describes a rare instance of MALT lymphoma, a low-grade mature B-cell neoplasm, developing in the dura. The patient was a 43-year-old woman who was admitted to the hospital after experiencing three tonic-clonic seizures on the same day. Neurological assessment revealed confusion and mild mixed-type aphasia. Magnetic resonance imaging (MRI) showed a contrast-enhancing, broad-based lesion along the dura mater in the left parieto-occipital region, raising suspicion of an en plaque meningioma. The

tumor was found to infiltrate the brain parenchyma, extending into the brain sulci. Significant brain edema surrounding the lesion caused a midline shift of 8 mm to the right. After stabilizing the patient's neurological condition with intravenous diuretics and steroids, surgery was performed. Pathological examination posed challenges in distinguishing MALT lymphoma from follicular lymphoma and required additional immunohistochemical (IHC) studies. Histological slides showed diffuse proliferation of small neoplastic cells in the dura, positive for LCA, CD20, PAX-5, MUM1 (positive in only 10% of cells), FOX-P1, and BCL2 antibodies. Tumor cells were negative for CD5, CD43, CD10, and BCL6 antibodies, and there was no restricted expression of either kappa or lambda light chains. The Ki-67 proliferation index was very low, approximately 1%. The diagnosis of dural MALT lymphoma was confirmed. Partial surgical resection, followed by R-CVP immunochemotherapy (rituximab, cyclophosphamide, vincristine, and prednisone), resulted in complete remission (with one year of follow-up). This case highlights that partial surgical resection combined with immunochemotherapy is a viable treatment option for these rare intracranial tumors. With only a little more than 100 cases reported in the medical literature to date, it is important to carefully evaluate each tumor to develop the best diagnostic and therapeutic approach.

