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LETTER TO THE EDITOR

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Ribosome-lamella complex in aggressive solitary fibrous tumour of the meninges

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Approximately 60 cases of solitary fibrous tumour (SFT) have been reported in the meninges [6]. Only a few cases have been malignant with local recurrences being the most common manifestation [2, 4, 6].

We report the biopsy and autopsy findings of a malignant SFT demonstrating multiple intracranial recurrences over a 17-year period, meningeal spread and invasion into brain tissue. The presence of ribosome-lamella complexes (RLC) in an SFT is described for the first time.

In 1985 a 59-year-old man presented with dizziness. A left-sided tentorial tumour compressing the cerebellum was completely removed. A recurrent tumour was subjected to surgery 13 years later. The patient died from acute myocardial infarction in 2002, 4 days after surgical resection of a leptomeningeal tumour compressing the cervical portion of the spinal cord (C7 level).

Samples from the three tumours removed over the 17-year period were fixed and embedded in paraffin wax according to our routine protocol. In addition to routine stainings, immunohistochemical analyses were performed using primary antibodies against the following epitopes: CD34 (Novocastra Laboratories, Newcastle upon Tyne, UK), bcl-2 (Novocastra), desmin (Dako, Glostrup, Denmark), epithelial membrane antigen (EMA, Novocastra), S-100 (Dako), muscle actin (Dako) and Ki67 (Dako). The use of McDowell's fixative precluded successful Ki67 immunostaining of the oldest biopsy specimen. Electron microscopic examination was performed in all cases.

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The histological appearances of the three surgical samples were almost identical. The tumours were composed of elongated cells arranged in fascicles between prominent eosinophilic bands of collagen (Fig. 1). No whorls, psammoma bodies, or necroses were observed. Mitoses were absent in the original tumour, whereas the first tentorial recurrence in 1998 demonstrated seven, and the intraspinal tumour from 2002 ten mitoses per ten high-power fields.

Immunohistochemical analyses demonstrated strong and uniform expression of CD34 and bcl-2. Scattered cells were positive for desmin. All samples were negative for EMA, S-100 and muscle actin. Approximately 5% of the cells in the recurrence from 1998, and 10% of the cells in the intraspinal tumour from 2002, expressed Ki67. Electron microscopic examination of the tumours from 1998 and 2002 showed cells without basal laminas connected by desmosome-like structures and surrounded by collagen fibres. Interdigitating cytoplasmic processes, typical of meningiomas, were not observed. The cytoplasm of the spindle-shaped tumour cells contained prominent rough endoplasmic reticulum (RER) arranged in concentric, multilaminar structures consistent with RLCs (Fig. 2). The RLCs were numerous and found in more than 50% of the tumour cells. Similar structures were not observed in the original tumour.

Autopsy findings demonstrated four tumours in the posterior fossa; a small recurrence of the left-sided tentorial tumour (diameter 0.8 cm) and three leptomeningeal tumours (2.1, 2.0 and 0.3 cm, respectively) outside the original operation field. Two of the latter compressed the brain stem (Fig. 3). A darker area inside the left inferior colliculus was noted.

The histopathological findings in all tumours diagnosed at autopsy were identical to those in the surgical specimens described above. Microscopy of the darker area of the left inferior colliculus showed an identical tumour within the brain tissue (Fig. 4). Mitoses were not observed in any tumour. The Ki67-index was less than 1%, but the other immunohistochemical findings were

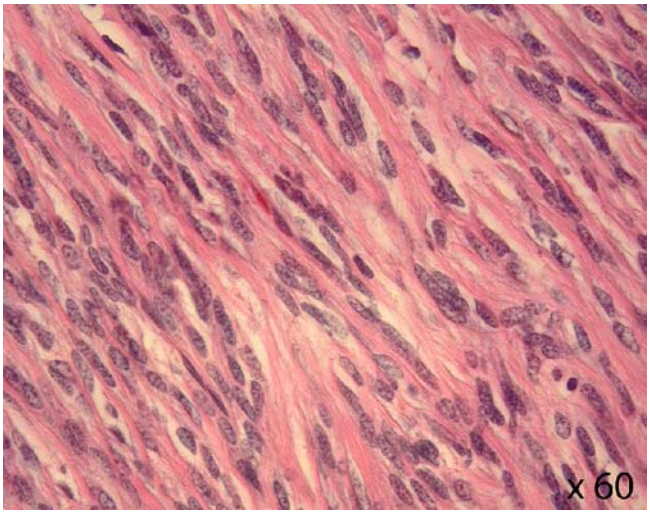

Fig. 1 Elongated cells are arranged in fascicles between prominent eosinophilic bands of collagen. Haematoxylin and eosin, ×60

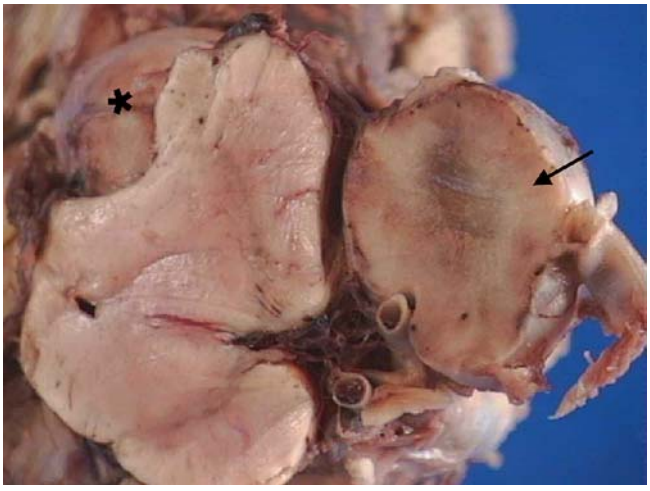

Fig. 3 Autopsy findings: Two leptomeningeal tumours are compressing the brain stem. One (marked with an *asterisk*) is located within a groove in the left, posterior portion of pons. The other (marked with an *arrow*) is located anterior to the left cerebral peduncle

the same as in the surgical samples. Electron microscopic examination was unsuccessful due to autolysis.

SFTs of the central nervous system show a predilection for the posterior fossa and the spinal canal [4]. Most intracranial SFTs show a dural origin but tumours located in the spinal canal often lack dural attachment [4]. In our case the primary tumour was clearly in continuity with tentorium cerebelli but the subsequent lesions were merely leptomeningeal.

SFT is characterised by fascicles of elongated cells that alternate with paucicellular, collagen-rich tissue. Whorls and psammoma bodies are absent and mitotic figures are usually scant [8]. CD34, which is considered the most sensitive immunohistochemical marker [8], has been demonstrated in dural fibroblasts and SFT cells may represent the neoplastic counterpart of such cells [3]. SFTs usually demonstrate positive staining for vimentin and bcl-2, but EMA or S-100 is not expressed [4]. Focal staining for desmin has been reported [4].

Electron microscopic examination of SFTs has demonstrated fibroblast-like tumour cells with well-developed RER. Cytoplasmic interdigitations and well-formed desmosomes are usually lacking, although occasional primitive junctions have been described [8], as in our case. Basal laminas are not present [5].

In our case the diagnosis of SFT was established by routine histological findings and positive staining for CD34. Absence of whorls, psammoma bodies and interdigitating cytoplasmic processes, as well as negative staining for EMA made us exclude the possibility of meningioma. As electron microscopic examination

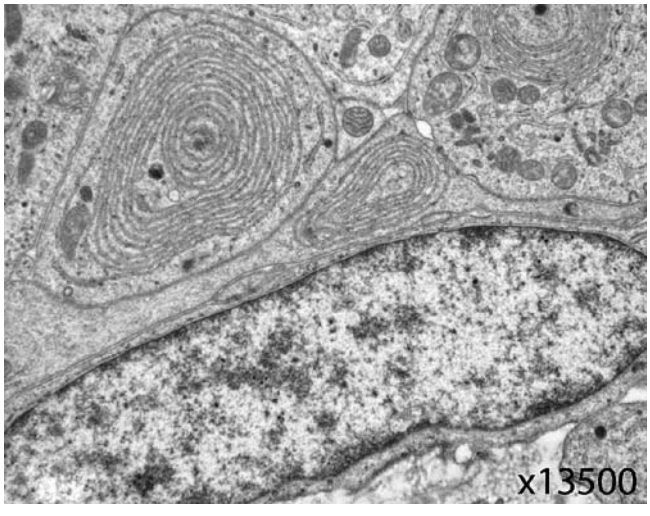

Fig. 2 Electron microscopic examination demonstrates ribosome-lamella complexes in spindle shaped tumour cells. ×13,500

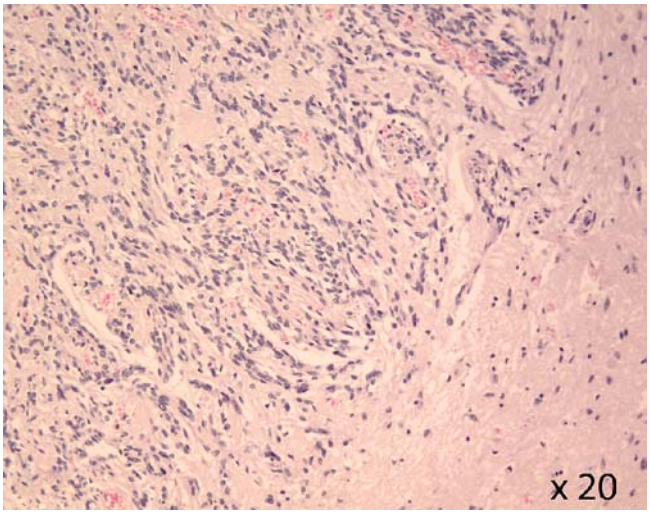

Fig. 4 Autopsy findings: Section from the left inferior colliculus demonstrates tumour invasion of brain tissue. Haematoxylin and eosin, ×20

failed to demonstrate basal laminas, haemangiopericytoma was considered unlikely.

Histological features reported to correlate with malignancy in pleural SFTs are increased cellularity, nuclear pleomorphism, increased mitotic activity and necrosis/haemorrhage [8]. The clinical behaviour of extrathoracic SFTs is unpredictable [4, 6, 8]. To our knowledge, only 5 of the 12 reported malignant SFTs of the meninges demonstrated histological features of malignancy in the original tumour sample [4, 6].

RLCs usually occur in haematological malignancies, especially in hairy cell leukaemia. In the intracranial compartment they have been described in one astrocytoma, two gangliogliomas and two haemangioblastomas [1]. Tani and Higashi [7] reported RLC in a meningioma but this tumour lacked the distinctive cylindrical architecture of an RLC [1]. Their functional significance is undetermined, but they probably signify aberrant protein metabolism [1]. Some authors regard RLC as a marker of malignancy [1]. However, since RLCs have been reported in both benign tumours and non-neoplastic cells, the prognostic impact of this structure remains to be established.

This case supports previous reports that the clinical behaviour of meningeal SFTs is unpredictable and that even tumours without histological features of malignancy may be biologically aggressive. As recurrences may occur many years after removal, careful and prolonged follow-up is mandatory.

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