Chordoid meningiomas of a different histopathological pattern. A report of two cases

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Abstract
Chordoid meningioma is an uncommon histopathological variant of meningioma with a peculiar chordoma-like appearance. Its association with systemic inflammatory disorder linked to Castleman’s syndrome was confirmed in the majority of young patients, however such a relationship in adults remains enigmatic.

We report two cases of chordoid meningiomas in adult patients without manifestation of Castleman’s syndrome. One tumour was almost totally composed of chordoma-like areas whereas the second one exhibited the unique combination of chordoma- and chondroma-like pattern. This is the first description of chordoid meningioma combined with extensive cartilaginous metaplasia. Both tumours exhibited histological evidence of infiltrative growth, accompanied by a relatively high proliferative index within structures of chordoid appearance. The designation of the chordoid component in meningioma is very important as this subtype of meningioma exhibits a more aggressive biological behaviour and higher risk of recurrence.

Key words: chordoid meningioma, cartilaginous metaplasia.

Introduction
Chordoid meningioma is a rare histopathological variant of meningioma, that comprises less than 0.5% of all meningeal tumours [6]. The meningiomas with distinct chordoma-like appearance were originally described by Kepes et al. [11] in young patients with manifestation of systemic disorders linked to Castleman’s syndrome. To date, two large series [6,11] and several individual cases of chordoid meningiomas have been reported [4,5,8,13,16,17,31,34,36-38]. However, the association of chordoid subtype of meningioma with hematological disorders in adults remains enigmatic.

We report two cases of chordoid meningioma in adult patients without manifestation of Castleman’s syndrome. In one tumour the peculiar coexistence of chordoma-like pattern and cartilaginous metaplasia occurred. This is the first description of the distinct chondro-chordoid pattern in meningioma.

Material and methods
The biopsy tumour tissue was fixed in 10% formalin, embedded in paraffin and stained with hematoxylin-eosin (H&E), Gomori’s method and mucicarmine. Immunohistochemical analyses were performed on...
The histopathological pattern of chordoid meningiomas was assessed using paraffin-embedded specimens according to the avidin-biotin complex method (ABC) with DAB as chromogen, using antibodies against epithelial membrane antigen (EMA), cytokeratins cocktail AE1/AE3, glial fibrillary acidic protein (GFAP), desmin and vimentin (all antibodies from Dako). The MIB-1 labeling index was established.

**Case 1**

A 47-year-old woman (KM) experienced recurrent sensory dysphasia for a few months and she was admitted to the Neurosurgical Department with a tumour in the left temporo-parietal region seen in MR imaging. On admission to the Neurosurgical Department she had no signs on the neurological exam. No physical abnormalities related to Castleman’s syndrome were detected. The MRI of the brain revealed a tumour of irregular outlines with homogeneous contrast enhancement accompanied by “dural tail sign” with marginal dural thickening that tapered peripherally (Fig. 1). The tumour was accompanied by marked oedema. EEG recording was borderline normal with a few slave waves after photostimulation. Macroscopically, the tumour of meningioma appearance was totally removed with the adherent dura mater, according to Simpson’s classification – grade I. Control CT scan proved total removal. She was discharged without neurological deficit and next she was submitted to conformal radiotherapy in a total dose a 54 Gy with no side effects.

**Microscopically**, the tumour displayed multilobular architecture that was almost totally composed of irregular cords and fascicles of spindle and/or epithelioid cells embedded in the loose matrix (Fig. 2A). The majority of tumour cells exhibited oval or round nuclei with finely dispersed chromatin surrounded by eosinophilic cytoplasm. The nest and cords of tumour cells were floating in abundant mucinous stroma in a chordoma-like pattern (Fig. 2B, 2C). The typical physaliphorous cells were not encountered, however some cells showed vacuolization of their cytoplasm. The neoplastic cells demonstrating classic cytologic features of meningothelial cells with round to oval nuclei and nuclear-cytoplasmic invagination were seen focally. The more typical meningothelial areas revealed marked nuclear atypia (Fig. 2D). The tumour contained little inflammatory lymphocytic and plasma cells infiltrates. There was evident invasion of the tumour into adjacent brain structure and dura (Fig. 2E). Mitotic figures could be found only occasionally. The MIB-1 labeling index (LI) was conspicuous (Fig. 2F, Fig. 7). The tumour cells exhibited strong immunoreactivity for EMA (Fig. 3A) and vimentin (Fig. 3B) but were negative for cytokeratins cocktail AE1/AE3, desmin and GFAP.

**Case 2**

A 47-year-old man (TD) was admitted to the District Hospital after loss of consciousness with 2 months history of grand mal seizure. CT scan revealed tumour mass in the right frontal lobe. On admission to Neurosurgical Department he had no signs on neurological exam. There were no laboratory findings suggesting haematologic disease. The magnetic resonance imaging (MRI) of the brain revealed a tumour mass in the right frontal convexity (Fig. 4). The tumour showed irregular outlines and heterogeneous signal intensity accompanied by local brain oedema. Right craniotomy was performed. During operation, the tumour revealed meningioma appearance with infiltration of the dura and adjacent bone. There was no arachnoid plane between tumour and surrounding...
Fig. 2. Case 1. Histopathological features of pure chordoid meningioma. **A.** Irregular cords and fascicles of spindle and epithelial tumour cells in the loose, myxoid matrix. H&E, x 200; **B.** Chordoid-like pattern with nest and cords of cells floating in the eosinophic matrix. H&E, x 400; **C.** Neoplastic cells embedded in abundant mucinous stroma. Mucicarmin, x 400; **D.** Typical meningothelial areas with marked nuclear atypia. H&E, x 400; **E.** Invasion of the tumour into the adjacent dura. H&E, x 100; **F.** The high MIB-1 labeling index. x 200
brain surface, also indicating the infiltrating tumour growth. The tumour was removed totally with resection of the infiltrated dura and bone, according to Simpson's classification for removal of meningiomas - grade I. Bone depletion was completed by artificial material – Codubix®. Control CT scans proved total removal. He was discharged without neurological deficit and next was treated with conformal radiotherapy in a total dose 54 Gy with no side effects. He returned to fulltime work.

Microscopically, the tumour displayed mixed pattern with both, chordoma- and chondroma-like appearance. The chordoma-like areas exhibited typical clusters and strands of spindle or epithelioid cells in myxoid matrix (Fig. 5A, 5B). The large parts of tumour presented cartilaginous metaplastic changes (Fig. 5C). The cartilaginous islands were composed of small nest of ovoid, polygonal, epithelioid cells embedded in abundant, eosinophilic homogenous stroma of chondroma-like appearance (Fig. 5D). The areas with meningothelial pattern containing whorls of more typical transitional meningioma were also seen (Fig. 5E). The tumour displayed tendency to infiltrate both, the adjacent brain and dura mater. Invasion of the underlying cerebral cortex is accompanied by extensive reactive gliosis with strong GFAP immunoreactivity (Fig. 5F). Immunohistochemically, the neoplastic cells in both histological patterns showed strong, diffuse reactivity for EMA (Fig. 6A, 6B) and vimentin but were negative for cytokeratins AE1/AE3, desmin and GFAP. The MIB-1 LI varied in chordoma- and chondroma-like areas (Fig. 7), being lower in cartilaginous tumour regions.

Discussion

Meningiomas are common primary intracranial tumours derived from meningothelial cells. They
Fig. 5. Case 2. Histopathological appearance of chordoid meningioma with advanced cartilaginous metaplasia; A, Parts of the tumour with a typical chordoid-like pattern. H&E x 100; B, The cords and nests of neoplastic cells in myxoid stroma. H&E x 200; C, Part of the tumour with prominent cartilaginous metaplastic changes. H&E x 100; D, Small groups and nest of ovoid, polygonal, epithelioid cells embedded in homogenous, eosinophytic background of chondroma-like appearance. H&E x 200; E, Cellular whorls of more typical transitional meningioma. H&E x 100; F, Tumour invasion of the underlying cerebral cortex accompanied by extensive reactive gliosis with strong GFAP immunoreactivity x 100
exhibited a wide spectrum of microscopic appearance and capacity for mimicking the histological features of other neoplasms [10]. The current WHO histopathological classification determines 15 separate histopathological variants of meningiomas that correspond with 3 grades of malignancy [12]. This histologically and biologically heterogenous group of neoplasms was still covered by some secrets [23] and is often related with clinical problems [12,26,27,35].

Chordoid meningioma is an uncommon subtype of this large group of meningeal tumours with a peculiar histological pattern. It has been suggested that the unique feature of chordoid differentiation is related with unbalanced translocation t(1;3)(p12-13;q11) in meningiomas [29].

The term “chordoid” was originally introduced by Kepes et al. [11] to describe the chordoma-like appearance of meningioma in 7 cases of young patients. These and other reported cases of chordoid meningiomas in childhood and adolescence were commonly associated with systemic symptoms i.e. microcytic anaemia, hypergammaglobulinemia and angiofollicular lymphoid hyperplasia, particularly linked to Castleman’s syndrome [2,11].

Histologically, the meningioma with the typical chordoma-like pattern in young patients was usually associated with prominent lymphoplasmacellular infiltrates, mostly composed of B-cells [37]. The manifestation of Castleman’s syndrome is suggested to be related to overproduction of pyrogenic cytokine interleukin 6 [2,7]. However, the large clinicopathological study of Couce et al. [6], including 42 cases of chordoid meningiomas, demonstrated that lymphocytic infiltrates varied, being absent in about 40.5% of tumours. Also, many other reports of adult cases did not reveal such association with systemic manifestation [14,17,21,28,36-38]. One case of chordoid meningioma of lung has been also described [25].

The histopathological diagnosis of chordoid subtype of meningioma is based on the characteristic chordoma-like appearance. The other tumours of this peculiar architectural pattern should be taken into consideration in differential diagnosis, particularly chordoma, myxoid chondrosarcoma, chordoid glioma and metastatic mucinous carcinoma [37]. The main histopathological feature of chordoid meningioma is
very similar to that of classical chordoma. The correct diagnosis might be confirmed by immunohistochemical studies. Chordoma is consistently immunopositive for cytokeratins and EMA and is usually located in the midline [20]. Chordoid glioma is a rare GFAP-immunopositive tumour limited to the third ventricle [19,24,30,33]. In contrary, chordoid meningiomas were positive for EMA and vimentin but negative for cytokeratin and GFAP. Moreover, in the majority of chordoid meningiomas it is possible to identify more or less distinctive areas of a conventional type of meningioma that is helpful in final histopathological diagnosis.

Our two cases of meningioma with chordoid components in adults did not reveal laboratory findings of hematologic abnormalities and histopathological evidence of chronic lymphocytic infiltrates. In both tumours, the chordoid pattern was associated with histological evidence of local invasion of surrounding structures suggesting aggressive clinical behaviour.

Moreover, the second case exhibited two different structural components such as chordoma-like and chondroma-like structures, probably reflecting the concomitant cartilaginous metaplastic changes. Meningiomas with a pattern corresponding to at least two histological variants have been sporadically reported [15]. The current WHO classification [12] considers meningeval tumours with pure cartilaginous mesenchymal differentiation as a metaplastic subtype. Various types of metaplasia including xanthomatous, osseous, lipomatous or cartilaginous differentiation might occur in meningiomas. However, the extensive cartilaginous metaplastic changes are extremely rare [3]. The peculiar association of chordoma- and chondroma-like structures in one of our cases resulted in some difficulty in its subclassification into the specific chordoid or metaplastic variant of meningioma. Its histological similarity to a very uncommon but controversial variant of chordoid meningioma might suggest that chordoid metaplastic changes are based on primary chordoid architecture of the tumour. The tumour was finally diagnosed as chordoid meningioma with advanced metaplastic changes. The cartilaginous parts of the tumour exhibited lower MIB1-labelling than chordoma-like regions but the tumour exhibited massive infiltration of adjacent structures. The MIB-1 LB in the meningiomas appears to be an important prognostic factor associated with potentially aggressive behaviour [1,9,18,22,32].

The correct diagnosis of the chordoid component in meningioma is very important as this subtype of meningioma exhibits more aggressive biological behaviour and higher risks of recurrence than conventional types of WHO grade I. It happens especially, when the tumour is subtotally removed [6]. The radiotherapy is often used for meningiomas that are incompletely excised, recurrent and histologically atypical and/or anaplastic [35]. In the current WHO classification, the chordoid subtype of meningioma corresponds to grade II and requires long-term follow up after surgery [12].

References