



A Rare Case Report of an Intradural Left Cerebellopontine Angle Chordoma

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AJNS 2022;17:134–136.

Abstract

Keywords

- ▶ CP angle tumor
- ▶ petroclival chordoma
- ▶ brachyury
- ▶ chordoma
- ▶ skull base tumor

Intracranial intradural chordomas are rare entities constituting 1 to 3% of primary bone tumors. The mainstay of treatment remains aggressive resection of the lesion followed by adjuvant radiation therapy. We hereby report a case of a 70-year-old gentleman with intracranial, intradural chordoma arising from the left cerebellopontine angle. We hope to add to the existing minimal literature on this subject by highlighting this case, the first reported one from Asia.

Introduction

Chordomas are rare, locally aggressive, notochord derived lesions constituting 1 to 3% of primary bone tumors.^{1–3} Intracranial chordomas with transdural extension are well described; however, reports on the ones within the intradural space are scant.^{1,3} The molecular and cellular mechanisms causing bony invasion remain poorly understood and require further research.^{2,3} Computed tomography (CT) and magnetic resonance imaging (MRI) are important diagnostic tools.^{3,4} The mainstay of treatment for this condition is surgical resection followed by adjuvant radiation therapy.^{3,4} We hereby report a case of a 70-year-old gentleman who presented to us with vestibular complaints and disequilibrium and was later diagnosed to have intracranial, intradural chordoma arising from the left cerebellopontine angle (CPA) with involvement of the petrous and the clival bone for which he had to undergo excision of the lesion. However,

despite the best medical and surgical efforts, he succumbed to his illness. This is the first case to be reported from Asia and an addition to the already available literature.

Case Report

A 70-year-old gentleman presented to an emergency department with progressive worsening of left-sided hearing, occasional vertigo, and disequilibrium for approximately 3 months. On physical examination, he was alert, conscious, and oriented with the Mini-Mental State Examination score of over 25/50. He was unable to sit erect and swayed to the left side on sitting up. Cranial nerve examination showed decreased hearing on the left side. Past pointing was present on the left side with bilateral horizontal nystagmus. Other general and systemic examinations were within normal limits. CT brain showed a space-occupying lesion in the left CPA, compressing the brain stem. An MRI axial fluid-attenuated inversion recovery (FLAIR)

published online
June 6, 2022

DOI <https://doi.org/10.1055/s-0042-1749128>.
ISSN 2248-9614.

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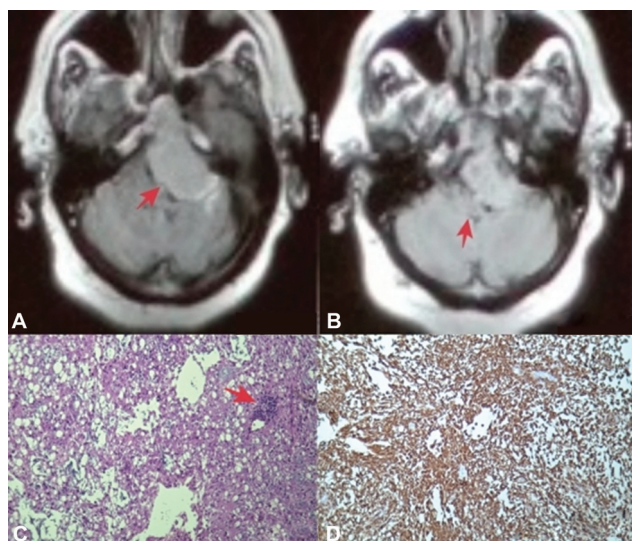


Fig. 1 (A) MRI axial FLAIR sequence showing a well-defined 4 cm × 2.5 cm hyperintense mass at the left cerebellopontine angle compressing the brain stem (red arrow). **(B)** The lesion was causing compression of the fourth ventricle (red arrow), and also showed evidence of brainstem edema with dilation of supratentorial ventricles causing obstructive hydrocephalus. **(C)** Photomicrograph showing a neoplasm composed of epithelioid cells with vacuolated eosinophilic cytoplasm (physaliferous cells) (red arrow) arranged in cords and sheets against a myxoid background (H&E stain; ×100); focal lymphocytic infiltration was also noted. **(D)** Tumor cells were diffusely positive for cytokeratin (immunoperoxidase; ×100). The histopathological analysis thereby confirmed the diagnosis of a conventional (classic) chordoma. FLAIR, fluid-attenuated inversion recovery; H&E, hematoxylin and eosin; MRI, magnetic resonance imaging.

sequence confirmed a well-defined 4 cm × 2.5 cm hyperintense mass at the left CPA (► **Fig. 1A** and **B**). He underwent a left retrosigmoid suboccipital craniotomy and excision of the lesion. Intraoperatively we noted a soft suckable vascular lesion compressing the lower cranial nerves (► **Fig. 2A** and **B**). Histopathological analysis confirmed the diagnosis of a chordoma (► **Fig. 1C** and **D**). Postoperatively, his neurological condition gradually deteriorated, and a noncontrast CT showed brain stem bleeding with intraventricular extension and features of uncus herniation. He was taken for immediate craniotomy, hematoma evacuation, and extraventricular drain placement. However, despite this rigorous medical and surgical management, his clinical condition gradually deteriorated, and he succumbed to his illness within 24 hours of the primary surgery.

Discussion

Chordomas are rare, aggressive, and locally invasive bone cancers that develop from notochordal remnants and preferentially affect the axial skeleton with the sacrum, skull base, and spine being the most common sites.³⁻⁵ Extradural chordomas are thought to develop from abnormal notochordal remnants in the ventral skull and are commonly seen at spondylocondroses in the ventral skull base, stretching from the dorsum sellae superiorly to the basion inferiorly.⁵ Development of an intradural chordoma has two possible theories.

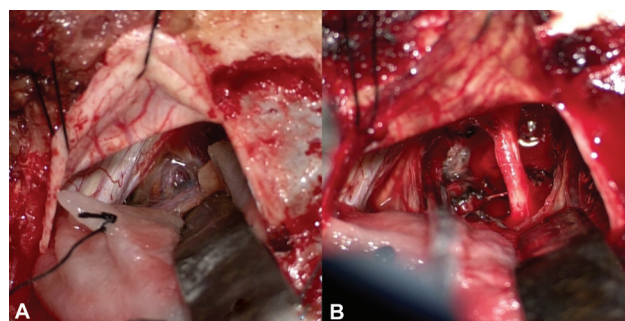


Fig. 2 (A) A soft suckable vascular lesion causing compression of the lower cranial nerves. **(B)** Postdebulking a substantial portion of the tumor and decompression of the brain stem; facial nerve and lower cranial nerves were preserved.

Some literature suggests that echordosis physaliphora (EP), a rare and benign intradural developmental condition derived from the notochord, can undergo degeneration and result in the development of an intradural chordoma. Whereas the other belief is that the displaced extradural residual notochord fragments can migrate into the intradural area especially in the event of trauma.^{4,5} However, the etiology remains poorly understood and requires further research.

Radiological imaging (CT and MRI) plays an important role in the evaluation and subsequent management of these patients. While schwannomas, meningiomas, and epidermoid cysts are common radiological differential diagnoses for a CPA tumor, intradural chordomas are rarely discussed in this list as they tend to be midline.^{3,5} Gross-total surgical excision with an increased emphasis on preservation of neurological function followed by adjuvant radiation therapy is the recommended treatment.^{4,5} Histopathological and immunochemistry analysis of the excised tissue is diagnostic where the genetic marker “brachyury” is proven to be unique, highly sensitive, specific, and plays an important role in confirming the diagnosis of this rare entity.⁶ However, in our case, it could not be done as the relatives did not consent for the same.

The location of our patient’s tumor at the left CPA with the involvement of the petroclival bone is extremely intriguing. According to our hypothesis, this lesion was most probably caused by residual notochordal tissue from the sphenococipital or petroocipital spondylocondroses. Literature review showed that out of 47 reported intracranial chordomas, the majority ($n = 32$, 68.0%) originated from the prepontine cistern in the mid axial plane, whereas some authors have reported tumor origination from the parasagittal plane along the petrous apex and tentorium.⁷ Only one report by Goodarzi et al had a case like ours where the tumor arose from the CPA and was managed with staged surgical resection.⁸ Our patient did not sustain the primary surgery well and suffered postoperative bleeding and despite best efforts, he succumbed to his illness.

Conclusion

Intradural CPA chordomas are rare entities that require CT and MRI imaging for initial diagnosis. Mainstay treatment remains

gross-total surgical excision of the lesion followed by radiation therapy. A histopathological analysis is diagnostic.

Authors' Contributions

D.H. contributed to the concepts, design, definition of intellectual content, literature search, manuscript preparation, manuscript review, and guarantor. C.B. contributed to the concepts, design, definition of intellectual content, literature search, manuscript preparation, and manuscript review. S.D. contributed to the concepts, design, and manuscript review. M.T. contributed to the concepts, design, and manuscript review. A.G. contributed to the concepts, design, definition of intellectual content, literature search, manuscript preparation, manuscript review, and guarantor.

Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has given their consent for their images and other clinical information to be reported in the journal. The patient's relative understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Funding

None.

Conflict of Interest

None declared.

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