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Case Report

Ear, Nose and Throat Section

Extraosseous Chordoma of Retropharyngeal Space: A Case Report

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ABSTRACT

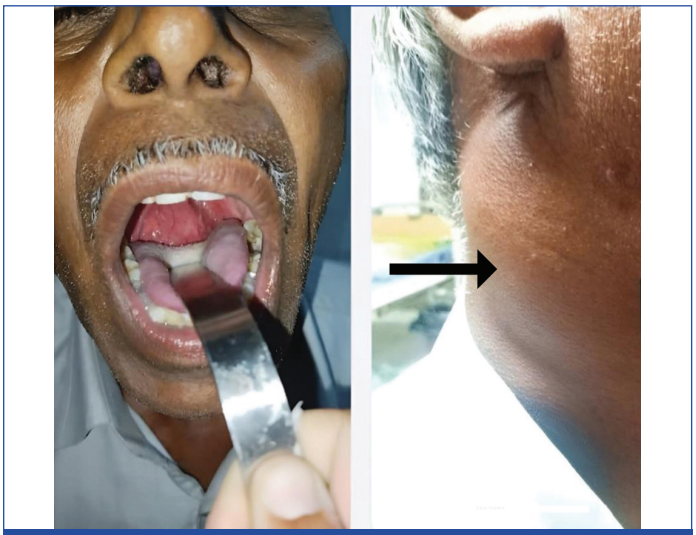
Chordomas are slow-growing malignant bone tumours derived from notochord remnants that occur infrequently, most commonly involving the axial skeleton. Extraosseous chordomas, particularly those located in the retropharyngeal space, represent a much less frequent presentation. Hereby, the authors described the initial presentation, diagnosis, and operative treatment of a 70-year-old man with progressive dysphagia and a right neck mass. Early imaging raised suspicion of a retropharyngeal abscess; however, operative exploration revealed a solid mass. Surgical resection and subsequent histopathological and immunohistochemical studies established the diagnosis of an unusual extraosseous chordoma. The novelty of present case lies in its rare presentation and location, which mimicked an infectious process typical of the retropharynx, thereby emphasising the importance of considering uncommon neoplasms in the working diagnosis. In clinical practice, present case highlights the necessity of thoroughly investigating all retropharyngeal masses, even those that appear to be infectious. The technical difficulties associated with tumour resection in this sensitive anatomical site, combined with the diagnostic challenges requiring sophisticated pathological evaluation, underscore the clinical significance of present case. Furthermore, this report contributes to the limited literature on extraosseous retropharyngeal chordomas, promoting enhanced clinical awareness and potentially informing future management strategies for this rare tumour.

Keywords: Debulking procedure, Dysphagia, Immunohistochemistry, Neck swelling

CASE REPORT

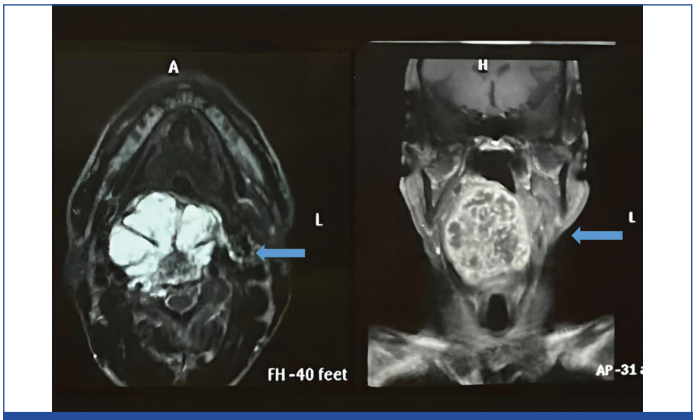
A 70-year-old male patient presented to the Department of Otorhinolaryngology with a history of difficulty swallowing for a month and right-sided neck swelling for 21 days. The symptoms had progressively worsened over this period. The patient reported a history of tuberculosis a year ago, having completed a course of anti-tuberculosis drugs (AKT), and had a history of chronic bidi smoking for 40 years.

On inspection, the oropharynx showed a bulge on the posterior pharyngeal wall [Table/Fig-1]. Clinical examination revealed a single, diffuse, non mobile, non tender, firm to hard swelling measuring approximately 4×4 cm in the right posterior triangle of the neck [Table/Fig-1].



[Table/Fig-1]: Preoperative image showing posterior pharyngeal wall bulge and right-sided neck swelling in posterior triangle.

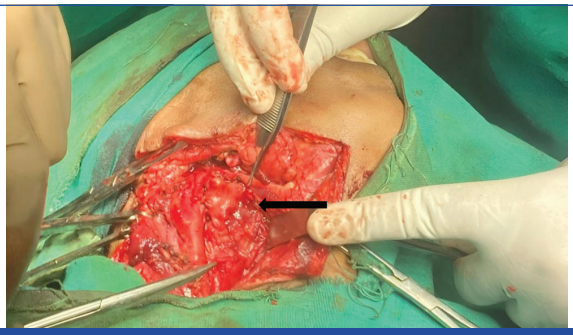
A contrast-induced Magnetic Resonance Imaging (MRI) of the neck suggested a pre-vertebral space/retropharyngeal space abscess measuring approximately 3.6×5.4×8.2 cm (anterior-posterior×transverse×cranio-caudal), extending from the lower end of the C1 vertebral body to the lower end of the C5 vertebral body [Table/Fig-2]. The lesion was noted to be causing compression and luminal narrowing of the oropharynx and hypopharynx, suggesting the possibility of a retropharyngeal abscess extending as a submandibular mass, schwannoma, or neurofibroma. The Ziel Nelson (ZN) stain for acid-fast bacilli from a sputum sample was negative.



[Table/Fig-2]: Preoperative MRI-T1 with contrast showing retropharyngeal mass.

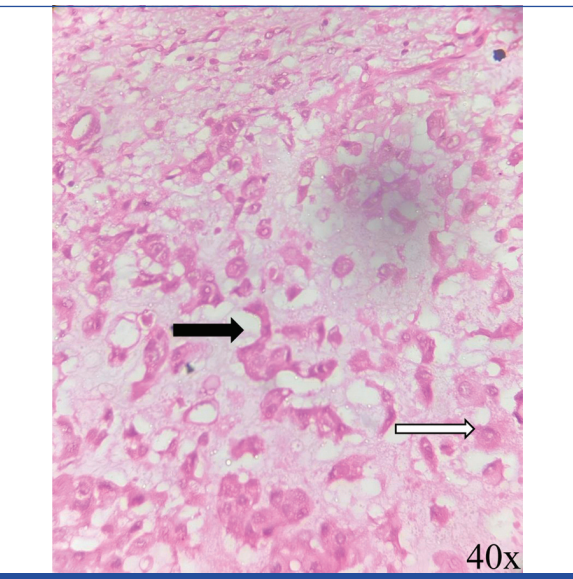
With the patient's informed consent, a debulking procedure was performed under general anaesthesia. A horizontal skin crease incision was made, and subplatysmal flaps were elevated. The right external carotid artery was identified and ligated, and the carotid sheath was exposed. The internal jugular vein, carotid artery, and vagus nerve were identified and preserved. Dissection of the strap muscles was carried out. A firm to hard mass was identified beneath the strap muscle, extending medially to the opposite side strap muscle, inferiorly to 0.5 cm above the right sternoclavicular joint, superiorly beyond the angle of the mandible, and posteriorly encroaching upon the sixth cervical vertebra. The tumour was dissected from all aspects except superiorly due to its attachment to critical structures. An incision was made over the capsule of the tumour, and a debulking procedure was performed [Table/Fig-3].

The excised tissue was sent for histopathological examination. There were no postoperative complications.



[Table/Fig-3]: Intraoperative image showing tumour.

The histopathology examination {Haematoxylin and Eosin (H&E)} report revealed a tumour composed of moderate to large-sized cells arranged in nests and single files within a myxoid matrix. These cells were large and polygonal, with some displaying vacuolated cytoplasm, imparting a "physaliphorous" appearance [Table/Fig-4]. Others exhibited eosinophilic cytoplasm. Mitotic activity was not discernible, and necrosis was not identified. Features of dedifferentiation were absent. To differentiate between chordoma and chondrosarcoma, further immunohistochemistry was conducted. The immunohistochemistry showed that the tumour cells expressed cytokeratin (Clone AE1/AE3; Dako) and Epithelial Membrane Antigen (EMA) (Clone E29; Dako). There was focal expression of S-100 (Clone S-100; Dako). The cells did not express p63 (Clone BC4A4; Biocare), Glial Fibrillary Acidic Protein (GFAP) (Polyclonal; Dako), or Smooth Muscle Actin (SMA) (Clone 1A4; Dako). A final diagnosis of chordoma was established.



[Table/Fig-4]: Histopathology specimen showing vacuolated cytoplasm (Black arrow) and myxoid background (White arrow) (H&E, 40x).

Postoperatively, the patient declined adjuvant radiotherapy. At the one-month follow-up, the patient reported relief from difficulties in swallowing, and no obvious recurrence was noted.

DISCUSSION

Chordomas are rare, slow-growing yet locally aggressive malignant tumours derived from notochordal remnants. These tumours account for approximately 1-4% of all primary bone malignancies, with an annual incidence of less than 1 per million population [1]. They predominantly occur along the axial skeleton, most commonly in the sacrococcygeal region, clivus, and vertebral column [2]. However, extraosseous presentations, where the tumour arises in soft tissue without any bone involvement, are exceedingly uncommon and may mimic neurogenic or inflammatory processes, posing significant diagnostic challenges [3,4].

This case report presents a retropharyngeal extraosseous chordoma, an atypical and rarely documented presentation. This tumour was located in the retropharyngeal space without evidence of bone erosion and clinically mimicked a benign neck swelling. Such presentations are often misdiagnosed as infectious or neurogenic lesions, which can delay appropriate management [4]. Similar to the previously documented case of extraosseous chordomas by Lee SJ et al., the initial misdiagnosis in this case as a retropharyngeal abscess underscores the difficulty in accurately identifying such tumours [4].

In contrast to spinal extraosseous chordomas, which may exhibit a characteristic dumbbell shape, retropharyngeal extraosseous chordomas may resemble common infectious or inflammatory conditions [4]. Both international and national literature has been reviewed and is presented in [Table/Fig-5] [3-6]. These cases consistently highlight symptoms such as neck swelling and dysphagia. However, subtle differences in anatomical extension, recurrence rates, and treatment modalities have been evident.

The main treatment option for chordomas is surgical resection; however, due to their proximity to vital structures and organs that limit the radiation therapy dose, complete resection is often not possible. Typically, tumour debulking is necessary prior to the application of radiation therapy, as tumour volume is a significant prognostic factor [7].

Histologically, both intraosseous and extraosseous chordomas exhibit identical microscopic features. Saint-Germain MA et al., summarised that lobulated architecture, vacuolated (physaliphorous) cells, and a mucinous background are key characteristics. Moreover, immunostaining for cytokeratin, EMA, and S100 is routinely positive [8]. In present case, S100 positivity aligned with these findings. Additionally, Brachyury, a notochord-specific transcription factor, plays a crucial role in chordoma oncogenesis and serves as a highly specific diagnostic marker [9]. Although chordomas typically exhibit moderate growth rates [10] and metastatic potential, if wide surgical margins are not achieved, relapse and local progression are frequent. Upto 40% of individuals may develop metastatic lesions, particularly in the later stages of the disease, affecting the lungs, lymph nodes, liver, and bone. The median survival is approximately 6.5 years, indicating a generally dismal prognosis [11].

New treatments have emerged for both the local and metastatic phases of the disease, but there remains a high degree of uncertainty in selecting the most appropriate treatment, leading to inconsistent adoption across the globe and suboptimal outcomes for many patients [11].

Authors name and year	Place	Age and gender	Clinical presentation	Diagnosis	Management and prognosis	Outcome
Wang Y et al., 2004 [5]	China	50/F	Sensation of lump in right-side of her throat	Extraosseous chordoma of the retropharyngeal space	Surgical excision	No recurrence at 1 year
Yang J et al., 2016 [3]	China	50/F	Right upper limb numbness	Spine extra-osseous chordoma mimicking a neurogenic tumour	Surgical excision	No recurrence at 1 year
		67/F	Right upper and lower limb pain and numbness			No recurrence at 1 year
		36/F	Saccrococcygeal discomfort			No recurrence at 1 year

Lee SJ et al., 2021 [4]	Korea	44/F	Submandibular mass and sore throat	Retropharyngeal chordoma extending to the spinal cord, mimicking a neurogenic tumour	Surgical excision	Follow-up incomplete
Vyas S et al., 2017 [6]	India	55/M 45/M	Dysphagia and a sensation of lump in throat Throat pain	Retropharyngeal chordoma Retropharyngeal chordoma	Surgical excision No mention on recurrence	Not reported
Present case	India	52/M	Dysphagia and submandibular region swelling	Retropharyngeal chordoma	Surgical excision	No recurrence at 1 month follow-up

[Table/Fig-5]: Comparative summary of extraosseous chordoma cases in literature [3-6].

It is imperative that patients with chordoma are referred as soon as possible after presentation to a skilled quaternary care facility, where all stages of the diagnostic and treatment process can be managed to achieve the best patient outcomes. The extent of resection and surgical margins are currently the most significant prognostic factors in chordoma. There is ongoing debate regarding the use of radiotherapy in the treatment of chordoma, and there is limited consensus on its role, with evidence supporting its benefits sometimes being conflicting [12].

CONCLUSION(S)

The present case of extraosseous retropharyngeal space chordoma highlights the complexities involved in diagnosis, as the mass was initially thought to be a retropharyngeal abscess based on imaging characteristics. During surgical intervention, it mimicked schwannoma, chordoma, and sarcochordoma. However, with further histopathological analysis and immunohistochemistry, it was confirmed to be a chordoma. Although extraosseous chordomas generally have a better prognosis and are less likely to recur, there is a need for more medical treatment options for this patient population.

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