Chondrosarcoma premaxilla

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Abstract We present a very unique case of a premaxillary chondrosarcoma arising in a relatively ‘younger’ female with apparently much ‘better’ disease free 7-year survival. The literature has been reviewed and a meaningful discussion regarding diagnosis and treatment is highlighted.

Keywords Chondrosarcoma premaxilla · Canine teeth · Anterior rhinoscopy

Introduction

Although chondrosarcoma is one of the common primary bone tumours afflicting the maxilla [1], yet it is a rare entity. Berktoed et al [2] concluded its incidence in maxillofacial region to the tune of 10 %. Owing to its rarity, the published series report only a few cases and hence considerable controversy exists in the literature regarding its clinical behaviour [3]. Despite maxillary chondrosarcomas being commonly seen in the anterior part of alveolus, the occurrence of such a tumor in premaxilla is still (extremely) rare.

Case report

A 58-year-old female presented to us with a facial (maxillary) swelling and nasal blockade from past 13 years. The history reveals the onset with loosening of upper incisors and canine teeth and a corresponding swelling. Its slow progressive and painless course lead to a gradually progressive nasal blockage with cosmetic disfiguration. This irregular (maxillary) swelling extended from nasal floor, involving the premaxilla and surrounding structures predominantly towards the right side to present as a bulge in the roof of the corresponding part of oral cavity, with oral mucosa being intact. It was of firm to hard consistency, 5 to 6 cm in maximum diameter, with overlying structures being freely mobile. The upper incisor and canine teeth were absent. Anterior rhinoscopy revealed a nasal mass displacing the nasal septum on left side resulting in a near total obliteration of both the nasal cavities. There was no significant evidence of regional and distant metastases and the routine hematology / urinalysis was normal.

She had been previously treated several times with some sort of incomplete surgery in the periphery, the records of which were not available at the time of admission. However biopsy report from the palate 6 years back revealed it to be a chondroma. Moreover about 2 years prior she had received one-year course of antitubercular treatment.

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The imaging revealed non-enhancing heterogeneous soft tissue mass with intervening areas of calcification, occupying midfacial skeleton predominantly anteriorly (Figs. 1–3) in the premaxillary region more towards the right side than left. The CT scan revealed the mass obstructing both the nasal cavities lateralizing the lateral nasal wall bilaterally more on the right side. A well-marked erosion of anterior nasal septum, medial part of superior alveolar arch, anterior

Fig. 1 CT Scan axial cut at the level of the palate showing a heterogenous mass involving the anterior part of the hard palate more on the right side. Appreciate the anticipated facial deformity

Fig. 2 CT Scan coronal cut at the level of the anterior half of the nasal cavity (eyeball visualized) showing a nasal mass, eroding the septum and extending to both the maxillary sinuses more on the right side along with the pent-up secretions. Note the severe compromise of the nasal patency

Fig. 3 Approach to the tumour via the midfacial degloving approach

part of hard palate presenting as a palatal bulge could be appreciated. The right maxillary and anterior group of ethmoid sinus revealed haziness. No evidence of intracranial and orbital extension was seen.

The mass was radically excised via a mid-facial degloving approach under general anesthesia. There was gross erosion of the structures of the lateral nasal wall as the growth pushed the septum to the extreme left side. The growth was well defined and was widely excised after a proper exposure (Fig. 4). The tumour revealed varied matrix calcification and hence variable consistency throughout to the extent that the hardness was confined to its peripheral part while the inner core comprised of greyish cheesy material (Fig 5). The caudal part of septum and the lateral nasal wall were excised with the tumour along with healthy margins and the remaining surrounding bony walls were further curetted to result in a fresh hard bleeding surface. Care was taken not to breach the palatal mucosa. Thus a resultant single large surgical cavity was packed with an antibiotic / steroid ointment soaked ribbon gauge and the wound closed. Postoperative phase was uneventful and the stitches were removed after 7 days. The patient was thereafter referred to prosthodontics for proper prosthesis fitting which she ultimately could not afford due to cost factor involved.

The histopathology revealed multiloculated mass composed of proliferated chondrocytes in an abundant chondroid matrix, showing cystic changes at places. In focally distributed areas the chondrocytes were crowded together and presented mild cellular atypia characterized by anisonucleosis and hyperchromatism. The tumour lobules were surrounded by fibrocollagenous septa, which in some areas showed increased cellularity. Thus a diagnosis of a low-grade chondrosarcoma was established. A 7-year follow-up did not reveal any recurrence as per the intranasal and oral examination.

Discussion

Chondrosarcoma as such arises de-novo in pre-existing cartilaginous rest or in a benign tumour. The maxillary involvement is twice as common as that of mandible and the former has been shown to have a female predilection. Although the usual age incidence is from 3rd to 6th decade but Berrone et al [4] have observed the premaxillary involvement to occur late in 9th decade in male with a rapid progressive course. On the contrary this case happened to be a female in her 6th decade that showed an extremely slow progression. Despite continuous progression from past 13 years, the patient apart from nasal obstruction did not reveal any other symptoms such as epistaxis, paresthesias, dental pain or any cranial nerve involvement. However a painless loosening of both the upper incisors during the onset of disease points to the origin of tumour at that particular site. The chondrosarcoma has a known predilection for the anterior area, palate and the vicinity of lateral incisors and canine teeth (Batsakis 1980). It may be possible that the cartilage remnants in between the maxillary processes and premaxilla during embryonic development (fusion), may give rise to chondrosarcoma. This case revealed a prior biopsy report at a peripheral center as a chondroma, the retrospective review of slides was however not possible. It may also be possible that the benign tumour initially may have undergone malignant transformation 6 years back. Saito et al (1995) in a series of chondrosarcoma of facial bones reported a median interval from first symptom to initial treatment of one year. This case with an interval of about 6 years indeed indicates an extremely slow progression of the tumour that could have been a benign counterpart to start with. However the reliability of the biopsy report coming out to be true chondroma is questionable especially that at the peripheral center given the expertise needed to differentiate the chondroma from grade I chondrosarcoma. Moreover an empirical course of anti-tubercular chemotherapy without any documented justifications further points out towards the irrationality of the treatment.

The presence of calcification on CT scan indicates the probability of well-differentiated tumour. Considering the occasional heterogenous nature of tissue in such a lesion a MRI may provide better soft tissue delineation. Hayt et al (1998) have recommended computed tomography with multiplanar reconstructions as an excellent way to image evolving or suspected lesions of maxilla, particularly for surgical treatment planning.

Midfacial degloving approach seems to be justified in tumours located in premaxilla. The previous undocumented surgical manipulations must have been undertaken with an intent of obtaining a biopsy or might have been incomplete attempts to remove the tumour. It is well known that the post operative prognosis in chondrosarcoma improves almost in a direct relationship to the width of margin of normal tissue included in surgical resection. Histologically it is

often extremely difficult to differentiate cellular chondroma and low grade chondrosarcoma (Zachariades et al 1987). However chondrosarcoma is twice as common as its benign counterpart in craniofacial region and hence Batsakis (1987) emphasized the importance of a high degree of suspicion for chondrosarcoma unless proved otherwise. Cytological grading based on cellularity, mitosis, and deviation from cartilage phenotype is of prognostic significance. Hence the overall prognosis depends on adequacy of surgical resection, anatomical location and histological grading.

There is no doubt that radical surgical resection with a wide margin of normal tissue is the treatment of choice since chondrosarcoma is very radioresistant. Although Paddison and Hanes (1971) have reported good response of radiotherapy as well but the general consensus is against radiation (Cohen et al 1984). Radiotherapy hence should be avoided as it may itself aggravate malignant potential of an otherwise low grade tumour or else should be given as a palliative measure if complete surgical excision is not possible.

Five year survival in case of chondrosarcoma maxilla has been reported to vary from 62% to 40% (Fu & Perzin 1974; and Kragh et al 1960). Since chondrosarcoma of premaxilla is still rarer entity there have been only a few reports to 'imperfectly' predict a 5 year survival. Whether the premaxillary involvement, or female sex predilection or a presentation in 6th decade (or younger) as such, either alone or in combination are associated with a better prognosis or not (as compared to maxillary counterpart) – can only be appreciated given a sufficient number of premaxillary chondrosarcoma in literature to conclude a 'perfect' survival.

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