A case of cystic form of angiomatous meningioma with prominent microvascular pattern mimicking haemangioblastoma

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A surgical case was reported of an unusual angiomatous variant of meningioma with predominant microvascular component and extensive cystic changes. The tumour was incidentally detected in a 79-year-old woman who was admitted to the hospital because of a head injury. The CT scan revealed in addition to bilateral subdural haematomas a large-sized (5 × 5 × 4 cm) multicystic tumour with a solid contrast-enhancing nodule in the right frontal region. Microscopically, the tumour tissue was composed predominantly of a dense meshwork of small, capillary-like and thin-walled dilated blood vessels and a relatively small component of intervening meningotheliomatous tumour cells. The resemblance of the presented case to some rare cases of cystic meningioma which were formerly classified as a haemangioblastic variant of meningioma or transitional forms between meningioma and haemangioblastoma is briefly discussed.

Key words: angiomatous meningioma, cystic meningioma

Cystic meningiomas are relatively uncommon tumours [2, 5, 6, 8, 24] which may mimic other neoplasms associated more frequently with cystic lesions such as gliomas [5, 6, 8, 9], metastatic tumours [6, 19] or haemangioblastomas [3, 5, 16]. Various histological variants of meningioma could demonstrate cystic changes but the meningothelial type has been found most frequently in the published series of cystic meningioma cases [3, 8, 19, 25]. Richly vascularised meningiomas of the angiomatos type were reported only in a few cases [1, 16, 24]. On the other hand, many cases in the reviewed series of cystic meningioma were histologically designated as the angiolastic type [2, 3, 8, 19, 24].

However, the term “angioblastic meningioma” has been used in the older literature for a broad group of highly vascularised tumours of the meninges comprising both angiomatic meningioma as well as haemangiopericytoma, supratentorial haemangioblastoma and some transitional cases between meningioma and haemangioblastoma [10, 12, 17, 20, 21]. Many reported cases of cystic meningioma termed as angiolastic type either failed more precise histologic subclassification or were synonymous with meningeal haemangiopericytoma. Since the two last WHO classifications of the tumours [13, 14] the histologic term “angioblastic meningioma” has been discarded as a designation of some meningeal neoplasms and the cases of highly vascularised tumours of meningothelial derivation are classified as angiomatous meningiomas. The literature concerning the angiomatic variant of meningioma is very sparse [11, 18] thus it seems to be justified to describe a case of the microvascular subtype of angiomatous meningioma in
association with prominent cystic changes, demonstrating some resemblance with supratentorial haemangio-blastoma in the preoperative and histologic pattern.

**CASE REPORT**

A 79-year-old woman was admitted to the emergency room after a head injury with a left-sided hemiparesis. She did not complain about any neurological deficit prior to the traumatic event. The admission CT scan of the head revealed thin, subdural haematomas bilaterally along with a large-sized (5 × 5 × 4 cm) tumour in the convexity of the right frontal region (Fig. 1A, B). The tumour was multicystic with a solid, contrast-enhancing nodule. The patient was operated upon on an emergency basis due to the progressive deterioration of her neurological status. After a frontal craniotomy a dural attachment of the lesion was found, fed by a pathological branch of the middle meningeal artery. Arterial supply of the tumour came also from the ipsilateral pericallosal artery. A puncture of the cysts containing a yellowish, transparent fluid facilitated debulking of the tumour mass. The tumour was excised completely with its dural attachment. The patient made a good recovery in the postoperative course. At discharge her neurological deficit resolved almost completely.

**MATERIAL AND METHODS**

The study was performed on the surgical material fixed in 10% formalin and embedded in paraffin. The sections were stained with haematoxylin and eosin (HE) and by Gomori’s and van Gieson methods. For immunohistochemistry the sections were stained by PAP method with DAB as chromogen and with the primary antibodies against vimentin (VIM), epithelial membrane antigen (EMA), factor VIII-related antigen (F-VIII), glial fibrillary acidic protein (GFAP), S-100 protein, (all products from DAKO) and CD34 antigen (from Immunotech).

**RESULTS**

On gross examination the smooth surfaced tumour showed vascular branches running on the surface (Fig. 2A) and large multilocular cysts with peripherally located brown-reddish nodule about 1.5 cm of length, on the cut sections (Fig. 2B).

Microscopically, the nodule exhibited a dense vascular meshwork interconnecting at many places with bands of hyalinised tissue forming the fibrous septa within central cystic spaces (Fig. 3).

Both the internal and external walls of cysts showed the presence of adhering vascular tissue of the tumour. The major part of the tumour tissue was composed of mostly small, capillary-like and thin-walled, often dilated blood vessels (Fig. 4), highlighted by reticulin fibres impregnation (Fig. 5), and of a relatively small proportion of the tumour cells between vessels. Some peripheral regions of the nodule displayed more numerous tumour cells presenting features of those of meningothelial meningioma, intervening with many small vessels and arterioles (Fig. 6) as well as large, thick-walled and hyalinised blood vessels (Fig. 4). In other areas, the cells with vacuolised cytoplasm and indistinct cytoplasmic borders were seen in the interstices between small blood vessels (Fig. 7). These cells were immunopositive for VIM, S-100 protein and EMA, whereas they were negative for GFAP and for the vascular markers used, F-VIII and CD—34 antigen, which disclosed immunoreactivity with the endothelia of blood vessels.

*Figure 1A.* Higher CT scan showing multicystic tumour with a contrast-enhancing nodule in the right frontal region. *B.* Lower CT scan showing thin, subdural haematomas bilaterally (arrows) along with a midline shift to the left side.
**Figure 2A.** Gross appearance of the tumour. **B.** On cut section large multilocular cysts surrounded by fibrous septa and peripherally located tumour tissue (x). Normal size.

**Figure 3.** Richly vascularised tumour tissue interconnecting fibrous septa within cystic spaces. HE, × 20.

**Figure 4.** Tumour tissue with vascular component of dilated, thin-walled, predominantly small blood vessels (top) and meningiomatic part comprising large hyalinised vessels (bottom). HE, × 100.

**Figure 5.** Dense aggregates of small blood vessels with dilated lumina and thin walls. Gomori's method, × 200.

**Figure 6.** Scarce meningothelial tumour cells between numerous capillary-like blood vessels and small arterioles. CD-34 immunostaining, × 200.
DISCUSSION

The reported case of meningioma presents angiomatous and cystic patterns, both of which rarely occur in meningiomas. The incidence of cystic meningiomas is of about 2–4% [2, 5, 6, 8] to 7.3% [24]. In this group of meningiomas different secondary cystic lesions of the tumours with both intratumoral and peritumoral location are included [1, 3, 5, 8, 24, 25]. The “true” intratumoral cysts in association with highly vascularised meningiomas have been thought to be exceedingly rare, accounting for about 0.2% [7] of all meningiomas. Although most of the reported cases of highly vascularised cystic meningioma were defined in the past as angioablastic meningioma and were indistinguishable from haemangioblastoma and haemangiopericytoma, there were some cases that could be reclassified as an angiomatous variant of meningioma [3, 24]. Angiomatous meningioma, when separated from the other types of angioablastic meningioma, constitutes 5.2% of all intracranial meningiomas and is found almost exclusively as convexity tumour [12]. According to the general recommendation for the revised WHO classification of meningeal tumours, the angiomatous meningioma is characterised by the abundance of well-formed vessels, sinusoids or capillaries in association with histologic pattern of meningioma usually of the meningotheliomatous type [4, 22]. Recently, Nolte and Paulus [18] reported the largest series of 40 cases of angiomatous meningioma and recognised two subtypes of this neoplasm, namely, a microvascular subtype with predominant component of small vessels (more than 50% with a diameter of below 30 µm) and a macrovascular subtype. In the microvascular subtype, the authors stressed frequent microcystic changes, foamy cells and a pattern resembling capillary haemangioblastoma.

The presented case, with high microvessularity, vascu- lisation of the tumour cell cytoplasm suggesting foamy cells and intrinsic cystic changes, shows also an initial similarity to supratentorial haemangioblastoma. In addition, the radiological feature of a large cyst with contrast-enhancing mural nodule mimics also a feature of haemangioblastoma. Particularly, the pre-operative appearance of cystic meningioma and supratentorial haemangioblastoma should be indistinguishable, since the tumours with dura attachment have been considered in some cases of supratentorial haemangioblastoma [7, 17].

Although angiomatous meningioma and haemangioblastoma are both classified as benign neoplasms (WHO grade I) and the distinction between them seems to be irrelevant for therapeutic purpose, it is important for patients with occurrence of supratentorial haemangioblastoma, which almost always implies the diagnosis of Von Hippel-Lindau disease [17]. Based on histogenetic criteria, haemangioblastoma is presently separated from the group of meningiomas and is classified as a meningeal tumour of unknown origin [13, 14]. In contrast, the cases formerly classified as a transitional form between haemangioblastoma and meningioma should be diagnosed as angiomatous meningioma, as far as the histological pattern of any meningioma variant is recognised in addition to haemangioblastoma-like features [4].

In the presented case, the histologic and immuno- histochemical evidence of meningothelial tumour cells prejudged a diagnosis of angiomatous meningioma with a predominant microvascular component. The accompanying large cysts could be regarded as secondary changes related to vascular permeability and transudation of plasma fluid resulting in micro- and macrovacu- itation of the tissue. A similar mechanism for the forma- tion of cysts has been previously postulated in microcystic and richly vascularised meningiomas [15, 23]. In our case the asymptomatic clinical course prior to the head injury might suggest a long-lasting process of development of the large cysts within the tumour with otherwise benign biological behaviour.
REFERENCES


