A Rare Case Report of an Intradural Left Cerebellopontine Angle Chordoma

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Abstract

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.

Keywords
► CP angle tumor
► petroclival chordoma
► brachyury
► chordoma
► skull base tumor

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)
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. Extramedullary chordomas are thought to develop from abnormal notochordal remnants in the ventral skull and are commonly seen at synchondroses in the ventral skull base, stretching from the dorsum sellae superiorly to the basion inferiorly. Development of an intradural chordoma has two possible theories.

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. Extramedullary chordomas are thought to develop from abnormal notochordal remnants in the ventral skull and are commonly seen at synchondroses in the ventral skull base, stretching from the dorsum sellae superiorly to the basion inferiorly. Development of an intradural chordoma has two possible theories.

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, definition of intellectual content, literature search, manuscript preparation, manuscript review, and guarantor. A.G. contributed to the concepts, 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.

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Conflict of Interest
None declared.

 References