Unusual Presentation of Chordoma in Nose

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Abstract Chordomas are uncommon tumors of the bone representing 1–4 % of all primary bone tumors. These typically involve axial skeleton. Primary nasal presentation of chordoma is a rare occurrence and is usually misdiagnosed till the histopathological confirmation. We present a case of chordoma in nasal cavity treated by wide local resection and also present a review of literature discussing various facets in its presentation and management.

Keywords Chordoma · Clival chordoma · Epistaxis · Nasal chordoma

Introduction

Chordomas are rare neoplastic lesions with an incidence of around 8.4 per 10 million population. Overall, the median survival is 7.7 years with better prognosis in younger patients, aged below 40 years [1]. Clival chordomas usually present with intracranial symptoms such as headache and cranial nerve dysfunction. Only rarely, they involve nasopharynx, nasal cavities and paranasal sinuses and present with sinonasal symptoms like nasal obstruction, anosmia and epistaxis [2]. Here is a case report of a rare

nasal presentation of chordoma in a middle-aged female in whom gross total removal of the tumor was done endonasally followed by radiotherapy.

Case Report

A 39 year old female presented to ENT OPD with complaints of nasal obstruction and epistaxis for 1 year, headache for 6 months and decreased right sided vision for 1 month. Anterior rhinoscopy revealed pinkish mass in right nasal cavity bleeding to touch. Right eye examination showed nonreactive dilated pupil and third cranial nerve palsy (Fig. 1). There was no other neurological deficit.

Computed tomography (CT) scan of head and paranasal sinus revealed well defined heterogeneously enhancing soft tissue lesion involving bilateral nasal cavities, bilateral ethmoidal air cells, sphenoid and nasopharynx with erosion of right lateral wall of sphenoid sinus and clival destruction. Magnetic Resonance Imaging (MRI) showed well defined lobulated lesion with epicentre in right nasal cavity and sphenoid sinus (Fig. 2). The radiological impression was of a nasal mass of unusual nature.

A biopsy from the nasal mass revealed a low to moderately cellular tumour composed of cords and lobules of large cells arranged in a vague lobular pattern. The cells were polygonal and round with vesicular nucleus in abundant extracellular matrix forms which showed affinity to alcian blue. The cells displayed immunoexpression for pancytokeratin and S-100. A diagnosis of chordoma was made based on the presence of classical physaliferous cells and coexpression of pancytokeratin and S-100 (Fig. 3). The tumor was completely removed piecemeal with a combined

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Fig. 1 Clinical photograph of the patient showing **a** pinkish mass (indicated by *arrow*) in *right* nasal cavity with deviation of septum towards *left* and **b** lateral deviation of right eye in neutral gaze indicating third cranial nerve palsy

Fig. 2 CT nose and paranasal sinuses (**a**, **b**, **c**) showing heterogeneously enhancing expansile lesion filling bilateral nasal cavities and bilateral ethmoid sinuses (**a**), sphenoid sinus with erosion in right lateral wall (**b**) and extending into nasopharynx (**c**). MRI (**d**, **e**,

f) demonstrating well defined lobulated expansile heterogeneously enhancing lesion in bilateral nasal cavities (**d**), intratumor septa and amorphous calcification (**e**) with sphenoid sinus and clival extension (**f**)

facial degloving and endoscopic approach, and the patient was referred for radiotherapy. She was treated with conventional fractionation (linear accelerator) to a median dose of 50 Gy/25 fractions/5 weeks. Her vision and third nerve palsy both improved.

Discussion

Chordomas are rare malignant tumors of axial skeleton, believed to arise from vestigial or ectopic notochordal remnants alongside the neuraxis at developmentally active

Fig. 3 Histopathological picture of chordoma showing **a** large vacuolated cells in abundant extracellular matrix (H&E, $\times 400$), **b** strong expression of cytokeratin (Pan Cytokeratin: streptavidin peroxidase with DAB: $\times 400$) and **c** strong nuclear expression of S-100 (S-100: streptavidin peroxidase with DAB: $\times 400$)

sites, such as the ends of the neuraxis and the vertebral bodies. 33 % cases arise in spine, 32 % in cranium and 29 % in sacral location [3]. Chordomas are considered low-grade neoplasms that display local malignant behaviour, characterised by aggressive local growth patterns and high local recurrence rates. Distant metastasis is rare and usually from sacrococcygeal chordomas [4]. Molecular research work to improve understanding of origin of chordoma is being carried out to improve the management of this tumor. One of the very novel finding is of the expression of the transcription factor *Brachyury*, found in 90–100 % chordomas and none in genitourinary

neoplasia, metastatic germ cell tumors and clear cell renal carcinoma [5].

Clival chordomas present most frequently in fourth and fifth decades of life, with a male predominance. The clinical presentation varies with the location and extension of tumor. In all the cases of chordoma with nose and paranasal sinus involvement, two patterns of involvement are seen [6]. In one group, tumors involve clivus, sphenoid and upper nasopharynx with more frequent cranial nerve involvement and symptoms ranging from headache, visual disturbances to nasal obstruction, hyposmia, rhinorrhoea and hyponasal speech. In the second group, tumors involve

the lower nasopharynx with erosion of vertebrae and symptoms consisting of nasal obstruction, dysphagia, dysphonia, dyspnea and hearing difficulties. Our patient belonged to the first group and presented with nasal complaints and visual disturbances with predominant radiological involvement of nasal cavity and sphenoid and associated evidence of clival destruction. Patients in the latter group have more favourable outcome since they can be resected more radically.

Histologically, chordomas consist of vacuolated physaliferous cells with enormous intracytoplasmic mucoid content and an eccentric nucleus, giving a signet ring appearance. Chordoma cells do not demonstrate intracellular mucin and hence, can easily be differentiated from mucin-secreting carcinomas. Chordoma can be differentiated from adenocarcinoma by the former's carcinoembryonic antigen (CEA) negativity. In the presence of large cartilaginous component (chondroid variety), chordomas can be confused with chondrosarcoma. However, the latter do not have multi-vacuolated cytoplasm or mucinous material and are cytokeratin (CK) and epithelial membrane antigen (EMA) negative, unlike chordomas [7].

CT and MR imaging features are non-specific. However, presence of a well-defined lobular or expansile soft tissue mass, focal amorphous calcification, intratumor septa, heterogenous hyperintensity on T2-weighted imaging, and mild to moderate enhancement may be suggestive of chordoma [8]. Our patient's radiological imaging showed most of these features. Infact, the honeycomb appearance created because of enhancement pattern of this tumor probably reflects the pathological feature of organization in lobules with mucinous and gelatinous contents.

The most effective treatment of the clival chordomas is maximal tumor resection followed by postoperative irradiation. Multiple modes of radiation therapy have been tried with proton-beam therapy offering best results because of superior dose distribution due to rapid radiation falloff beyond the target. This enables higher dose delivery to tumor while decreasing injury to the vital neurological structures in vicinity [9]. The most important cause of treatment failure continues to be local recurrence but seeding along the surgical pathway may become more common with advent of more precise conformal radiation delivery methods. Hence it is advisable to consider

coverage of complete operative route in radiation treatment field [10].

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed Consent Informed consent was taken from the patient before sending this article to the journal. Additional informed consent was obtained for the possible identifying information included in this article.

Ethical Approval All procedures performed were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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