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Case Report

A rare case of large retropharyngeal pre-vertebral chordoma presented as oropharyngeal swelling

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ABSTRACT

Posterior oropharyngeal swelling can result from various pathologies, including chordomas. Chordoma is a notochordal remnant origin tumor. It is a rare bone and central nervous system tumor, especially in the cervical region. Chordoma is a tumor with properties such as slowly growing and frequently recur in most patients. This case report aims to comprehensively understand the clinical presentation and radiological features of a rare C2 retropharyngeal pre-vertebral chordoma. A 24-year-old male with a two-year history of dysphagia presented with progressively worsening symptoms, including dyspnea, dysphonia, and weight loss. On examination, a large mass was observed in the posterior oropharynx. Radiologically, a large soft tissue mass involves the C2 vertebral body, extending into the pre-vertebral soft tissue and compromising the airway. The mass appears hypointense in T1 weighted images, heterogeneously hyperintense in T2, and shows heterogeneous contrast enhancement. The chordoma was confirmed through histopathological analysis, showing characteristic physaliferous cells within a myxoid stroma. Posterior oropharyngeal masses require prompt and accurate evaluation. Radiological evaluation is crucial in identifying the tumor's origin, extent, and vascularity, aiding in diagnosis and treatment planning. Surgical resection is the primary treatment, but chordomas have a high recurrence rate even after complete removal. Radiotherapy can improve survival and reduce recurrence. This case highlights the importance of multidisciplinary collaboration in providing individualized care for rare chordoma presentations.

Keywords: Spinal chordoma, Notochordal remnant, Oropharyngeal mass, Radiological evaluation, Dysphagia

INTRODUCTION

Posterior oropharyngeal swelling can be attributed to various pathologies originating from the retropharyngeal, danger, or pre-vertebral spaces and from medial extensions of pathology from the parapharyngeal or carotid space.[1] The presence of mass in this area warrants prompt evaluation, with airway obstruction being a primary concern.[2] Differential diagnoses include edema, abscess, primary tumors, and lymph node metastasis.[3] Spinal chordoma is a differential diagnosis of posterior pharyngeal swelling, as discussed in this case report.

Chordoma is a rare, slow-growing tumor that tends to return even after treatment. It is thought that chordomas originate from the residual tissue of the notochord during embryonic development.[4] Although they are often discovered accidentally as large, compressing masses with no symptoms, they can cause significant discomfort and disability. Chordomas are most commonly found in the sacrococcygeal and sphenooccipital regions, with cervical region chordomas accounting for only 6% of all cases.[5]

Cervical chordomas might present as retropharyngeal swelling.[4] The primary management of retropharyngeal swelling depends on the degree of airway compromise and patient stability. When airway compromisation occurs, the management ranges from observation to endotracheal intubation or surgical airway.[2,3] Accurate

assessment, including a thorough history and examination, followed by radiological investigations and histopathological analysis, is crucial for proper diagnosis and subsequent management.[1,3]

This article presents a case study of a young male with a rare C2 retropharyngeal pre-vertebral chordoma-a location infrequently reported concerning these tumors. We discuss the patient's clinical presentation, etiology, radiological and histological features, and relevant findings from previously reported cases. Aim is to comprehensively understand this rare chordoma presentation and contribute to existing literature on topic.

CASE REPORT

The present case report describes a 24-year-old male patient from Saudi Arabia who visited the otolaryngology clinic with a two-year history of dysphagia. The patient's condition had progressively worsened, particularly affecting solid foods, and over the last two months, he experienced severe dysphagia, dyspnea, dysphonia, and weight loss. Mild neck pain was also reported upon extending the neck, but the patient had no history of stridor, cyanosis, cough, hemoptysis, fever, night sweats, or neurological deficit.

The patient had a medical history of type 1 diabetes mellitus for the last decade but no prior trauma, surgery, or intubation history. The general examination was unremarkable, and the vital signs were normal.

On examination, a large mass was observed in the posterior oropharynx when he opened his mouth. The surface of the mass appeared smooth, with intact mucosa. A flexible nasal endoscopy could only pass from the sides of the mass, which extends from the soft palate to the tongue's level. However, the head, neck, and cranial nerve examination revealed no abnormalities.

Further investigations revealed a soft tissue swelling at the level of C2 and C3 with a widening of the prevertebral soft tissue, causing narrowing of the airway column on Lateral X-ray. At the same time, the height of the vertebral bodies appeared preserved (Figure 1).

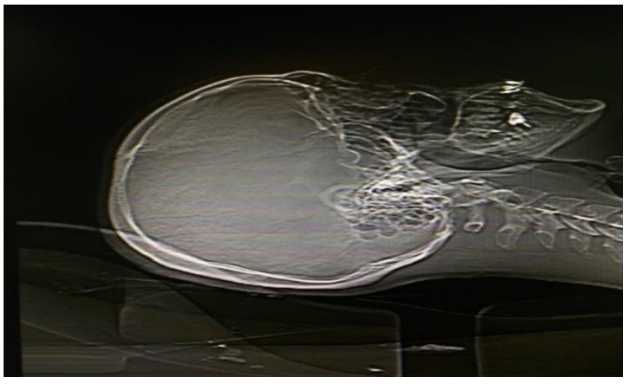

Figure 1: Lateral head and neck X-ray.

The MRI scan of the cervical spine, with and without IV contrast, showed a large soft tissue lesion that is destructive and hypointense on T1 and heterogeneously hyperintense on T2 (Figure 2 and 3). The lesion involves the C2 vertebral body and extends to the anterior and right lateral pre-vertebral space. The lesion showed a lobulated and well-defined anterior margin and appeared to arise from the C2 vertebral body. It measured about 3.1 cm in AP dimension, 5.3 cm in length, and 4.5 cm in width (Figure 2 and 4). It extended to the right transverse foramen, posteriorly displacing the right vertebral artery but appearing patent (Figure 5 and 6). There were multiple internal linear hypointensities on T2, representing multiple internal septations with heterogeneous content (Figure 2 and 6). Lesion appeared hypointense in T1 weighted images and heterogeneously hypointense in fluid-sensitive sequences, showing heterogeneous contrast enhancement (Figure 3 and 5).

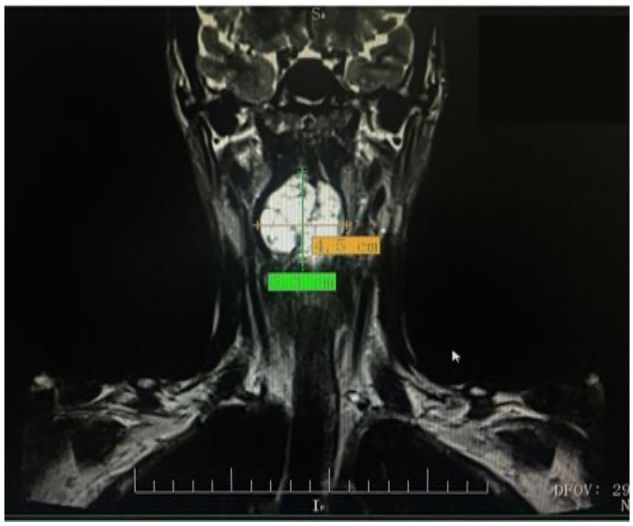

Figure 2: Coronal T2-weighted MRI of hyperintense mass in the oropharynx with multiple hypointense internal lines representing internal septations.

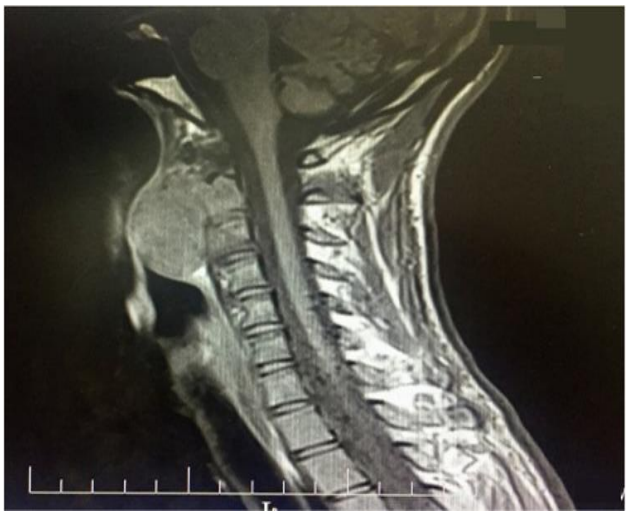

Figure 3: Sagittal T1-weighted MRI of heterogeneously hypointense mass.

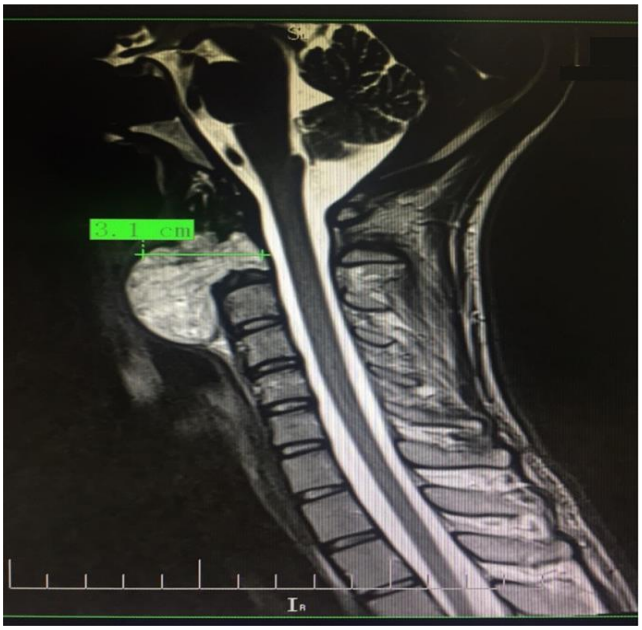

Figure 4: Sagittal T2-weighted MRI of heterogeneously hyperintense mass arising from C2 vertebra.

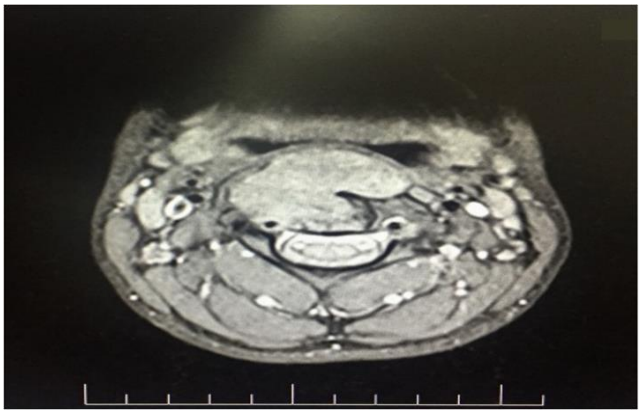

Figure 5: Axial T1-weighted MRI of heterogeneous contrast enhancement.

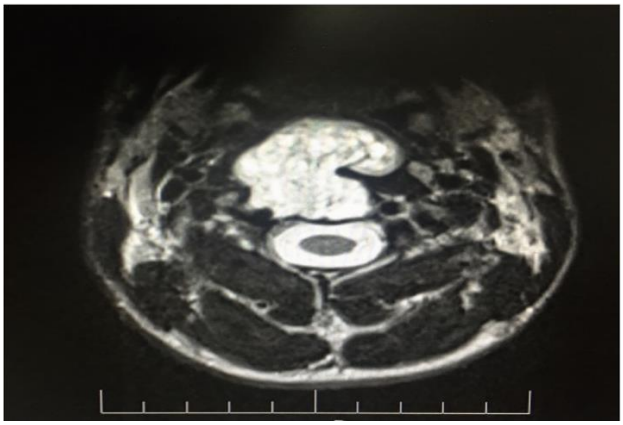

Figure 6: Axial T2-weighted MRI showing heterogeneously hyperintense mass appears compressing the airway.

The mass appeared to compress parapharyngeal spaces and the carotid vessels bilaterally. It extended from C1-C2 to the laryngeal inlet. There was no interspinal canal or epidural extension. The lesion was also seen eroding the right lateral surface of the C3 vertebral body. The rest of the vertebral bodies appeared normal. All the findings were consistent with a C2 vertebral body chordoma with pre-vertebral soft tissue extension.

The diagnosis was confirmed by a histopathological examination, which revealed a chordoma with a lobulated appearance in a cord-like fashion consistency of large epithelioid cells with calcification and sequestration of bone fragments. Characteristic physaliferous (blister-bearing) cells were seen, containing a peripheral nucleus and numerous vacuoles within the cytoplasm (Figure 7 and 8).

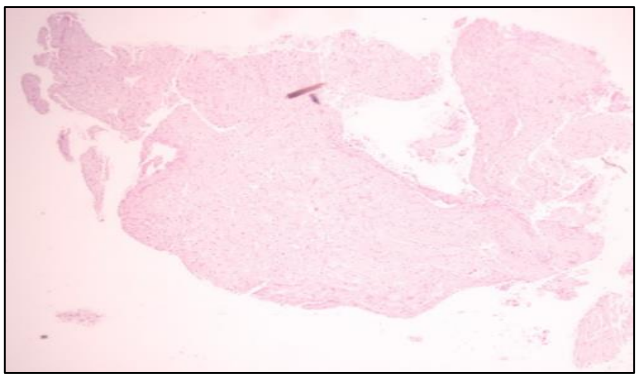

Figure 7: Histopathology with H and E stain showing tumor cells inside an extensive myxoid network material with cords of physaliferous cells.

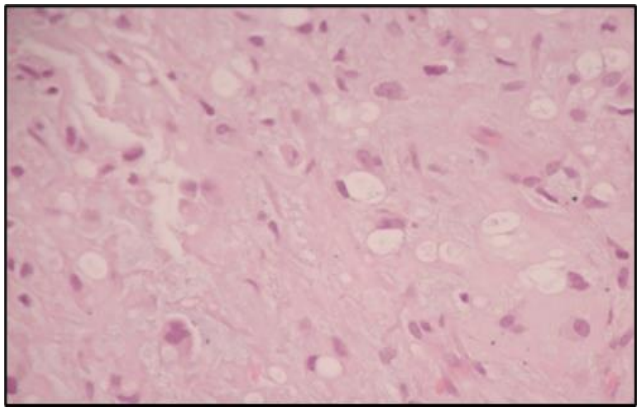

Figure 8: Histopathology with H and E stain showing numerous physaliferous cells with vacuoles within the cytoplasm and peripheral prominent nucleus.

DISCUSSION

Chordoma is a rare type of tumor that accounts for only 1% of CNS tumors and is considered one of the main causes of midline tumors affecting the axial path.[4,5] It is a notochordal remnant and is more common in males over 40, with less than 1% of cases occurring in patients under

20.[6] Chordomas are most commonly found in the sacrococcygeal and spheno-occipital regions and the cervical region of the spine, accounting for about 6% of all cases, with a higher tendency in the upper cervical region.[7]

Radiological evaluation plays a vital role in identifying the mass's origin, extent, and vascularity before any invasive procedures, such as biopsy, are considered. A contrast-enhanced CT scan is particularly valuable in providing detailed information about the location and extension of the tumor. It aids in differentiating between benign and malignant lesions and guides further diagnostic steps.[8] Subsequent MRI can offer additional clarity and help refine the diagnosis by delineating the mass and its relationship with surrounding structures.[9]

Tissue biopsy, guided by radiological findings, allows for a precise lesion characterization. Histopathology is the definitive diagnostic tool for confirming the diagnosis. The histopathology of chordoma is characterized by physaliferous cells with vacuolated cytoplasm and a prominent nucleus arranged in cords and lobules within a myxoid stroma.[10]

Surgical intervention, either via transoral or external approaches or a combination of both, is often required for definitive treatment.[4,6] While complete surgical resection of the tumor is the most effective method to decrease recurrence, radiotherapy improves survival and reduces recurrence after surgical removal.[11] Chordomas are slowly growing, locally aggressive tumors with a high recurrence rate within two years, even after complete surgical resection. The median survival rate for most patients is 6.3 years, irrespective of age, gender, and race.[12]

CONCLUSION

Posterior oropharyngeal masses require prompt and accurate evaluation. Radiological evaluation is crucial in identifying the tumor's origin, extent, and vascularity, aiding in diagnosis and treatment planning. Surgical resection is the primary treatment, but chordomas have a high recurrence rate even after complete removal. Radiotherapy can improve survival and reduce recurrence. This case highlights the importance of multidisciplinary collaboration in providing individualized care for rare chordoma presentations.

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