

CASE REPORT

Retropharyngeal Chordoma: Uncommon Tumor at Uncommon Site

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ABSTRACT

Retropharyngeal space is a rare site of occurrence of chordomas. We present two cases of retropharyngeal chordoma with their imaging features. A brief review of the chordomas is discussed along with role of imaging in extraosseous chordomas.

Keywords: Chordomas, Extraosseous, Retropharyngeal.

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INTRODUCTION

Chordomas are rare tumors. They are typically malignant in nature. These tumors occur along the axial skeleton. The two common sites for occurrence of these tumors are the sphenoid-occipital region in the skull base and the sacrum. Chordomas have a notochordal origin.¹ Usually, these tumors are described as lesions arising from the bone. Clinical reports of nasopharyngeal chordomas, generally describe tumors with both bony and soft tissue component.²

CASE REPORTS

Case 1

A 55-year-old male patient presented with complaints of dysphasia and a sensation of lump in his throat. On clinical examination a bulge was noted in the posterior wall of the nasopharynx. Contrast enhanced magnetic resonance imaging (MRI) of the cervical spine showed a well-defined oval shaped lobulated lesion in the retropharyngeal region

at C1– C2 level (Fig. 1). The lesion was predominantly iso- to hyperintense on T2-weighted images with a few hypointense bands within. On T1-weighted images the lesion was predominantly hypointense with central isointensity. The lesion was hyperintense on fat suppressed T2-weighted images. Moderate heterogeneous contrast enhancement was noted on gadolinium administration. The underlying bone did not show any evidence of alteration in bone marrow signal intensity or abnormal enhancement on post-contrast imaging. Biopsy of the tumor showed chordoma.

Case 2

A 45-year-old male patient presented with throat pain. On examination, a lump was noted in the posterior wall of the nasopharynx. Contrast enhanced computed tomography (CT) revealed a hypodense (Fig. 2), mildly enhancing mass with calcific specks and enhancing septa within the retropharyngeal and perivertebral space. The lesion was causing expansion of bilateral neural foramina with extension of the tumor into the extradural space at C3 level. Destruction of the adjacent vertebral bodies was also seen. Subtotal excision of the tumor was done which revealed chordoma on histopathology.

DISCUSSION

Chordomas of the head and neck usually arise from the sphenoid-occipital synchondrosis in the clivus.³ They arise from the remnants of the notochord.⁴ The notochord is a group of primitive cells from which the skull base and axial skeleton develop.⁵ Chordomas usually occur in adults, most commonly in the 4th decade. Intracranial chordomas are more likely to occur in males with a male to female ratio of 2:1.¹ The tumor tends to spread locally and erosion of the surrounding bone and soft tissue may be seen. Rarely, chordomas may arise only in soft tissue without bony involvement. These are known as extraosseous chordomas and are embryologically derived from ectopic rests of notochord within the soft tissue.⁶ The most common sites of occurrence of extraosseous chordomas are retropharyngeal space, tentorium cerebelli, intradural foramen magnum, cervical and thoracic regions, and filum terminale of spinal cord.⁷ Rare sites of origin also include maxilla and paranasal sinuses.¹ Chordomas

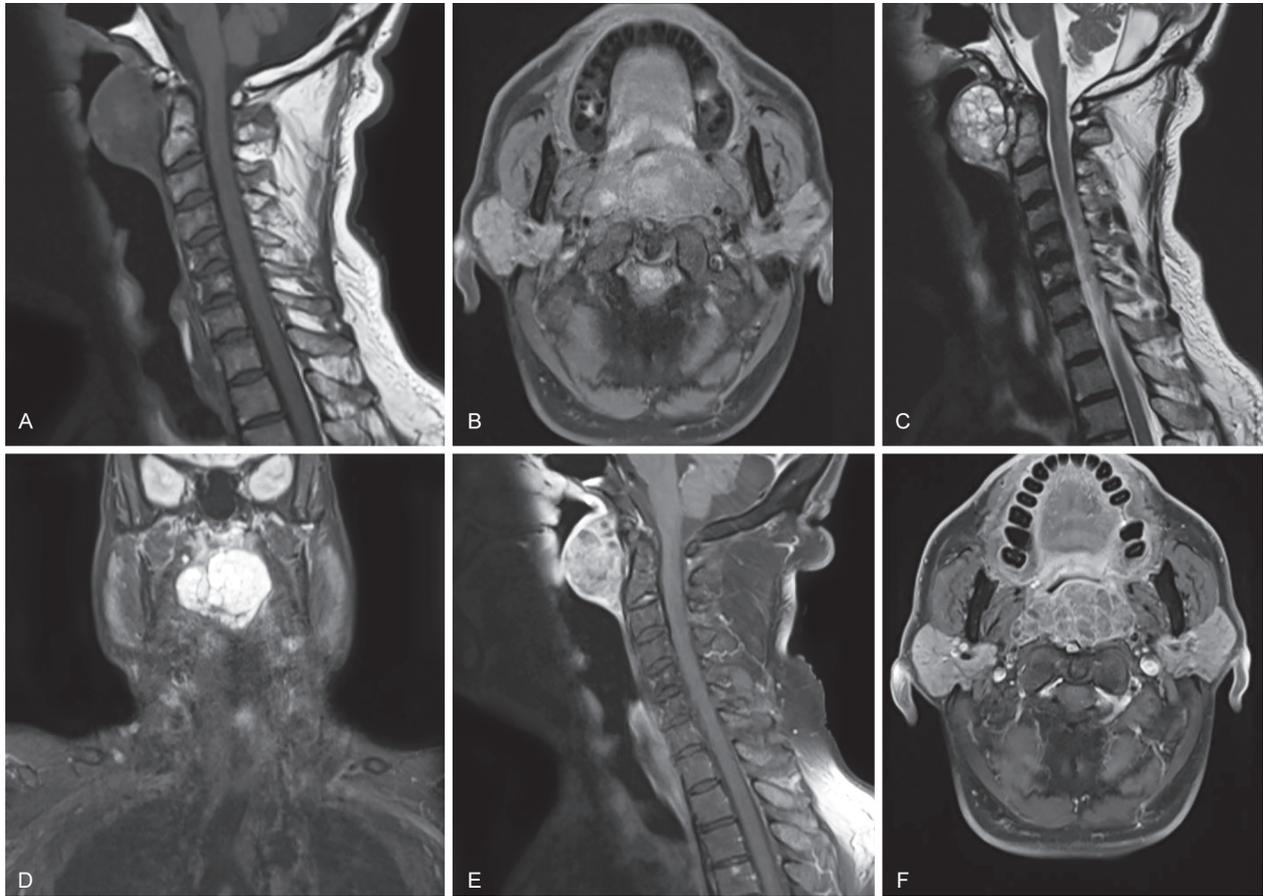
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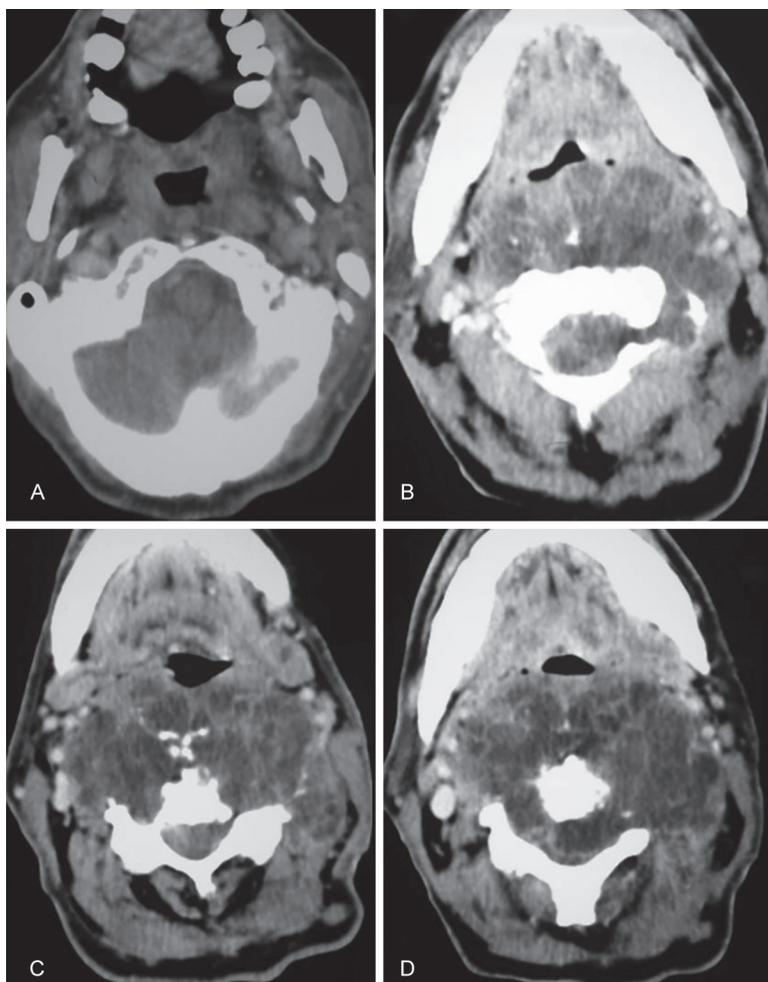
Figs 1A to F: Magnetic resonance imaging of cervical spine showing a well-defined oval-shaped lobulated lesion, i.e., predominantly hypointense with central isointensity on: (A) T1W Images; (B) T1-fat saturated images; (C) the lesion is iso- to hyperintense on T2W Images with hypointense bands within; (D) it is hyperintense on T2-fat saturated images; and (E, F) moderate heterogeneous contrast enhancement is seen

are less likely to metastasize. Distant metastasis to lung, bone, soft tissue, lymph node, liver, and skin have been reported in up to 43% of patients.⁸ Histologically, Chordomas are divided into conventional, chondroid, and dedifferentiated types.⁸ Most common amongst these are conventional chordomas. This subtype has neither cartilaginous nor additional mesenchymal components. Chondroid chordomas contain both chordomatous and chondromatous features. These are usually seen in the spheno-occipital region of the skull base. Around 5 to 15% of all chordomas and up to 33% of cranial chordomas are of the chondroid subtype. In addition, this subtype shows low grade behavior with a favorable outcome. Dedifferentiation or sarcomatous transformation occurs in 2 to 8% of chordomas.⁸ Physaliferous cells with large, sharply defined, clear vacuoles with a bubbly appearance are characteristic of chordoma.

Both CT and MRI are useful in imaging chordomas. Bony involvement is better appreciated on CT, while MRI allows better delineation of the soft tissue component. On CT, extraosseous chordomas of the retropharyngeal region appear as a well-circumscribed, expansile, hypoattenuated soft tissue mass with a lobulated

morphology. Areas of dystrophic calcification and lytic areas in the adjacent bone are seen. Areas of hyperattenuation may be seen within the lesion representing blood products and hemorrhage.⁵ A midline sinus leading from the soft tissue component may be seen in some cases representing the medial basal canal. This canal is the cephalad exit tract of the notochord when it moves from the clivus into the midline nasopharyngeal soft tissues. Tumor recurrence may result from incomplete excision of this tract.⁵

The tumor is usually hypo to isointense on T1-weighted. On T2-weighted images, high heterogeneous signal intensity is seen, representing the high fluid content of vacuolated cellular components. Heterogeneous hypointense areas due to calcification, haemorrhage, and proteinaceous content may also be seen. Fibrous septa dividing the gelatinous components of the tumor are seen on the T2-weighted images as areas of linear low signal intensity. These septa have been reported in 70% of chordomas and are characteristic features of chordomas.¹ Moderate enhancement is seen postgadolinium injection. However, calcification and cortical bone destruction may be missed MR imaging. Abnormal enhancement of the tissue on



Figs 2A to D: (A) Axial contrast enhanced CT images showing a large retropharyngeal and perivertebral hypodense soft tissue mass with enhancing septa within; (B) showing hyperdense specks within; (C) bony destruction; and (D) spread into the extradural space via enlarged neural foramina

the opposite side of the bone on contrast administration confirms bone involvement even though the cortex is not actually visualised.¹

Our first case is of a primary retropharyngeal chordoma without destruction of the clivus. The tumor shows characteristic lobulated architecture with septa on T2-weighted imaging. The second case also shows a predominantly soft tissue mass in the retropharyngeal region with destruction of C2 and C3 cervical vertebrae. The midline sinus mentioned by Nguyen et al was not seen in either case. The differential for a retropharyngeal chordoma includes nasopharyngeal carcinoma and non-Hodgkin lymphoma. To conclude, though retropharyngeal chordomas are uncommon, they should be included in differential diagnosis of retropharyngeal lesions in adults.

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