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Chordoid Meningioma with Hyperostosis of The Skull Bone: A Case Report

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Abstract

We are reporting a 39-year-old woman diagnosed with chordoid meningioma with hyperostosis of the skull bone. A red-greyish, encapsulated brain tumor was located in the parietal lobe of the right hemisphere of the cerebrum. A large high density right pariteal mass was disclosed by CT scan and destruction the skull bone. The tumor revealed a multilobular arrangement of two types of neoplastic cells, and the surrounding myxoid stroma was separated by incomplete fibrous septa. Neoplastic cells consisted of myxomatous and meningothelial cells. The former made up about one-third of the tumor, had a vacuolar cytoplasm, and were arranged in a chordoma-like cord pattern. They were floating in myxoid stroma. The latter had an eosinophilic spindle or epithelioid cytoplasm and were disposed in lobules. Both neoplastic cells were positive for vimentin and EMA, and were consistently negative for cytokeratin, GFAP and Ki-67 showed <20%(low proliferation)

Key words

Chordoid meningioma, hyperostosis, skull bone

Introduction

Meningiomas are tumors of the meninges with an incidence ranging from 13-19% of all primary brain tumors. Meningioma microscopically has a very varied picture and gives an appearance of massive classification. Cushing and Eisenhardt's types of meningiomas present nine main types and twenty subtypes. Meningiomas can undergo metaplastic and degenerative changes. Chordoid meningioma is a rare variant of meningioma and has a histological appearance that resembles chordoma and other chordoid neoplasms. Tumors have a well-documented tendency for local recurrence and aggressive behavior. Many cases of this tumor occur around the cerebral convex, and in a location similar to the classic form of meningioma. We report a case of cordoid meningioma with hyperostosis of the right parietal bone.

Case report

A 39-yr-old woman visited Siloam Hospital Jambi and her chief complaint was severe head ache about 6 months and became in the last 14 days. Neurological examination showed no focal deficit. In a routine laboratory test, the hemoglobin, mean corpuscular volume and mean corpus-cular hemoglobin were 15.2 g/dL, 85.60 fL and 30.0 pg, respectively. Therewas no laboratory finding suggesting hematologic abnormality or dysgammaglobulinemia. The CT-scan imaging revealed right parieto-occipital mass, size 77 x 58 mm, enhanced with contrast injection with severe midline shift to the left (Fig.1). Thus the preoperative diagnosis was SOL supratentorial at right parietal et causa suspect meningioma.

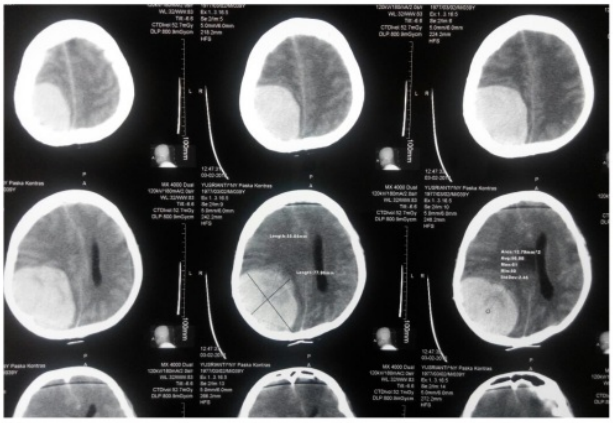

Figure 1. Ct-Scan

Right parietal craniotomy was performed, and reg greyish rubbery mass was seen with high vascularity. There was no gross invasion to adjacent brain tissue, and the mass was removed totally with adhering dura. (Fig.2)

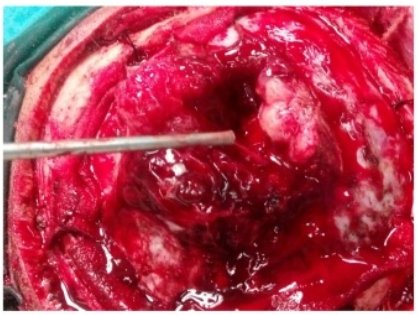

Figure 2. Chordoid meningioma mass

Pathological finding

Histologically, the tumor mainly consisted of trabeculae or cords of eosinophilic vaculoated cells in the abundant mucoid matrix. This chordoid area comprised the majority (about 95%) of the tumor, which intermixed with small areas of conventional meningioma. There were prominent inflammatory cell infiltrates within the mass as well as around the tumor margin. Neither nuclear pleomorphism including mitosis nor necrosis was found. (Fig 2a.). Tumor cell found in duramater and there was invading to the bone sample. Immunohistochemically, Ki-67 showed <20% (fig 2b). Tumor cells showed diffuse cytoplasmic staining for vimentin (fig 2c) and epithelial membrane antigen (Fig. 2d). None of tumor cells expressed glial fibrillary acidic protein (fig 2e) and cytokeratin (fig 2f). All these findings were consistent with those of chordoid meningioma.

DISCUSSION

The designation of meningioma has been extended through the years to diverse neoplasms sharing only a tendency to arise within the histogenetically complex tissues of the leptomeinges or duramater .(1) The chordoid meningioma was first reported by Kepes et al in young patients who had iron-refractory hypochromic microcytic anemia and/or dysgammaglobulinemia. In his report, he explained that chordoid meningeal tumor displaying histological characteristics closely imitating those of chordoma. (2) This type of meningioma is a rare meningioma according to World Health Organization grade II meningioma because of its more aggressive behavior and increased likelihood of recurrence.

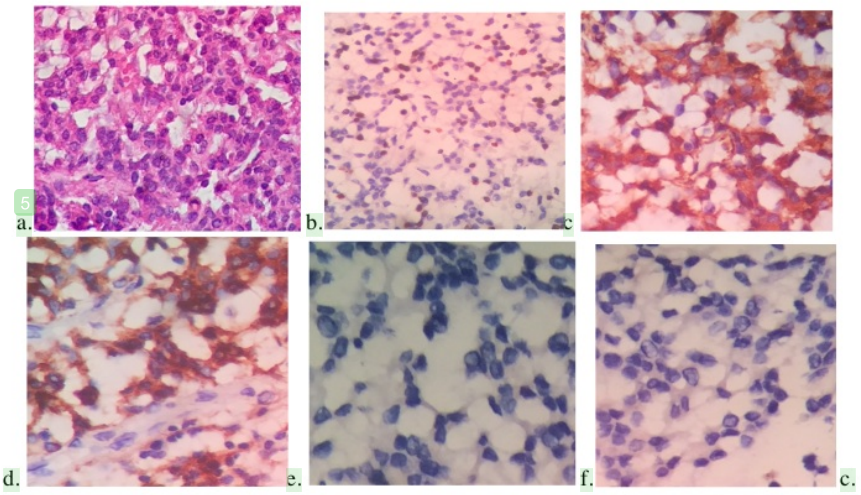

Fig 2. Chondroid meningioma a. Hematoxilin-Eosin stain b. Ki-67 showed < 20% immunoreactive c. Immunoexpression of epithelial membrane antigen (immunohistochemical stain, ×400). D. Immunoexpression of Vimentin (immunohistochemical stain, ×400) e. Immunoexpression of cytokeratin (immunohistochemical stain, ×400) f.Immunoexpression of GFAP (immunohistochemical stain, ×400).

In a study by Couce et al in 2000 reported that there were a total of 42 chordoid meningiomas and 0.5% of all meningiomas operated on at the Mayo Clinic from 1975 to 1997. From the conclusion, in this case, there was no significant association with abnormalities that were systemic or hematologic findings. Likewise, Kepes et al stated that patients of a younger age range were more susceptible to the systemic effects of lymphoplasmicellular infiltrates.

Microsopically, the chordoid meningioma gives a lobular-lobular image of septa and a tendency to form an eosinophilic mass, sometimes the cells have vacuoles and resemble chordomas. Partial chordoid areas are found throughout the tumor or occur partly or interspersed with classic meningioma areas.

The differential diagnosis of chordoid meningiomas includes chordoma, extraskeletal myxoid chondro sarcoma, chordoid glioma, and metastatic mucinous carcinoma. Immunohistochemical features along with the location of the tumor are helpful in the differential diagnosis between chordoid meningiomas and chordomas. In chordoid meningiomas, tumor cells are strongly positive for EMA and vimentin, but negative for S-100 protein and cytokeratin. In contrast, the latter two antibodies are characteristically expressed in chordomas. Additionally, chordomas usually locate in the midline structures such as the clivus or sellar area. Extraskeletal myxoid chondrosarcomas are rare and have not been described in the central nervous system. In contrast to chordoid meningioma, they are positive for S-100 protein (19). Chordoid glioma is a recently described entity consisting of cords and clusters of epithelioid cells with abundant eosinophilic cytoplasm in a mucoid matrix, simulating chordoid meningioma. In contrast to chordoid meningioma, the tumor cells in chordoid glioma are strongly glial fibrillary acidic protein-positive (20). Metastatic mucinous carcinoma differs from chordoid meningioma by its strong and uniform Cam 5.2 positivity and by the presence of intracellular mucin in tumor cells (6). In our case, the presence of focal areas of meningothelial pattern, positive immunostaining for vimentin and EMA, and negative immunostaining for S-100 protein, cytokeratin and glial fibrillary acidic protein facilitated the differential diagnosis.

The major clinical parameter for predicting recurrence in chordoid meningiomas is the extent of resection (6), like other meningioma variants (21). Chordoid meningiomas exhibit a high rate of recurrence following subtotal resection. In the series of Couce et al. (6), one or more recurrences were noted in 14 (42%) of the 33 cases with available follow up, and of those, 13 (92%) had been subtotally resected. The tumor grade is the most important histological parameter to determine the risk of recurrence in meningiomas (22). Although most meningiomas are benign and are graded into WHO grade I, the chordoid variant is associated with a less favourable clinical outcome and is graded into WHO grade II, but inti this case was graded into WHO grade I because Ki-67 showed < 20% of tumor mass. The extent of chordoid pattern is also important in predicting prognosis. Couce et al. (6) observed that in the majority (85.7%) of recurred cases, the primary tumors show chordoid pattern in more than 50% of the tumor tissue. Proliferation indices have also been used to predict recurrence in meningiomas. Ki-67 proliferation index show a highly significant increase from benign, to atypical, and anaplastic meningioma (23).

The conclusion of this study is that chordoid meningioma is a morphological variant of meningioma that often occurs. Histopathological misdiagnosis is very possible, so it needs to be avoided, supported by positive immunostaining for EMA and vimentin. One important parameter is the determination of tumor grade to determine the potential risk of meningioma recurrence.

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