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[A1]

Meningiomas and other tumours of meninges and inter/perimeningeal spaces – a review of experience and examples of diagnostic difficulties based on our own material

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Meninges of the brain and spinal cord are a place of origin of tumours (mainly meningiomas) but also are an anatomical structure where non-meningeal tumours occur, both localized and diffuse (like leukaemic, carcinomatous or gliomatous infiltrations). Moreover, the diagnosing pathologist must always remember non-neoplastic tumour-like pathologies (e.g. haemangiomas, inflammatory reactions).

The subdural space (which is in fact “intermeningeal”) is a typical place of formation of chronic post-traumatic haematomas that result in a very active productive and inflammatory process leading to a characteristic membranous capsule surrounding extravasated blood and containing numerous different cellular components.

Well known is a rich phenotypic diversity of meningiomas and hence existence of many subtypes of this neoplasm. From a practical point of view, especially important is differentiation of rarer and more aggressive types like *meningeoma clarocellulare*, *m. chordoides* and *m. atypicum* and those regarded as the most aggressive types like *meningeoma papillare*, *m. rhabdoides* and *m. anaplasticum*.

Meningioangiomatosis is another important and interesting outgrowth which though being quite unique may be mistaken for infiltrating (invading) meningioma. The type of the outgrowth of meningioma and its relation to brain parenchyma (invasion vs. “adhesion”) also matters. In cases of especially diffuse infiltration within meningeal spaces with encroachment of brain and its feeding blood vessels (which supposedly could be called “meningiomatosis”) total resection is either almost impossible or burdened with high risk of serious complications.

Artifactual changes in neoplastic tissue caused by high temperature (electrothermic damage due to coagulation of bleeding vessels during surgery) may lead to mistakes in diagnosis. This sort of artefacts may even lead to the mistaken diagnosis of carcinoma instead of meningioma and vice versa. Arachnoid cyst and many other cystic non-malignant lesions that can appear in meninges and in the borderline zone with brain and spinal cord parenchyma make a separate group of pathologies which the diagnosing pathologist should be aware of. The aforementioned meningeal and perimeningeal/intermeningeal pathological changes will be presented and discussed on the basis of own material.

[A2]

Optic nerve sheath meningioma – a report of 3 cases

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We describe 3 cases of optic nerve sheath meningiomas (ONSM). All the patients were 14-year-old girls at the time of surgery. The common feature of the clinical cour-

se was that progressive loss of vision preceded the diagnosis on imaging studies (CT, MRI). The first case with type 2 neurofibromatosis (NF2), and type IIb location of tubular primary ONSM (according to Schick, Dott, Hassler) with diffuse, grass-like intracranial extension underwent subtotal resection of the transient meningioma without subsequent radiotherapy. No tumour progression was observed in 3 years of follow-up. The second girl without NF2 suffered from a type IIIb tumour. She underwent only palliative partial resection of the psammomatous, nodular (globular) meningioma and was subsequently locally irradiated, also without progression of the disease during 3 years of follow-up. The third girl had severe expression of NF2. Near-total resection of type IIb nodular, globular ONSM was performed. Transient meningioma was diagnosed on pathological examination. No radiotherapy was performed because the patient had refused her consent. During the period of 8 years we could observe very extensive intracranial progression, and the tumour can be classified now at least as IIIb Schick, Dott and Hassler type. The 2nd case could also be considered as a tuberculum sellae meningioma with secondary intraorbital extension, and the 3rd case as group 3 of Al-Mefty's clinoidal meningioma. ONSMs in children occur frequently in NF2 patients but sporadic cases are also observed. Classifications of ONSM locations including Schick, Dott and Hassler's are first of all gradings of clinical progression. The problem of treatment strategy including indications and timing of surgery and radiotherapy in children is still being discussed.

[A3]

Clear cell meningioma – diagnostic difficulties

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Clear cell meningioma is an unusual subtype of meningiomas that affects children and young adults. The cerebellopontine angle and *cauda equina* region are the most frequent localizations of this tumour. Clear cell meningioma is associated with more aggressive behaviour and frequent recurrences in spite of its typically benign-appearing histopathology and corresponds to WHO grade II. Clear cell meningioma is composed of sheets of elongated or oval cells with clear, glycogen-rich cytoplasm and prominent perivascular and interstitial collagen. Because of its clear cell morphology, the correct diagnosis might be difficult. In differential diagnosis tumours of different histogenesis ought to be considered, i.e. microcystic meningioma, low grade astrocytoma or metastatic renal carcinoma.

[A4]

Intrahemispheric papillary meningioma in a ten-year old girl – clinical implications

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A 10-year old girl was admitted to the Department of Paediatric Neurosurgery of the Silesian Medical University in Katowice due to one-week history of headaches and divergent squint. CT and MRI examinations revealed a large tumour involving the left parietal and temporal lobes surrounded by brain oedema. The child was operated on through left temporal craniotomy. Gross total tumour resection was performed. In histopathological examination diagnosis of papillary meningioma, WHO Grade III was established. After the surgery the girl was treated with chemotherapy according to the current protocol for high-grade paediatric brain tumours. At the same time radiation therapy was performed; a total dose of 54 Gy was delivered with 1.8 Gy per fraction to the tumour bed with margin. After 8 months' follow-up the girl has no new neurological deficits and in control MRI examinations no signs of recurrence were detected. The authors present clinical consideration and treatment options of this very rare

type of malignant meningioma which was located within the cerebral hemisphere.

[A5]

Mast cells in meninges and brain tissue in neurocysticercosis

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Mast cells are a heterogeneous cell population, whose precursor cells undergo development and differentiation in response to local tissue requirements. Thus, in different body sites, mast cells reveal heterogeneous morphology and size of cytoplasmic granules.

Mast cell heterogeneity can encompass significant differences in the cell mediator content, responses to proliferation/survival factors, and patterns of sensitivity to agents that induce or pharmacologically modulate mast cell degranulation and release their powerful mediators to the extracellular environment. The secretory granules of human mast cells contain various amount of two proteolytic enzymes: chymase and tryptase. These two enzymes differentiate human mast cells from other immune cells. The relative content of chymase and tryptase in mast cells can also distinguish two main phenotypes of these cells. Mast cells containing both enzymes belong to tryptase-chymase phenotype (MC--TC) and those that contain mostly tryptase are considered as cells of the tryptase phenotype (MC-T) .

In normal human brain mast cells are tryptase-chymase phenotype, few in number and can be found in the meninges, choroids plexus and in the perivascular area of some blood brain vessels. It is now generally accepted that these cells in the normal brain mediate alternations in blood flow, neurotransmission and local immune responses.

The aim of the present study was to examine phenotype, number and distribution of mast cells in human brains with neurocysticercosis.

The study was performed on brain specimens fixed in formalin and embedded in paraffin. Mast cells were identified with two monoclonal antibodies generated against human tryptase and chymase.

The immunohistochemical technique for mast cell localization was compared to other conventional stainings that include acidified toluidine blue, alcian blue microwave and sudan black B. The mast cells were counted at high power field (HPRx400) for each specimen. The data were pooled and mean values were calculated.

The results show that numerous tryptase phenotype mast cells infiltrate the areas of meninges and brain parenchyma which were infested by cysticerci. The cells were accumulated around cysts containing parasites, in the wall of blood vessels and were scattered in different inflammatory cell types infiltrations. Mast cells demonstrated various morphological appearances, from cells showing densely stained intracellular tryptase to those showing extracellular release of tryptase-positive granules and cells shrunken following degranulation. Such observations suggest that some mast cells undergo activation/degranulation at the infected brain regions . In non-infected regions of the brain, number and phenotype of mast cells were as in the control, normal brain age-matched patients.

In summary, the results of the present study confirm our previous findings that reactive mast cells infiltrating the brain under pathological circumstances are numerous and tryptase phenotype. In neurocysticercosis, the presence of these cells is clearly connected with the local defence reaction of the brain to the toxic effect of the parasite.

[A6]

Rhabdoid meningioma with advanced histological malignancy – case report

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Rhabdoid meningioma is a rare, aggressive variant of meningioma with a great risk of recurrence. It was included in the 2000 WHO classification of tumours

of the nervous system as a subtype corresponding to WHO grade III.

We present a case of rhabdoid meningioma in a 51-year-old man. MRI exhibited a large tumour mass located on the cranial base and right cerebral hemisphere. Histologically, the tumour showed large parts of rhabdoid morphology accompanied by conventional meningioma of various morphological pattern. The central part of the tumour was composed of necrotic tissue containing well-preserved sheets and clusters of typical rhabdoid cells. These cells exhibited large, vesicular, often eccentric nuclei with distinct nucleoli and abundant cytoplasm containing cytoplasmic eosinophilic inclusions. Rhabdoid tumour cells were positive for vimentin and S-100 protein and showed focal epithelial membrane antigen and cytokeratin expression. The rhabdoid parts of the tumour exhibited a high proliferation rate (80% of MIB1-positive cells).

The presented case documents that rhabdoid phenotype represents features of malignant transformation in meningiomas.

[A7]

Posterior cranial base meningiomas

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We present our experience in the surgical treatment of posterior cranial base meningiomas with special consideration to the evaluation of the surgical outcome and operative technique.

A series of 30 patients treated surgically for posterior cranial base meningioma in the Department of Neurosurgery, Medical Academy of Warsaw in the years 1997-2006 is reported. There were 17 patients presented with posterior surface of the pyramid meningiomas, which were further divided into anterior pyramid, posterior pyramid and jugular foramen meningiomas (9, 5 and 3 patients, respectively). Furthermore, there were 8 petroclival meningiomas and 5 foramen magnum meningiomas. Combined supra- and infratentorial approach and retrosigmoid approach were suitable for petroclival and posterior pyramid meningiomas. We used far lateral retrocondylar approach for foramen magnum meningiomas. Meningiomas were resected incompletely (Simpson IV surgery) in 10 patients (six of them were petroclival meningiomas); in the other patients complete tumour removal was made (Simpson I and II surgery).

Postoperative facial nerve dysfunction appeared in 11 and further deteriorated in 2 patients. Trigeminal hypesthesia appeared as a new finding in 5 patients and 4 patients showed aggravation of pre-existing deficit. Lower cranial nerve deficits occurred in 5 patients. In our series, hemiparesis developed in 4 patients, abating in three within a few weeks. Perioperative death occurred in four patients: two of them were large petroclival meningiomas, another one was posterior pyramid and the last one was foramen magnum meningioma. All other patients were in good clinical and neurological condition on discharge from hospital. Fortunately, many patients with cranial nerve deficits showed some evidence of improvement in the follow-up.

Surgical treatment of posterior cranial base meningiomas has become increasingly safe; however, these tumours still remain a surgical challenge because of the relatively high incidence of permanent complications associated with their removal.

[A8]

Operative treatment of parasagittal and falx meningiomas: prognostic factors and long-term results

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The purpose of this study is to evaluate results, prognostic factors and complications following surgical treatment of parasagittal and falx meningiomas. A retrospective analysis of 87 consecutive patients with parasagittal and falx meningiomas operated on at the Department of Neurosurgery, Medical University of Warsaw between 1987 and 2001 is presented.

There were 50 parasagittal and 37 falx meningiomas (17 bilateral falx meningiomas). 4 patients included in the series had atypical and 8 patients had anaplastic meningiomas. Meningothelial (27 cases) and fibrobla-

stic (22 cases) meningiomas were the most common benign meningiomas. The tumour resection was graded according to Simpson's classification. In this series, 25 procedures were Grade I (complete tumour removal with resection of dura and abnormal bone), 55 were Grade II (complete tumour removal with coagulation of dural attachment) and 7 were Grade IV (subtotal tumour removal).

Early postoperative results were assessed according to the Glasgow Outcome Scale (GOS) and Karnofski index. 76 (87%) patients had satisfactory outcome (good recovery or mild disability and Karnofski index values of over 70 points) on discharge from hospital, 7 (8%) patients were disabled and 4 (5%) patients died. Bilateral falx meningioma and incomplete removal of tumour were significant predictive factors for unfavourable outcome. Lack of focal deficits and location of meningioma beyond eloquent areas were significantly related to very good outcome (good recovery). In 34 (39%) patients the early course after the operation was disturbed. In 24 patients postoperative temporary deterioration was recorded. In 6 patients neurological deficits remained permanent and 4 patients died. Analysis revealed two main mechanisms of postoperative deterioration: haemodynamic complication and deterioration after resection of tumour in the eloquent area. Haemodynamic complications were the main cause of postoperative death. Intraoperative obliteration of the patent superior sagittal sinus was an independent factor influencing haemodynamic complications.

The follow-up period ranged from 22 to 196 months with a mean of 87 months. Late results were achieved in 79 out of 83 (95.2%) patients discharged home. Tumour recurrence is the most important factor influencing late results of operative treatment. At the time of analysis 14 (18%) patients showed evidence of recurrence. Tumour recurrence was a significant factor for unfavourable late outcome. Male gender, partial removal of meningioma (Simpson Grade IV) and bilateral falx meningioma had a statistically significant influence on recurrence.

[A9]

Optic nerve sheath meningioma (ONSM)

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Meningiomas represent approximately 1.7% of all orbital tumours and 35% of all intrinsic tumours of the optic nerve. Optic nerve sheath meningiomas (ONSM) account for 1 to 2% of all meningiomas. Classic primary ONSMs originate in the orbit and may extend along the optic canal intracranially. Secondary ONSMs extend from the planum sphenoidale into the subdural or subarachnoid spaces surrounding the nerve within the optic canal and, ultimately, within the orbit. Tumours originating at the optic foramen represent group 3 of Al-Mefty's clinoidal meningiomas. Optic nerve sheath meningiomas occur predominantly in middle-aged women or in children. In children a more aggressive course has been noticed. Schick, Dott and Hassler divided ONSMs according to their location into 3 types: type I lesions are located purely intraorbitally, type II lesions are located intraorbitally with extension through the optic canal or superior orbital fissure, and type III are located intraorbitally with far-reaching intracranial extension. Intracranial extension could be diffuse grasslike (65%) or nodular (35%). Saeed et al. described two distinct types of growth within the orbit: tubular, characterized by minimal widening of the length of the optic nerve and more frequently with evidence of calcification; and globular, with growth outside the dural sheath. An optic nerve sheath meningioma is a benign tumour whose management remains controversial. Surgical resection should be reserved for patients who are already blind or who have severe exophthalmos or intracranial involvement because surgery is associated with a significant rate of morbidity, including blindness or total loss of remaining vision. The standard surgical approach is the intradural pterional approach with decompression of the optic canal and no resection of the intraorbital flat tumour around the optic nerve. The extradural approach with posterior orbitotomy is restricted to purely intraorbital meningiomas. Radiotherapy decreases the rate of local failure or progression after subtotal surgery to less than 20% at 5 years. Irradiation of the growing orbit may lead to hypoplasia and gross facial asymme-

try in children as well as cataracts and retinopathies in various age groups. In newer techniques such as three--dimensional conformal radiotherapy and stereotactic radiosurgery, practitioners seek to decrease these risks by minimizing irradiation of normal tissues; however, further studies of these modalities are needed. The loss of vision in patients with ONSM is only a matter of time. In patients with good vision the role of radiotherapy becomes more important. Surgery is recommended for intracranial tumours to prevent contralateral extension. Patients with ONSM receiving radiation alone demonstrated the best visual outcome during the follow--up period. We recommend that fractionated external beam radiation (5000-5500 cGy) be considered as an initial treatment in adults in selected cases of ONSM when preservation of visual function is a reasonable therapeutic goal.

[A10]

Idiopathic hypertrophic pachymeningitis

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Idiopathic hypertrophic pachymeningitis (IHPM) is a rare pathological state, with still unclear aetiopathogenesis. We present a case of a 63-year old woman with cranial variety of that disease. The manifestations of the disease included headaches, paresis of VI, IX, X nerves and cerebellar ataxia. The disease was diagnosed with magnetic resonance imaging (MRI) and histopathological assessment of the pachymeninx biopsy specimen. The MRI revealed significant thickening of the cranial basis pachymeninx, compressing the pons and medulla oblongata. MRI examinations could be misinterpreted as extensive meningioma of the skull base. Dura mater biopsy revealed however inflammation with abundant lymphocytic infiltrations. Clinical improvement was obtained after the application of corticosteroids. We noted the subsidence of all symptoms of the disease, as well as radiological improvement, manifested through substantial regression of the described changes in the pachymeninx. The patient is presented in the context of 65 cases of idiopathic hypertrophic pachymeningitis, described in the literature of English-speaking countries in the last five years. Recently, the importance of autoimmunogenic background of IHPM has been underlined. In that respect IHPM becomes an interdisciplinary problem. Its diagnosis and treatment requires not only radiologists, neurologists, pathomorphologists and neurosurgeons, but also specialists in internal medicine, including immunologists, allergologists and rheumatologists as well – in other words, physicians that rarely take part in the diagnosis and treatment of intracranial pathologies.

[A11]

Lectin histochemistry and ultrastructure of secretory meningiomas

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Secretory meningioma is a rare variant of benign meningiomas of grade I malignancy, characterized by appearance of pseudopsammoma bodies in the histological pattern. In this report we present an examination of lectin glycoconjugates and ultrastructure of pseudopsammoma bodies in biopsy material of 7 cases of secretory meningiomas. Clinically, there were female patients aged from 50 to 73 years; the tumours involved the region of the sphenoid ridge in 4 cases, frontal convexity in 1 case and temporal lobe in 1 case and were associated with prominent peritumoural oedema in 5 cases. Histologically all tumours revealed a multitude of eosinophilic globular pseudopsammoma bodies, strongly stained by PAS and immunopositive for carcinoembryonic antigen (CEA). The biotinylated lectins used for the study, namely Peanut agglutinin (PNA), Soybean agglutinin (SBA), Dolichos biflorus agglutinin (DBA), Wheat germ agglutinin (WGA) and Concanavalin A (Con A), showed different reactivity with pseudopsammoma bodies and tumour cells. Binding by PNA and WGA was intensive in both pseudopsammoma bodies and within

surrounding tumour cells and resembled the expression of CEA. Staining with Con A and DBA was positive in some pseudopsammoma bodies but negative in tumour cells. SBA stained mostly tumour cells bearing small pseudopsammoma bodies. Ultrastructurally, intracellular membrane-bound lumina lined by microvilli and filled with fine-granular osmophilic or vesicular material were observed in tumour cells. Their cytoplasm consistently showed high electron density, well developed Golgi complex and many tonofilament fascicles. Our studies confirm previous reports indicating unique epithelial and glandular differentiation of tumour cells and further characterize the secretory products of these cells as glycoproteins, particularly rich in galactose and N-acetylglucosamine residues.

[A12]

Intacranial haemangiopericytoma, related to meninges

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Haemangiopericytoma (HPC) of the central nervous system (CNS) is a vascular neoplasm derived from the pericytes. HPS is a highly cellular and richly vascularised tumour, frequently attached to the dura mater. It can be classified as WHO grade II or III. HPC is not specific radiologically, but contains numerous vessels with angiography.

Two cases of haemangiopericytoma are reported. Patient 1, aged 58, had symptoms, such as progressive visual disturbances, vertigo, dementia, gait abnormalities and urinary incontinence, that began 3 month before the operation. Cranial CT scan showed masses of tumour in the right region of the fronto-parietal lobes and in the right base of the temporal lobe. Patient 2, aged 56, complained about headache and symptoms of epilepsy. Extensive parasagittal tumour in the left region of the fronto-parietal lobes was demonstrated on CT scan. In both these cases changes were related to the dura mater; therefore menigioma was suggested.

The neoplasms were removed neurosurgically. The postoperative course was complicated by the presence of blood in post-neoplasm regions.

Both removed tumours were studied with histological and immunohistochemical methods. Paraffin-embedded fragments from tumours were examined with antibodies against vimentin, GFAP and CD34, Ki-67, EMA.

Morphological examination of the surgical specimens revealed a pattern of compact and looser texture areas of oval or spindled cells and numerous vessels. Neoplastic cells proliferated between vessels. The cells usually had scanty cytoplasm and indistinct cell boundaries. The reticulin stain demonstrated reticulin surrounding the cell and vessels. Antibodies CD34 and vimentin showed positive immunoreaction with neoplasms and endothelial cells. The reaction with antibodies GFAP and EMA were negative. Proliferation index Ki-67 was lower than 4%. The tumours were diagnosed as haemangiopericytomas.

Hypervascularity seen on MRI angiography may distinguish haemangiopericytoma from meningioma. Differentiating between these two neoplasms before an operation is essential for preventing postoperative complications.