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CASE REPORT

Recurrent Multiple Atypical Meningiomas Despite Neurosurgical Resection

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Summary
Meningiomas constitute 20-38% of primary intracranial neoplasms. Among these, atypical meningiomas (WHO grade II) comprise 4.7-7.2% of symptomatic cases. The primary goal of surgery is the complete removal of the meningioma, including the attachment to dura mater and bone. Subtotal resection as well as more aggressive histological type and grade are associated with higher recurrence risk. Here we report a 68-year-old male who suffered from multiple (5) meningiomas which necessitated 3 resections within 4 years. The simultaneous occurrence of different tumour grades of malignancy as well as grade progression was evident.
Key words: meningioma, recurrence, multiple meningiomas, surgical treatment

AIM OF THE DEMONSTRATION

The aim of the demonstration is to present a clinical case of recurrent multiple meningiomas showing the phenomena of simultaneous occurrence of different tumour grades and the histological grade progression over the time course.

CASE REPORT

A 68-year-old male was admitted to a university hospital for elective resection of right-sided meningioma. The patient complained of progressing weakness in the left hand, episodes of syncope and epileptic seizures, nausea and vomiting. Computed tomography of the brain showed right sphenoidal meningioma with wide perifocal swelling. The oncologic history was remarkable for previous bilateral meningioma resections 1 and 3 years earlier. During the first operation, 2 meningiomas were resected. From the right parietal area, multiple reddish-grey tissue fragments, measuring up to 3 cm, were removed. Histological examination showed transitional meningioma with stromal hyalinosis, grade I by World Health Organisation (WHO) classification (Fig.1A-B). From left parietal region, a semilunar, grey, rough mass, measuring 2 cm, was extirpated and diagnosed as atypical meningioma, WHO grade II, by pathology examination. One year later, magnetic resonance imaging (MRI) showed no evidence of recurrence. However, in the next year the symptoms recurred and MRI revealed right frontal and parietal tumours. The largest lesion was associated with perifocal swelling. Right frontoparietal trepanation was performed and 2 meningiomas were resected in 225 minutes. Histological examination showed medium-sized polygonal cells, focally interspersed with many small vascular channels. There was no necrosis (Fig.1C). By immunohistochemistry, the tumours expressed epithelial membrane antigen and vimentin. The proliferation fraction by Ki-67 was up to 35%. The final diagnosis was atypical meningiomas, WHO grade II, and radiotherapy was recommended.

At present, the general condition was estimated as average. The Glasgow coma scale score was 15. Paresis of the left hand and delayed cognitive response were found. The arterial pressure was 140/90 mm Hg, pulse rate was 96 bpm and breathing sounds were normal having frequency of 16 times per minute. Right pterional craniotomy was performed and convexial meningioma was extirpated within 120 minutes. During the operation the brain was visibly oedematous (Fig. 1D); dehydration and corticosteroid treatment was applied. The postoperative period was smooth. The histological examination of surgical material showed high-cellularity tumour with loss of tissue architecture (Fig. 1E), consisting of medium-sized and small neoplastic cells with marked nucleolomegaly (Fig. 1F). The final morphological diagnosis was atypical meningioma, WHO grade II. Thus, the patient has had multiple (5) meningiomas, experiencing 2 recurrences and grade progression at the right side.

DISCUSSION

Meningiomas constitute 20-38% of primary intracranial neoplasms. As autopsy studies have revealed undiagnosed asymptomatic meningiomas in 2.3% of individuals, the true prevalence is likely higher. These tumours arise from arachnoid meningothelial cells and occur mostly over cerebral convexities and at the skull base in the parasellar region (3). They are benign in more than 90% of the cases. However, atypical meningiomas, WHO grade II comprise 4.7-7.2% of symptomatic cases and exhibit increased tissue and cell abnormalities, such as higher mitotic activity or at least 3 of the following histologic features: increased cellularity, small cells with a high nuclear: cytoplasmic ratio, prominent nucleoli, patternless growth, and spontaneous necrosis (3,7). The most common histological types include meningotheliomatous (32%), followed by transitional (13.5%), fibrous (14.5%), and psammomatous (14.5%) meningiomas (9). Meningiomas are more common in older age and in females (3).

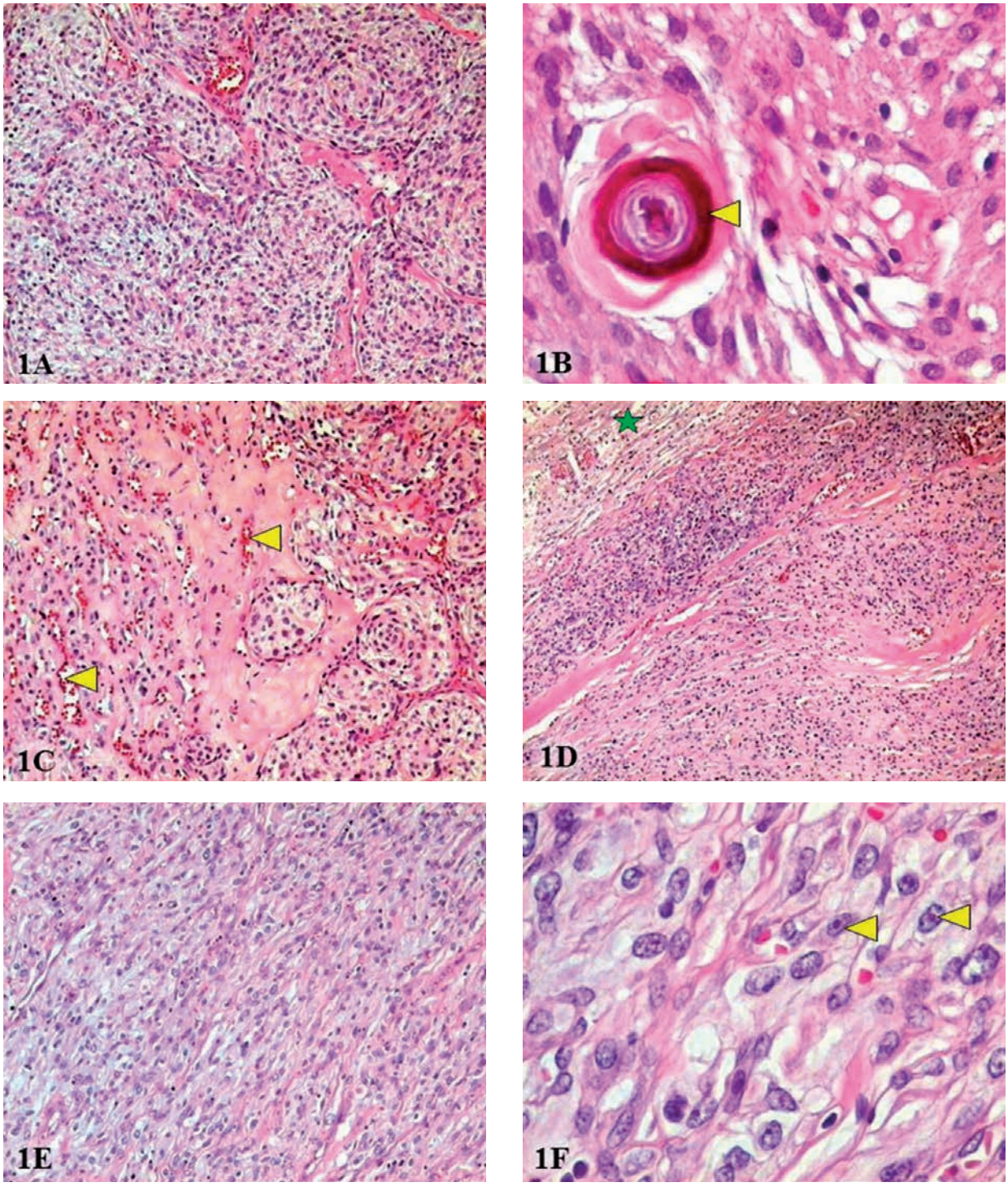

Fig. 1. The tissue structure of multiple recurrent meningiomas. 1A, Transitional meningioma, grade I. 1B, Psammoma body (arrow) in meningioma, grade I. 1C, Grade progression in meningioma showing focal loss of architecture and high vascularity (arrows). 1D, Atypical meningioma, grade II, exhibiting invasive growth, areas of high cellularity and loss of architecture. Note the highly oedematous brain tissue (star). 1E, Atypical meningioma, grade II, showing loss of architecture and high cellularity. 1F, Nucleolomegaly (arrows) in an atypical meningioma. Haematoxylin-eosin, original magnification 50x (1D), 100x (1A, 1C, 1E), 400x (1B, 1F).

Only 1-9% of meningioma patients have multiple synchronous tumours. Approximately one third of multiple meningioma cases show simultaneous occurrence of different WHO grades that can be associated with partially different somatic genetic changes (8). Multiple meningiomas can be associated with neurofibromatosis type 2 (NF2) due to the inactivation of the *NF2* gene on chromosome 22, encoding the tumour suppressor protein merlin (8). However, only 1% of meningioma patients have multiple tumours in association with NF2 while 4% have multiple tumours but lack NF2 (2). The clinical features of NF2 comprise bilateral vestibular schwannomas or family history of NF2 in association with either vestibular schwannoma, or two from the following tumours: meningioma, glioma, neurofibroma or schwannoma (6). Thus, the clinical data do not confirm NF2 in our patient.

Neurosurgical resection is standard therapeutic option and is aimed at complete removal of meningioma, including the dural attachment and invaded bone, if applicable. Complete resection is difficult or impossible in approximately 20-30% of patients because of multiple tumours in different regions, severe adherence to or invasion in the brainstem or cranial nerves, or entrapment of the vertebrobasilar blood vessels. Either subtotal resection, or gross total resection without radiotherapy for atypical meningioma correlates with higher recurrence rate (1,4,9). The recurrence rates of atypical meningioma are following: 7% within 1 year, 41% in 5 years, and 48% in 10 years after surgical resection. The 5- and 10-year overall survival rates are 78.4% and 53.3% (1,5).

In conclusion, we present a rare case of multiple recurrent meningiomas with simultaneous occurrence of different histological grades and grade progression.

Conflict of interest: None

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