ORIGINAL RESEARCH PAPER

ENT

KEY WORDS:
Parapharyngeal Chordoma, neck mass, Brachyury

AN ATYPICAL NECK MASS: PARAPHARYNGEAL CHORDOMA: DIAGNOSTIC DILEMMA IN DEEP NECK SPACE

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ABSTRACT

Chordoma is a rare malignant tumour arising from embryonic remnants of notochord. Most commonly occurring in Clivus and Sacrum. This is case report of a 73-year-old female who to ENT OPD with complains of dysphagia and odynophagia. On examination right posterior pharyngeal bulge was seen. After further investigations, including imaging and biopsy, it was found to be Parapharyngeal Chordoma. The aim of this study, is to present the clinical features, diagnostic procedures, histopathological findings and surgical management options of Parapharyngeal Chordoma.

INTRODUCTION

Chordomas are slow-growing, locally aggressive malignant tumours that represent 1% of all primary tumours arising from embryonic remnants of the notochord along the neuroaxis occurring in Clivus & Sacrococcygeal region. Rarely, it occurs in an extraosseous location and present as a parapharyngeal mass with an associated clival sinus tract. A few cases of soft tissue chordomas have been reported in the head and neck region involving the paranasal sinuses, parapharyngeal space, and soft tissues of the neck. Parapharyngeal chordomas account for approximately 1-4% of all chordomas. Extra axial Chordoma were first described by Laskowski in 1951 as '*chordoma periphericum*'. Age group 40-70 yrs.

CASE REPORT:

A 73-year-old female presented with complaints of cervical pain for 3 months, progressive dysphagia for 1 months and odynophagia for 3 days. She was a known case of T2DM and hypertension on medication. On examination, a bulge on right posterior pharyngeal wall was seen with mucosa intact. (fig1) MRI revealed a fairly circumscribed T2 hyperintense lobulated mass lesion in right parapharyngeal space without intramuscular invasion extending from skull base up to C6 vertebra. (fig 2) CECT of neck showed no bony erosion. Punch Biopsy done after incision in the mucosa under local anesthesia. The tissue was sent for histopathological study with Brachyury which confirmed the diagnosis to be Parapharyngeal Chordoma. (fig3-6) Due to large size of mass, extreme age and association of comorbidities like uncontrolled diabetes and hypertension Radiotherapy was given for 33 cycles. Dysphagia improved after 6 weeks and tumor size also decreased. Regular follow up done in every 4 weeks.

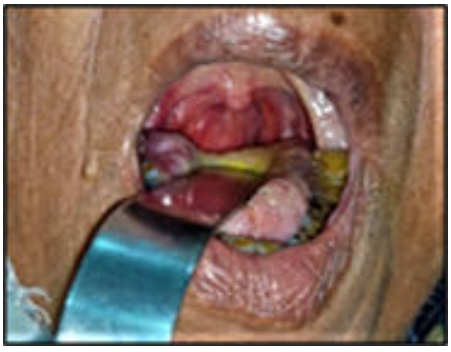

Fig 1: Clinical picture showing a Bulge on right posterior pharyngeal wall

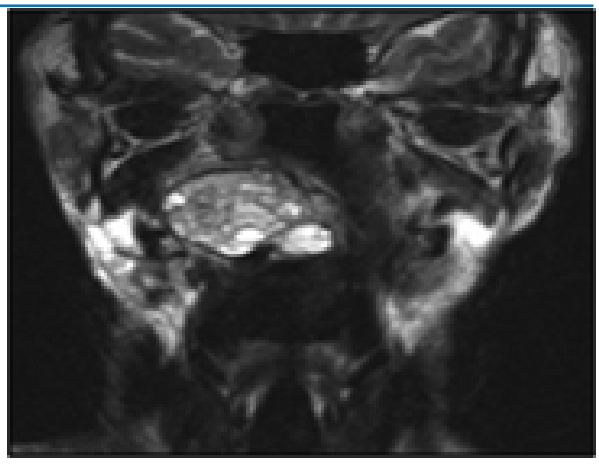

Fig 2: MRI revealed a fairly circumscribed T2 hyperintense lobulated mass lesion in right parapharyngeal space

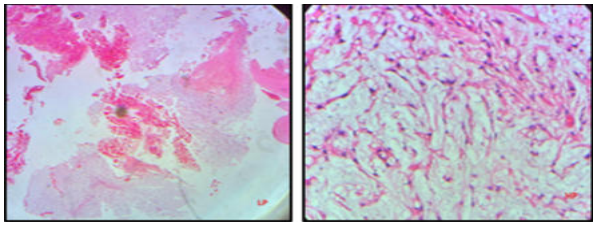

Fig 3 & 4: Histopathological examination in 10x and 40x

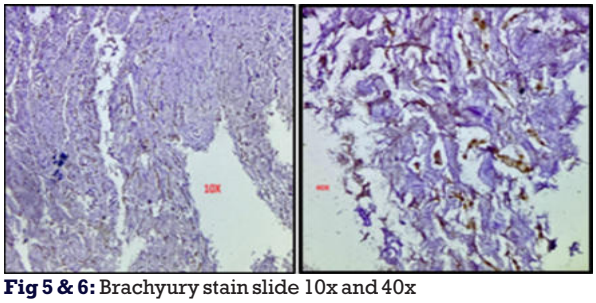

Fig 5 & 6: Brachyury stain slide 10x and 40x

DISCUSSION:

Chordoma is a rare tumour that originates from the remnant of the primitive notochord in the embryo, and it is locally aggressive and develops along the cranio-spinal axis. (4) Incidence rate of Chordoma is 0.1 per 10,000 per year. The most common location of chordoma is in the sacrum, and it

accounts for 50-60% of the cases, followed by the skull base, which accounts for 25-35% (near the sphene-occipital area). The cervical vertebrae account for 10%, and thoracolumbar vertebrae account for 5% of cases.(5) S.evans et al did a retrospective study in 131 patients which showed 2.3% of extra-axial chordoma. The onset is usually asymptomatic, so it is difficult to diagnose chordoma early. For the evaluation of chordoma, CT scans and MRI imaging are usually required. Due to the involvement of the bone, a CT scan is needed, and for the involvement of the nearby structures, MRI imaging is needed. The gold standard modality that is used in the evaluation of pre- and post-treatment is MRI imaging. On a CT scan, the tumour will appear centrally located with expansile soft tissue that originates from the clivus with the association of the destruction of the lytic bone. On T2-weighted MRI, there will be high signal intensity (4).

Chordomas can be classified histologically into three categories: classical (conventional), chondroid, and dedifferentiated. The classical (conventional) type is slow-growing and consists of physaliphorous cells within a myxoid matrix, which may show cellular proliferation. Furthermore, the chondroid type resembles a hyaline cartilage neoplastic. Moreover, the dedifferentiated type is rapidly growing and can lead to metastatic spread with the worst prognosis, and it consists of a mesenchymal component and sarcomatoid appearance. Immunohistochemical study shows chordoma will show positive stains for S100 protein, vimentin, epithelial membrane antigen, and cytokeratin . (6,7) The primary modality in management is surgery. But in 70% case negative margin is not achieved. Boriani et al reported 100% recurrence in patients treated with radiation alone. Radio therapy can be used as adjuvant treatment.

CONCLUSION:

Parapharyngeal chordomas are uncommon, often mimicking neurogenic or salivary tumors. It should be considered in the differential diagnosis of parapharyngeal masses. Due to their slow growth, it usually presents late with symptoms of dysphagia and neurological deficit. Early imaging, histopathology with brachyury, and multimodal management offer the best outcomes. The treatment options include surgical resection and radiotherapy and follow up with serial MRI. This case contributes to the understanding of chordomas in uncommon anatomical locations, warranting further research.

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