CHORDOID MENINGIOMA

Report of two cases

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ABSTRACT - We present CT scan, MRI and histopathologic findings of two patients harboring a rare type of meningioma. In the first case, a 52 year-old male patient, a large parasellar mass invading the cavernous sinus, infiltrating the infratemporal fossa and extending as low as C2 was founded. The tumor was isointense on T1, enhanced strongly with gadolinium injection, and was hyperintense on T2. In the second case, a 19-year-old male patient, a large high density temporal right mass was disclosed by CT scan. Both patients were taken to surgery. In the first case, only a partial removal was possible to be accomplished due to a severe intra-operative bleeding. In the second case, the tumor was totally removed. Both showed characteristic pathologic findings of a meningioma resembling a chordoma. Meningioma is a relatively common intracranial tumor, occurring most frequently in adults, showing a wide variety of growth patterns. We described a pattern that had a peculiar chordoma-like appearance. The pathological findings and the differential diagnosis from chordoma are discussed.

KEY WORDS: chordoid meningioma.

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Meningiomas range from 13 to 19 per cent of all primary brain tumors1. The microscopic appearance of meningiomas is highly variable and has led to several rather complex and confusing classifications. The classic monograph on meningiomas by Cushing and Eisenhardt presents nine major types and twenty subtypes. Meningiomas may undergo a variety of metaplastic and degenerative changes, which are not important if differences in biologic behavior are taken into account. However, this wide variety of forms gave rise to a bewildering array of histologic patterns.

The last classification of the World Health Organization listed 11 types of meningiomas. One of these variants is the chordoid meningioma, a very rare type, that resembles a chordoma with its large areas of lymphocytic and plasma cell infiltration1,2. This special group presents a remarkable myxoid and chordoid pattern, described in children and adolescents by Kepes et al.3. The resemblance of the tumor cells to those of a chordoma can be striking1,4. We report on two cases.
CASES

Case 1 - This 52 year-old male patient was referred to us complaining of severe pain on the right side of the face for the last two weeks, involving mainly the frontal and maxillary region. The patient underwent an operation six years ago for a right cerebellopontine angle tumor. Histological examination revealed it to be a neurinoma. On neurological examination he had right hypoesthesia on the first and second division of the trigeminal nerve and no hearing on that side. There was also some impairment of the lower cranial nerve function, with some dysphagia and nasal speech. MRI revealed a large parasellar mass invading the cavernous sinus, infiltrating the infratemporal fossa, extending as low as C2. The tumor was isointense on T1, enhanced strongly with gadolinium injection and was hyperintense on T2 (Fig 1). The patient was submitted to a cranio orbito zygomatic approach, with exposition of the superior orbital fissure and foramen rotundum. The middle menigial artery was ligated, permitting the visualization of the third division of the trigeminal nerve. Extensive drilling between V2 and V3 was done, but we decided to proceed the approach intradurally. The tumor was rather soft, very well vascularized and was removed piecemeal. Since there was a lot of bleeding, homeostasis was greatly required so we decide to perform a partial removal. Most of the tumor was extradurally removed. Fat and surgicel were left in the supratentorial part of the tumor. The tumor located in the infratemporal fossa was debulked without extirpation of the capsule. MRI was performed after the operation and showed a mass in the parasellar region around the fat tissue (Fig 2). The patient is doing well 3 years after the operation.

Case 2 - This 19 year-old male patient was referred to us complaining of severe right temporal headache from the last two months. Neurological examination was nor-
mal. The CT scan showed a large high density right temporal mass, enhanced by contrast administration (Fig 3). A fronto-pterional craniotomy was performed and the tumor totally removed. The microscopic appearance was described as secretory meningioma. The patient was discharged home free of symptoms one week later. On his follow-up two month later a CT scan was performed, showing no abnormalities (Fig 3). The patient was asymptomatic. Five months later he returned complaining of severe headache from the last five days. The patient presented inflammatory and febrile syndromes but the neurological examination was still preserved. At that time a CT scan demonstrated a huge renewed tumor. The patient died a few hours later before surgery.

A microscopic appearance slide review showed a mixoid matrix surrounded by a small strand of meningothelial cells areas, resembling chordoma histology. This was diagnosed as a chordoid meningioma.

**Histopathological findings** - Multiple fragments of tumor were fixed for classical histological study in 10% formalin; the sections were included in paraffin block, and stained with hematoxylin-eosin (HE). Immunohistochemistry was performed on paraffin sections using monoclonal antibodies to epithelial membrane antigen (EMA) and vimentin. All antisera were used with the avidin biotin complex technique and were visualized using 3,3 diamino benzidine tetrahydrochloride substrate. Appropriate positive and negative controls were also used. Light microscopy examination revealed tumor characterized by cohesive strands of epithelial cells in a myxoid Alcian-blue positive matrix and a chronic inflammatory response consisting of lymphocytes and plasma cells but without the formation of follicles or germinal centers (Fig 4). Immunohistochemical stains showed a membranous pattern of immunoreactivity for epithelial membrane antigen and diffuse immunoreactivity for vimentin.

![Fig 3. Case 2. CT Scan showing a large high hyperdense temporal mass after constrast injection. Normal CT Scan after two months.](image)

![Fig 4. Light microscopy examination revealed tumor characterized by cohesive strands of epithelial cells in a myxoid Alcian-blue positive matrix and a chronic inflammatory response consisting of lymphocytes and plasma cells but without the formation of follicles or germinal centers. H & E 100x, 400x.](image)
DISCUSSION

Burger, Scheithauer and Vogel called attention to this atypical form of meningioma in their Surgical Pathology of the Nervous System and its Coverings. Meningiomas are most common in the middle decades of life, usually after the third decade, and show a marked preference for women with an overall male: female of 1:2 (intracranial meningioma, 3:2; intraspinal meningiomas, 1:10). In order of incidence, intracranial meningiomas occur in the following locations: parasagittal and free convexity accounting for almost 50% of the cases, followed by sphenoid ridge, tuberculum sellae, olfactory groove, foramen magnum, optic nerve, tentorium cerebelli, and choroid plexus. The commonly recognized histological subtypes display both mesenchymal and epithelial features. Mesenchymal metaplastic changes include xanthomatous, lipoblastic, myxoid, chordoid and osteoblastic differentiation, while evidence of epithelial differentiation includes the presence of papillary structures, “epithelial” markers (EMA, EGF), and desmossomal attachments.

The chordoid meningioma is a rare variant with a myxoid pattern and cords of epithelioid cells similar to that a chordoma. A prominent lymphoplasmocytic infiltrate may be seen in some. Patients can present with iron-refractory anemia and polyclonal gammopathy (Castleman’s syndrome), both of which remit with resection but reappear upon recurrence. This variant may have prognostic significance since a number of patients with meningiomas has been a controversial issue. Only a histological study can confirm this situation. The association of pathological examination and immunohistochemical study can make a diagnosis of chordoid meningioma. Civit et al. say that the literature has shown that chordoid meningiomas display several areas of physaliferous cells, which give the tumor a chordoma-like aspect. The patient presenting with an intracranial meningioma usually has an indolent course of symptoms depending on its location. A critical evaluating of the two cases here presented and the recurrence after surgery call attention to a non benign form of meningioma of chordoid type.

REFERENCES