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Hrushikesh Kharosekar,
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Paediatric intradural cerebellopontine angle chordoma mimicking meningioma

Hrushikesh Kharosekar, Debabrata Patra, Mazar Khan Mulla, Vernon L. Velho

Dept of Neurosurgery, Sir J.J. Group of Hospitals and Grant Medical College, Mumbai, INDIA

ABSTRACT

Chordomas are rare, notochord-derived neoplasms, most commonly affecting the sacrum and clivus, and exceedingly rare in the cerebellopontine angle (CPA), especially in children. This report describes a 10-year-old male presenting with hearing loss, giddiness, vomiting, and left-sided facial palsy, who was found to have a CPA chordoma that was SMARCB1-deficient, confirmed via histopathology. The clinical presentation and management are discussed, along with a review of the scant global literature.

Keywords
chordoma,
paediatric,
intradural

INTRODUCTION

Chordomas are rare malignant tumors arising from ectopic remnants of notochord. They account for 1–3% of primary bone tumors, mostly extradural with sometimes trans-dural extension. They have a predilection for the midline particularly the sacrum and spheno occipital synchondrosis of clivus (1). Primarily intradural chordomas are rarely reported in literature, less than 10 case for our knowledge. Most of these are seen in adult patients. Paediatric intradural chordomas only 4 cases have been reported till now. Cerebello pontine angle (CPA) chordomas are very rare, with only a handful of cases reported globally, and even fewer in paediatric patients. Intradural chordomas are usually benign lesions with no bony involvement. As there was no bony involvement in our case, we suspected tentorial meningioma as primary diagnosis. (1,2)

CASE PRESENTATION

A 10-year-old male child presented to us with gradually progressive hearing loss, giddiness for 3 months, intermittent episodes of vomiting. On neurological examination child had left-sided Grade 3 facial palsy and sensorineural hearing loss. Fundoscopy was suggestive of Grade 2 papilledema. On radiological examination Contrast-enhanced CT/MRI demonstrated a large heterogenous solid-cystic lesion in the left CPA with hydrocephalus. The patient underwent surgical resection of left CPA lesion with CSF diversion in form of ventriculo- peritoneal shunt.

Corresponding author:
Hrushikesh Kharosekar

Dept of Neurosurgery, Sir J.J. Group of Hospitals and Grant Medical College, Mumbai, India

hkharosekar@gmail.com

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Intraoperatively lesion was based on tentorium cerebelli, reddish grey in colour, highly vascular and suckable mimicking a meningioma. Complete excision of lesion as achieved with coagulation of dural attachment. All cranial nerves were preserved. Postoperatively, clinical recovery was achieved, and the patient was eventually discharged after suture removal. (Fig 1,2,3)

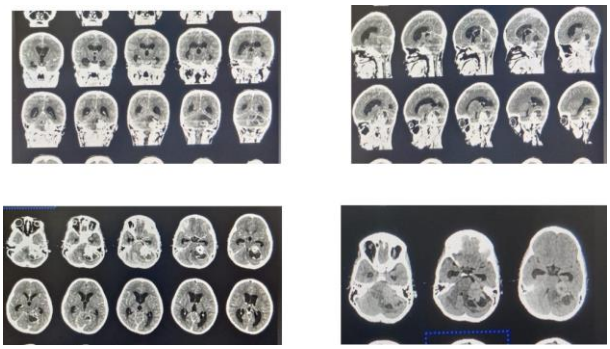

Figure 1. Preop CT scan of patient showing the left CP angle lesion

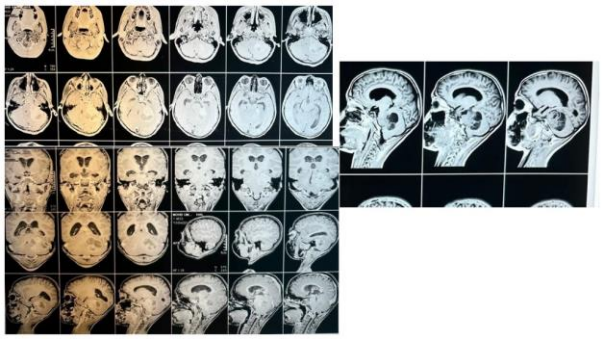

Figure 2. Pre op MRI with contrast

Histopathological evaluation of the resected tissue revealed poorly differentiated chordoma, with loss of SMARCB1/INI1 nuclear expression, confirming the diagnosis of a SMARCB1-deficient chordoma. The tumor cells were strongly positive for brachyury, CK pan, and EMA, consistent with chordoma. (4)

Discussion

Chordomas develop from remnants of notochord throughout the axial skeleton. Chordomas are rare slow growing primarily bony tumors with potential for trans-dural extension. They have malignant potential; symptoms are primarily due to compression of surrounding structures as they are slow growing. Rarely they can show acute progression of symptoms due to intertumoral haemorrhage.

Chordomas are usually extradural lesions with transdural extension in some cases. Purely intradural chordomas are rare, less than 4% of chordomas. Purely intradural chordoma in paediatric patients is even more rare as only 4 cases have been reported in literature till today. Purely intradural chordomas are usually well-defined lesions compared to extradural chordomas, they have no bony invasion, less adherent to surrounding structures. That is the reason these lesions can be resected completely causing less recurrence. (1,2)

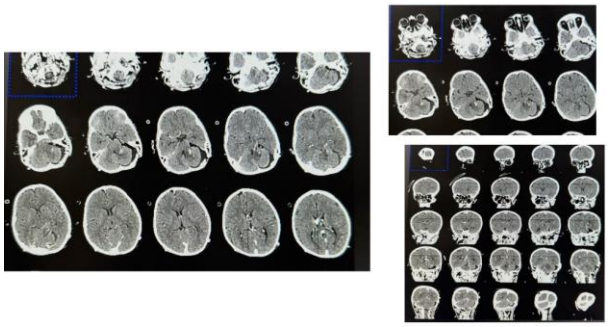

Figure 3. Post Op CT scan showing complete excision

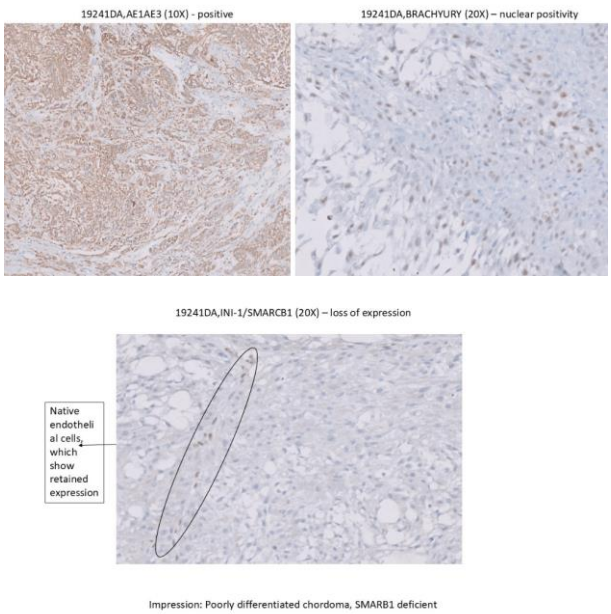

Figure 4. Histopathological image

Pathogenesis of intradural chordomas is not clear, various theories have been proposed. According of "migration" theory, some remnants of embryological

notochord are displaced intradural, as in cases of head trauma, from which intradural chordomas can originate. Other theory suggests malignant transformation of ecchordosis physaliphora (EP), a benign developmental ectopic remnant of intradural notochord tissue attached to the clivus. Even though EP and intradural chordomas are similar histologically, EP are smaller lesion(<2cm) which are usually asymptomatic. The differential diagnosis of these lesions includes dermoid, neurenteric and arachnoid cysts. (2,4)

CPA chordomas are notably rare, with only 2–3 cases described worldwide to date, and this case is among the first reported in Asia. The mainstay of treatment is surgical excision aiming for maximal safe resection. Role of adjuvant radiotherapy is debatable due to local recurrence rate shown even after subtotal resection. Ito et al reported zero recurrences in 18intradural chordomas among 17 patients with 5 to 144 months of follow-up while Vellutini et al reported a case of tumor recurrence two years after the initial diagnosis. So, role of adjuvant chemotherapy is still debatable. But considering the potential side effects of radiation on paediatric brain we propose careful observation rather than radiotherapy in these patients. (1,2,3)

Conclusion

Paediatric CPA intradural chordomas are rarely reported. Our case demonstrates the need to consider these lesions in the differential diagnosis of lesions of the CPA. As these lesions are slow growing and surgically completely resectable, role of adjuvant radiotherapy is debatable. Case pooling and further research are required to better understand and manage this entity.

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