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CASE REPORT

SUPRASELLAR CHORDOID MENINGIOMA- HISTOLOGICAL MIMICKER OF CHORDOID TUMORS: A RARE CASE REPORT

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ABSTRACT

We present a case of a 40 year old male who presented to the neuro clinics with the complaints of headache for three months. The clinic-radiological differential diagnoses were Craniopharyngioma, Pituitary macroadenoma, meningioma, chordoma and Epidermoid cyst. On histopathological and detailed immunohistochemical examination, this case was diagnosed as Chordoid meningioma. Since the chordoid features are related with a rapid recurrence after incomplete removal, meticulous histopathological and immunohistological examination is crucial for the adequate postoperative

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INTRODUCTION

Meningiomas are dural-based neoplasms that account for ~37% of intracranialtumors in the adult population (Wang, 2021) They can occur anywhere within the central nervous system and have a predilection for females (Saraf, 2011) Meningiomas has been classified into 3 grades according to WHO, based on increased risk of recurrence and its associated mortality in grade III tumors has been observed. Although most tumors are categorized as low-grade (Nanda. 2017). Meningiomas originate from arachnoid cap cells, (Toland, 2021) and are the most common primary neoplasms of the central nervous system with an annual incidence of 8.83 per 100 000 (Reynoso-Noverón, 2021). Most meningiomas occur over the age of 65 yr, with frequency continuously increasing in subsequent years.(4) Of note, males generally tend to have higher-grade tumors as compared to females (Toland, 2021; Perry,1997). The majority of tumors are supratentorial. mostly localizing to parasagittal, falcine, and skull base (Cucu, 2019) Uncommon sites include intraventricular, orbit, intraosseous, and subcutis/dermis of the scalp. Although meningiomas are known as benign tumors, they are up to 15% atypical and up to 2% anaplastic (Arslantas, 2002).

Common symptoms are headache, focal cranial nerve deficiency, seizures, cognitive change and weakness and sometimes asymptomatic (Geçit, 2022). Meningiomas account for about 1% of sellar masses. Although they can mimic pituitary adenomas, but are more vascularised and invasive (Kwancharoen, 2014; Hershey, 1993) The term chordoid meningioma was first introduced by Keeps *et al*., in 1987 to describe a meninges tumour in young patients associated with microcytic anemia and/or dysgammaglobulinemia (Sangoi, 2009) Chordoid meningioma (CM) was classified separately for the first time in the 1993 World Health Organization (WHO) classification of tumors of the central nervous system.(14)Among grade II group, besides atypical and clear cell meningiomas, the rare type tumors constitutes chordoid meningiomas (CMs). CMs represent approximately less than 1% of intracranial meningiomas.(14,15) Chordoid meningioma (CM) is a meningioma containing regions that are histologically similar to chordoma, with trabeculae of eosinophilic, vacuolated cells in a myxoid background and corresponds to WHO grade II (Prokopienko, 2022; Ozen, 2004)

Case report: A 40 year male presented to out patient department of neurosurgery with complain of headache for 3 months. He also complained of vomiting associated with blurring of bilateral vision.

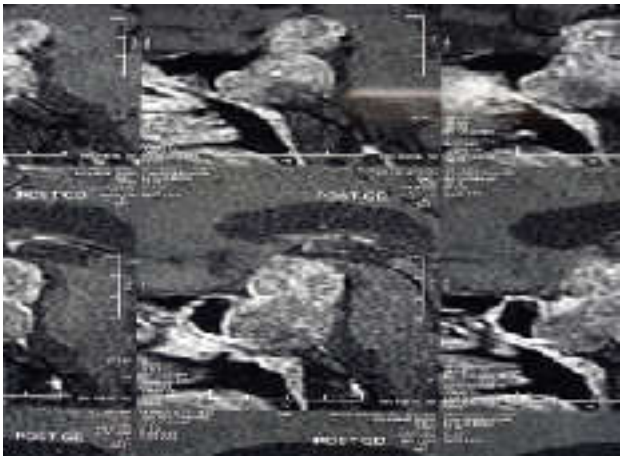

Figure 1. MRI Brain showing suprasellar and sellar mass

Figure 2. Gross image showing multiple creamish white tissue pieces measuring 4x3cms

There was no history of vomitingor no history related to hypergonadismor any sensory and motor weakness. Neurological examination revealed left temporal hemianopia and decreased visual acuity (3/6). Routine laboratory investigations including Hb, MCV, MCH, total protein, albumen, and globulin were all normal. The patient was advised MRI and serum prolactin level. The serum prolactin level was within normal limit of 33.6ng/ml. The MRI findings revealed a large well defined oval shaped T2/ Flair hyperintense mass lesion involving sellar and suprasellar region. Presence of heterogeneous post contrast enhancement noted within the lesion. The lesion is causing enlargement of sella with erosion of sellar floor and clinoid process.The differentials on CE-MRI included Pituitary Adenoma / meningioma /chordoma and epidermoid cyst (Figure1). Further the patient underwent surgery in the department of neurosurgery. We received multiple creamishwhite tissue pieces aggregate measuring4x3 centimetres. (Figure2). Histopathlogical examination revealed ill defined lobular architecture along with sheets, trabecular and chords of cells having moderate amount of eosinophilia to vacuolated cytoplasm, nuclear hyperchromasia and mild to moderate nuclear pleomorphism with interviewing stroma showing lymphocytes. 2-3mitosis/HPF were seen. No necrosis was appreciated (Fig 3,4). The histopathological differentials included chordoma, chordoid glioma, chordoid menigioma. Further immunohistochemistry was advised for the confirmativehistopathological diagnosis. Immunohistochemistry applied were GFAP, EMA, S100, Vimentin, brachyury and Ki-67. The neoplasm came out to be strong membranous positivity for EMA and moderate nuclear and cytoplasmic positivity for S100 IHC (figure 5,6), rendering a confirmative and final diagnosis of chordoid meningioma. Focal and weak cytoplasmic positivity was seen with vimentin.

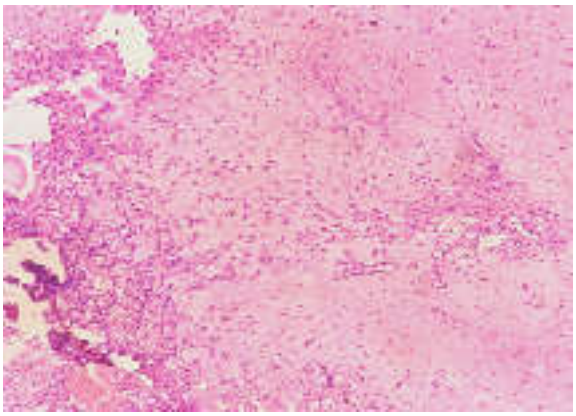

Figure 3. Sheets, trabecular and chords of cells arranges in lobulated architecture (H&E X100)

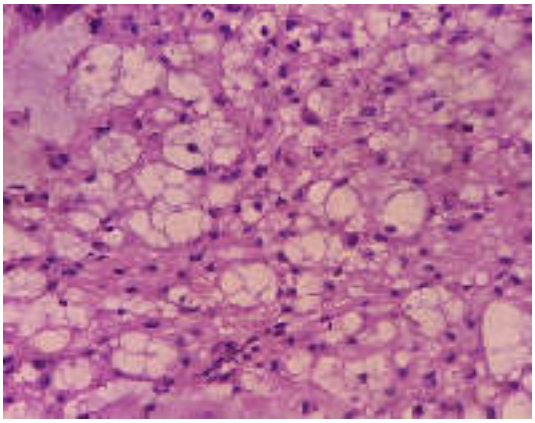

Figure 4. Cells with vacoulated to eosinophilic cytoplasm (H&E X400)

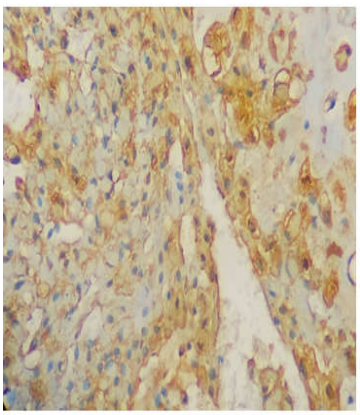

Figure 5. Diffuse membranous EMA positivity (IHC X400)

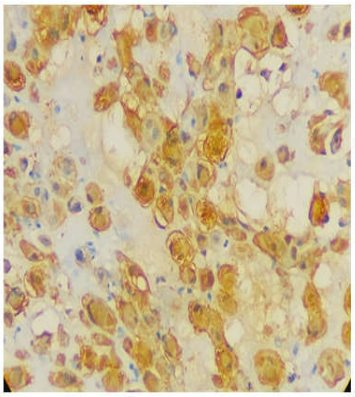

Figure 6. Diffuse cytoplasmic and nuclear S100 positivity (IHC X400)

GFAP and brachyury was negative thus excluding the possibility of chordoid glioma and chordoma respectively. Ki-67 was low, <2% indicating a low to grade neoplasm. Following surgery the patient had an uneventful postoperative recovery. The headache subsided and the visual acuityimproved to 5\6. Further the patient didn't developed similar complain as seen in the past and the post-op period of 6 month was uneventful.

DISCUSSION

Meningiomas are common intracranial tumors that originate from the arachnoidal cap cell of the meninges and have a large variety of histopathologic appearances (Mawrin, 2010) Most of the histopathological variants are only for descriptive purpose and carry no prognostic significance. However, some subtypes, such as chordoid and clear cell, have different clinical associations or prognostic implicationsand may show an aggressive clinical behaviour (Lin, 2010). It constitute 13–20% of all primary intracranialtumors (Epari, 2006). Microscopically, chordoid meningioma exhibits trabeculae of eosinophilic. vacuolated cells in a myxoid background mimicking chordoma (Lane, 2021; Yano, 2020). The present patient also showed histological features like those of chordoma on hematoxylin and eosin staining. However, intracranial chordoma is likely to grow in the skull base or the sella turcica in case of suprasellar chordoma and are more aggressive clinically (Stüer, 2006). The differential diagnosis with chordoma is of particular importance given its often midline skull base location, which can radiographically mimic pituitary adenoma (Lee, 1998; Santegoeds, 2018). The presence of a WHO grade II meningioma, including chordoid meningiomashould guide clinicians down a different management path, possibly resulting in closer follow-up and a lower threshold for re-operation in the event of recurrence.[14,27] Surgery is the treatment of choice for all grade of meningiomas, and gross total resection should always be the goal of surgery. However, some meningiomas, particularly those involving the cavernous sinus, petroclival region, posterior aspect of the superior sagittal sinus, or optic nerve sheath, cannot be completely removed due to their relationship to vital neural or vascular structures.[28,29] The recommendation for such circumstances is surgery plus postoperative radiotherapy, especially when dealing with WHO grade II or grade III meningiomas (Song, 2008; Fischer, 2022).

CONCLUSION

Chordoid meningiomas are rare subtype of meningiomas which has an increased propensity to recurr and carry an aggressive clinical behaviour. This subtype has been seen associated with Castleman's Syndrome and can be associated with T-cell rather than B-cell infiltration. This histologic category can be seen in the elderly as opposed to only in younger age groups.In conjunction with clinical and radiographic findings, immunohistochemical evaluation with a panel of podoplanin, D2-40, EMA, cytokeratins, GFAP, and CD10 is very helpful in distinguishing chordoid meningiomas from chordoma, chordoid glioma, skeletal myxoid chondrosarcoma, extraskeletal myxoid chondrosarcoma and low-grade chondrosarcoma. Differential diagnosis should be be excluded, as each one of them carry a different clinical course and treatment.

Conflict of interest: No conflict of interest.

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