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Case Report

Clival chordoma presenting as a parapharyngeal mass: a diagnostic challenge

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ABSTRACT

Though the parapharyngeal space is sites of primary involvement by neoplastic process, they can rarely house masses descending from a base of skull tumour. Chordoma is an uncommon tumour of the skull base and sacrococcyx. Originating from the notochordal remnants, they are locally aggressive causing lytic destruction of the adjacent bony structures, particularly in the base of the skull. The use of surgery and adjuvant high-dose proton RT is documented to produce best results. Here we report a diagnostic challenge posed by a chordoma occurring as a parapharyngeal mass in a 68 year old male.

Keywords: Notochordal, Dysphagia, Sphenoid sinus, S100

INTRODUCTION

Chordoma, initially described by Virchow, is a rare, low-grade, malignant, locally aggressive bone tumour. Usually seen in the elderly, they are most often found in the axial skeleton along the line of the notochordal remnants. Though sacroccogygeal region lesions are most frequent, they can occur at atypical locations, like nasopharynx or oropharynx, wherein they are called as ectopic chordoma.[1] During the intracranial growth phase, it can cause a myriad of symptoms.[2] There are scant isolated reports documented regarding the same.[3] As they are difficult to access and give adequate clearance, it requires the close long term monitoring due to their progressive nature.[4] Herewith we present a diagnostic challenge posed by a chordoma occurring as a parapharyngeal mass in a 68 year old male.

CASE REPORT

A 68 year old male presented with the chief complaints of difficulty in swallowing and breathing for one year. On examination there was a growth in the nasopharynx. MRI of neck (plain and contrast study) showed a large enhancing heterogeneous space occupying lesion extending from the naso- and oro- pharynx upto C2 vertebral level measuring 8.0×3.6×4.0cm causing destruction of sphenoid sinus and clivus and abutting adjacent portion of internal carotid artery (Figure 1). The strap muscles of the neck, great vessels of the neck, submandibular region muscle and soft tissue are normal. Pre and para-vertebral spaces are normal. The larynx showed no abnormality or mass lesion. MRI brain showed bilateral cerebral hemispheres, brainstem and posterior fossa structures, pituitary gland, cranial vault and sella within normal limits. Chest X ray showed mild cardiomegaly and bronchitis.

Based on the clinicoradiologic findings, functional endoscopic sinus surgery was planned. Under general anaesthesia, by endonasal endoscopic approach, sphenoidectomy was done. Tissue was removed from the whole of sphenoid sinus and parapharyngeal space of left side. Hemostasis was achieved and patient was stable.

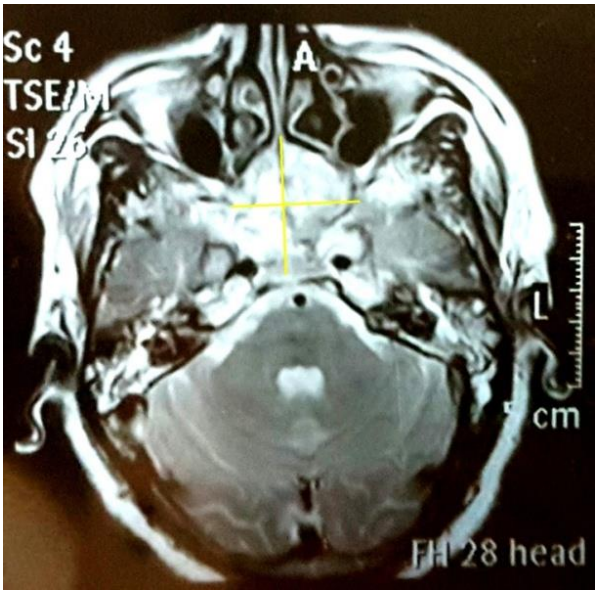

Figure 1: MRI of neck showing enhancing heterogeneous space occupying lesion extending from the naso- and oro-pharynx upto C2 vertebral level causing destruction of sphenoid sinus and clivus and abutting adjacent portion of internal carotid artery.

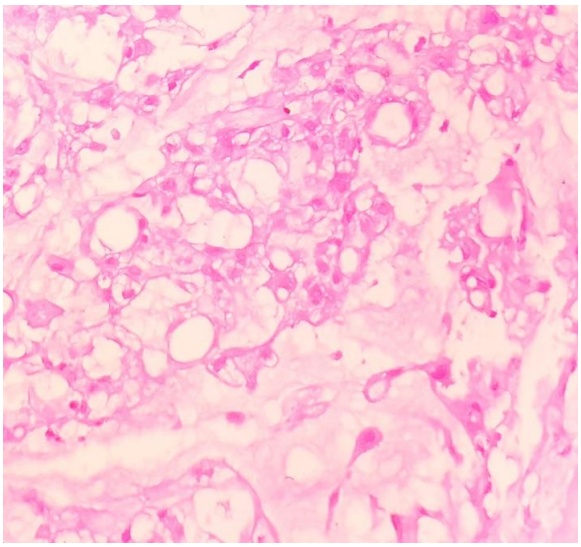

Figure 2: Section from tumour cells arranged in nest and cord pattern in a myxoid background containing large round to polygonal cells with eosinophilic to vacuolated cytoplasm and bland nucleus. (H&E, 400X).

Grossly the tissue removed were multiple irregular grey white to grey brown tissue pieces ranging in size from 0.8 cm to 2×1.5×0.5 cm. All tissue bits were embedded. On microscopic examination, sections revealed a predominantly diffuse, at places vaguely lobular, myxoid lesion, having fibrocollagenous tissue on one side. The tumour cells arranged in nest and cord pattern are large round to polygonal with eosinophilic vacuolated cytoplasm and bland nucleus. Few cells having large bubbly cytoplasm suggestive of physaliferous cells are seen (Figure 2). Mild nuclear atypia and few mitotic figures were also appreciated. As the morphologic features were suggestive of chordoma, immunohistochemistry was performed with S100 and EMA, both of which were found to be positive in tumour cells (Figure 3).

Patient was discharged and was planned for further management and is currently on follow up for the same.

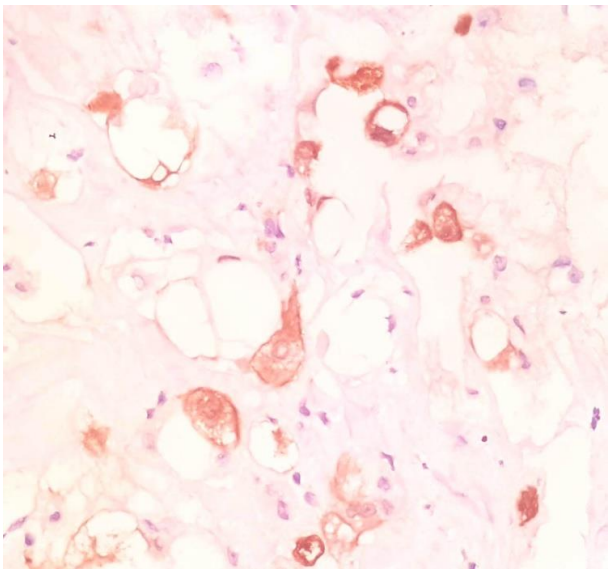

Figure 3: Section from tumour showing positive staining for S100 on immunohistochemistry (400X).

DISCUSSION

Chordoma, first described by in 1857, is an uncommon, primary malignant bone tumour, which is low grade. Conventionally they are seen in elderly patients but pediatric and younger cases are also known to occur. It is frequently found in the axial skeleton from the notochordal remnant, with majority of the cases occurring in the sacroccogygeal region, followed by spheno-occipital and spinal lesions. However, less commonly they can occur at atypical locations, like nasopharynx or oropharynx, wherein they are called as ectopic chordoma.[1]

The clinical manifestations of chordomas are dependent on the site and size of the tumour along with the direction of spread. Most of the cases of the base of skull lesion show intracranial extension which can result in vision problems, neurologic deficits and headache. It is locally aggressive nature and has documented tendency to metastasize in late stages to bone, lung, lymph node, soft tissue, skin and liver. Hence, it is considered as malignant despite its slow growth. Its deep seated nature makes it inaccessible for complete cure. Oropharyngeal chordomas are sparsely documented in literature, with about 12 cases in literature. As they present with compressive features and an intact overlying mucosa,

they spectrum of clinical presentation varies from dysphagia to nasal obstruction and breathlessness, rarely even epistaxis.[1,5,6] Cell of origin of chordomas as described by Virchow are physalipherous cells. These are found along the midline from the skull base to the axial skeleton which corresponds to the notochordal elements. It is postulated to be from the medial basal canal which can form a sinus tract.[7]

MRI and CT are useful for imaging chordomas. While MRI is useful for soft tissue extension, CT is more helpful in gauging the bony lesion.[5] Many of the cases in the base of skull lesions and head and neck region have involvement of the clivus.[8] CT shows a hypoattenuated lobular with areas of bone erosions and calcification. On MRI they are heterogeneous showing intratumoral septations.[7] Still, it is challenging to make an accurate preoperative diagnosis of chordoma.[1] As per the presentation, the possibilities of other masses in the parapharyngeal space should be considered as differential diagnosis namely, tumours of neural origin like neurilemoma, lymphoma and nasopharyngeal carcinoma.[1] For isolated sphenoidal masses rare pathologies like Langerhans cell histiocytosis, solitary plasmacytoma, chordoma, pituitary adenoma, leiomyosarcoma, fungal infection, and mucocele should be considered in the list of differential diagnosis.[9]

Histopathology shows a tumour having cells in cords and nests which are dispersed in a characteristic myxoid stroma separated by fibrous bands.[5] The cells are polyhedral with eosinophilic cytoplasm and are interspersed with a variable number of classic physalipherous cells, containing abundant vacuolated bubbly cytoplasm and a bland nucleus. Cellular atypia, mitotic figures and areas of necrosis may be present as was seen in the given case.[9] To differentiate chordoma from its mimics having chordoid morphology, positive immunohistochemistry with S100, EMA and panCK are useful.[1,10]

Despite the limitations posed by the deep seated location, surgery remains the primary modality for debulking. However, clear margins are practically not feasible, necessitating adjunct radiotherapy. Best results have been found with the use of surgery along with adjuvant high-dose proton RT.[5] After the surgical excision, disease free period of follow-up ranges from 12 months to 4.5 years.[11] The patient is planned for further management and is currently for follow up regarding the same.

CONCLUSION

Chordomas are per se, unusual entities, which orginate form the notochordal remnants along the midline. This report showcases an enigmatic clinical presentation in the form of dysphagia and dyspnea and no neurological manifestations, with a mass in sphenoid and parapharyngeal space which can readily confuse with other parapharyngeal masses. High index of clinical suspicion, consideration in differential diagnosis, radiologic information regarding the involvement of clivus and/or sphenoid, characteristic histopathologic and an appropriate immunohistochemistry panel can help in confirming and or refuting the same.

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REFERENCES

1. Li X, Wang Y, Wang F, Li B, Sun S, Yang H. An unusual case of oropharyngeal chordoma: a case report and literature review. Medicine (Baltimore). 2017;96:e8963.
2. Erazo IS, Galvis CF, Aguirre LE, Iglesias R, Abarca LC. Clival Chondroid Chordoma: a Case Report and Review of the Literature. Cureus. 2018;10:e3381.
3. Bharti JN, Dey B, Khurana N. Clival chordoma presenting as sphenoid sinus mass. Head Neck Oncol. 2014;6(2):14.
4. Zhou Y, Hu B, Wu Z, Cheng H, Dai M, Zhang B. Clival chordoma: long-term clinical outcome in a single center. Medicine (Baltimore). 2018;97:e12207.
5. Kataria SP, Batra A, Singh G, Kumar S, Sen R. Chordoma of skull base presenting as nasopharyngeal mass. J Neurosci Rural Pract. 2013;4:S95-7.
6. Khurram SA, Biswas D, Fernando M. A Parapharyngeal soft tissue chordoma presenting with synchronous cervical lymph node metastasis: an unusual presentation. Head Neck Pathol. 2016;10:400-4.
7. Nguyen RP, Salzman KL, Stambuk HE, Ahuja AT, Harnsberger HR. Extraosseous chordoma of the nasopharynx. AJNR Am J Neuroradiol. 2009;30:803-7.
8. Sajisevi M, Hoang JK, Eapen R, Jang DW. Nasopharyngeal masses arising from embryologic remnants of the clivus: a case series. J Neurol Surg Rep. 2015;76:e253-7.
9. Sadashiva N, Nandeesh BN, Shukla D, Bhat D, Somanna S, Devi BI. Isolated sphenoid sinus lesions: experience with a few rare pathologies. J Neurosci Rural Pract. 2017;8:107-13.
10. Cho HY, Lee M, Takei H, Dancer J, Ro JY, Zhai QJ. Immunohistochemical comparison of chordoma with chondrosarcoma, myxopapillary ependymoma, and chordoid meningioma. Appl Immunohistochem Mol Morphol. 2009;17:131-8.
11. Coppens JR, Ric Harnsberger H, Finn MA, Sharma P, Couldwell WT. Oronasopharyngeal chordomas. Acta Neurochir (Wien). 2009;151:901-7.

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