Myxoid Chondrosarcoma of Nasomaxilloethmoid Region with Intracranial Extension

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Abstract Chondrosarcoma of the faciomaxillary area constitute only 4 % of non-epithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx, making it one of the rare malignancy (Indian J Otolaryngol Head Neck Surg 60(3):284–286, [1]), with its myxoid variety still rarest. It is a slow-growing tumour, occurring mostly in middle-aged men. Primary chondrosarcoma of the nasal and paranasal sinus region, including the nasal septum, rarely extends into the cranial or intracranial areas unless there is recurrence (Indian J Ophthalmol 41(4):189–191, [2]). When it does occur, early diagnosis is difficult because patients generally present with common, nonspecific sinonasal complaints. A 45 year male patient came to ENT OPD with complaints of epistaxis, diplopia, facial swelling and loosening of teeth. On examination extensive swelling was present involving right maxilla, palate and ethmoid with shifting of Rt eye laterally. FNAC of swelling was non specific infected cystic lesion. On endoscopic examination there was erosion of Rt lateral wall of nose with mucoid material filling maxilla. Its wall was expanded & lined with velvety red mucosa. Maxillary antral biopsy report was myxoid chondrosarcoma. CT scan revealed extensive lesion of maxilla, ethmoid going up to optic nerve and brain. This case of myxoid chondrosarcoma is presented as it is a rare diagnosis. It presented with

advanced disease involving nasomaxilloethmoid region and extended up to optic canal and middle cranial fossa. In a thorough review of Indian literature this was a rarely diagnosed tumour.

Keywords Myxoid chondrosarcoma · Maxilla · Optic nerve · Epistaxis · Nasal obstruction · Diplopia

Abbreviations

rt	Right
CT scan	Computerized tomography
PNS	Paranasal sinus
FNAC	Fine needle aspiration cytology
NAD	No abnormality detected

Introduction

Myxoid Chondrosarcoma are malignant bone tumours in which malignant cell produce abnormal cartilage exclusively. It is an uncommon bone neoplasm of jaw usually of anterior maxilla, consisting of proliferating plump of chondroblasts. This tumour usually involve pelvis, ribs and long bones of extremities, scapular and sternum with only 5–10 % occurring in the head and neck region [3]. It is a slow-growing tumor, occurring mostly in middle-aged men. Primary chondrosarcoma of the nasal and paranasal sinus region, including the nasal septum, rarely extends into the cranial or intracranial areas unless there is recurrence [2]. Nasal obstruction, discharge, and bleeding were the major symptoms and a nasal mass was the most common sign. The typical chondrosarcoma is malignant tumour but low in grade and it presents in the nasal cavity as a

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large, pale, glistening mass usually in advance stage. We report a case of huge myxoid chondrosarcoma involving rt maxilla, ethmoid, orbital apex and middle cranial fossa.

Case report

A 45/year male patient, farmer by occupation came to ENT opd with complaints of intermittent nasal bleeding and nasal blockage in right nose since 1 year, epiphora and diminished vision right eye since 1 year, Loosening of tooth since 1 year, headache since 5 month, along with swelling of right side face which was gradually increasing in size to superior directions to involve root of nose thus shifting the eye laterally. It also extended inferiorly to involve right half of hard palate (Fig. 1) and floor of left nostril giving rise to bilateral nasal obstruction.

On examination there was a diffuse swelling involving right side of face right cheek, lateral wall of nose, root of nose thus shifting the eye ball outward and laterally (Fig. 2). There was broadening of root of nose, with increase in inter canthal distance to around 7 cm, the swelling is extending from root of nose downward to involve the hard palate, laterally up to lateral extend of maxilla, involving anterior wall of maxilla and right gingivo buccal sulcus. The surface of swelling is irregular, consistency is firm. The skin overlying swelling is free but tense.

The swelling is firm and fixed to deeper structure per oral examination the finding were rt canine and premolar teeth were loose, however no fistulous tract is present in oral cavity nor there is any ulcerative growth. The palate swelling is soft, cystic just reaching the midline. Alveolar margins are well defined. On anterior rhinoscopy there is smooth rounded swelling is seen causing bulging of floor of nose to fill the nasal cavity hence causing nasal obstruction. There is blood stained, thick copious nasal discharge present.

The hematological investigation reports were within normal limits, FNAC was inconclusive suggestive of inflammatory lesion, X ray PNS showed cystic expansion of right maxillary sinus. C T scan of P N S showed lytic lesion involving rt maxilla, rt palate, rt ethmoid, rt frontal bone and extending upward to involve rt eye displacing it laterally (Fig. 3). Rt optic nerve canal was also involved. The lesion was extending up to middle cranial fossa. X ray chest was NAD.

A diagnostic nasal endoscopy was done. The right nasal cavity had hard bulge on floor. The nasal cavity was narrow with lateral wall pushed medially there was erosion present in medial wall of maxilla at inferior turbinate area. Through this defect right maxillary sinus was approached by endoscope, it was found to be expanded and filled with thick mucoid material. The lining of cavity was red velvet

Fig. 1 Clinical photograph showing palatal bulge on right half of palate

Fig. 2 Showing shifting of eye

like. The mucoid material was sent for fungal culture in sabourds medium and bacterial culture. Biopsy of curettage specimen was sent for histopathology. The bacterial and fungal culture were negative.

The biopsy report was suggestive of myxoid chondrosarcoma. Histopathological sections showed lobules of hyaline type cartilage which was calcified at many places. The tumor periphery showed an invasive growth of neoplastic lobules into adjacent narrow spaces of ossified cartilage. The cells containing enlarged nuclei were arranged haphazardly (Fig. 4). Increased cellularity and

cells in lacunae were noted, some of which showed multinucleation. Mild cellular and nuclear pleomorphism was noted. The stroma was scanty and myxoid areas were present. Mitotic activity was low to almost absent. Neoplastic cells did not show osteoid formation. Treatment of choice for myxoid chondrosarcoma is surgery as it is relatively radioresistant. In our patient considering clinical extent and CT scan finding of extensive disease with involvement of skull base, complete curative resection was not possible and patient was unwilling for surgery; hence patient was referred for palliative radiotherapy and chemotherapy which he refused.

Discussion

The true incidence of extraskeletal myxoid chondrosarcoma in head and neck is difficult to define. In 1986, M. John G. Batsakis has mentioned four cases in head and neck. Chondrosarcoma of long bone and hip is commonly reported. In the face, they commonly involve the nasal cavity, paranasal sinuses, nasopharynx (ethmoid sinus (50 %), maxilla (18 %), nasal septum (17 %), hard palate and nasopharynx (6 %), alar cartilage (3 %), larynx, and mandible [4]. Gallagher and Strome in 1972 stressed the need of several intranasal biopsies to confirm the diagnosis of chondrosarcoma [5].

The usual age of presentation varies between 40 and 50 years, with higher incidence among males. It is an uncommon bone neoplasm in jaws usually of anterior maxilla, where preexisting nasal cartilage is present and premolar areas of mandible, a site of Meckel's cartilage. Lesions are expansile masses that produce distortion of areas. Lesions may be primary chondrosarcoma [arising directly from bone cells as malignant neoplasm] or secondary chondrosarcoma [arising in a pre-existing benign cartilaginous lesion]. In jaws, all chondrosarcomas arise *de novo* without preexisting benign chondroma. Due to their

Fig. 3 CT scan showing expansion of maxilla, ethmoid, and involvement of nose

Fig. 4 HandE stain microphotograph x 100 large chondroid cell seen in lobules in abnormal cartilage matrix with marked hypercellularity. Nuclear atypia is also seen

slow growing nature, chondrosarcomas tend to be large at presentation [2]. Symptomatology varies according to the site and size of the tumor. Usually nasal obstruction, nasal discharge, facial asymmetry, headache, restricted ocular movements, diplopia, proptosis, facial pain, and ear fullness are presenting symptoms. Extension of the lesion with intracranial involvement is characterized by loss of vision, proptosis, and multiple cranial nerve involvement [2].

Radiological imaging is essential before attempting surgical intervention. Computer Tomographic (CT) scan in coronal and sagittal planes is the most informative investigation [1] in evaluating the extent and exact location of the tumor [6]. Radiographic appearance can be variable. It may appear as an expansile 'moth-eaten' radiolucent area with indistinct boundaries containing flecks or blotchy radiopacity throughout, widening of the periodontal membrane of associated teeth is a common finding [18]. A relatively low grade of biological activity characterizes the clinical course of most myxoid chondrosarcomas. Bony destruction and calcification with an isodense or hypodense nonenhancing mass are common findings on CT scan. Chondrosarcomas with an extreme myxoid component frequently lack calcification. These tumors in the base of the skull cannot be radiologically distinguished from chordomas [3].

In general, slow growing tumors cause reactive thickening of the cortex, whereas a more aggressive high-grade neoplasm destroys the cortex and forms a soft tissue mass. The more radiolucent the tumor, the greater the likelihood that it is high grade [8, 19]. In this presently discussed case, CT imaging showed the exact extent of the lesion, which was much more extensive than what was visualized on conventional radiograph.

Magnetic Resonance Imaging, if available, is equally helpful. The CT and MR images were carefully inspected to determine the surgical margins.

Chondrosarcomas arises from primitive mesenchymal stem cells or may arise in tissues known to be formed of cartilage from nests that remain after ossification. In their gross appearance the neoplasm is well circumscribed, ovoid, lobular, gelatinous masses. A fibrous capsule may be present. Hemorrhage may occur giving it a look of hematoma. Average size of lesion may be 3–8 cm on presentation the resemblance of neoplastic cell to chondroblast, typical staining characteristic and finding areas of more mature identifiable cartilage point to chondroblastic origin of myxoid chondrosarcoma. On Low power magnification there is characteristic multinodular pattern. Nodules demonstrate round to elongated cells set in mucoid matrix. Histological criteria for the proper diagnosis of this neoplasm is the presence of many cells with hyperchromatic nuclei, two or more such chondrocytes within a lacuna and giant cell cartilage with the large single or multiple nuclei with clumps of chromatin and mitotic activity [7]. Lesion are graded 1–3 depending on amount and maturity of cartilage and anaplasticity of connective tissue. In Grade 2 and 3 areas of myxoid tissue and cystic degeneration are present consisting of proliferating plump chondroblast or spindle shaped mesenchymal cell and abnormal cartilage. The histological grading by cellularity, nuclear size and mitotic rate is reported to correlate with the rate of distant metastasis and overall survival [8].

Although the histological appearances of this tumour are generally characteristic, a differential diagnosis of chondroma, chordoma, and chondromyxoid fibroma should be considered. It is not unusual for a fine needle biopsy or even an excision biopsy to be unreliable since the specimen may not be representative of the lesion as areas of cellular atypism may be restricted only to certain parts of a well-differentiated chondrosarcoma [9]. Therefore, it is important to evaluate multiple fields to grade these tumours accurately [10]. Biopsy of the lesion is the only way to confirm the diagnosis.

Uncontrollable local diseases are the most common cause of death in these cases [11], and the rate of local recurrence is primarily dependent on the adequacy of surgical removal [8]. These tumours are not routinely offered post-operative adjuvant radiation therapy. Adjuvant radiation therapy or chemotherapy may have to be utilized for residual, recurrent disease or for palliation. Since it is described as a radio-resistant tumor due to its prolonged response time to radiation [12], The treatment consist of wide surgical excision. The extend of surgical margin depend on size and grade of lesion. Chondrosarcoma can also get implanted in an operative scar [13] or even along the tract of a needle biopsy [14]. The implanted cartilaginous cells survive even in the most hostile environments making it essential for a meticulous clearance of this tumour. Metastasis occur to lungs and other bone.

Recurrence occur after many years of primary diagnosis and treatment. The least cellular and most myxoid tumour have better prognosis. The prognosis for jaw lesion is worse than lesion at other site.

The prognosis depends upon the location and extent of the lesion, adequacy of treatment and degree of differentiation. Patients with incomplete resections requiring further radiotherapy or chemotherapy have a bad prognosis [11]. Generally, the posterior nasal cavity and nasopharynx are the sites with poor prognosis because of late presentation [12]. The worst prognosis has been implicated in centrally occurring chondrosarcomas of the pelvis, trunk, proximal extremity and head and neck areas especially the nasal cavity and nasopharynx [15]. 5 year survival rate for all stages combined is between 54 and 81 % in recent series, but disease free rate is lower distant metastases occur in 18 % patient and usually involve lung [16]. Since the grade of the tumour is an important prognostic factor [17], high-grade lesions should be treated aggressively. This case of myxoid chondrosarcoma is presented here because of rarity of presentation.

In Indian literature a few cases of head and neck myxoid chondrosarcoma are published, with a limited form of lesion of maxilla, hyoid and arytenoids tumour. Advanced myxoid chondrosarcoma of maxilloethmoidonasal area with lesion extending up to optic canal with intracranial extension is not reported in india [20].

Conclusion

This case of myxoid chondrosarcoma is presented here because of rarity of presentation, and extensive lesion extending up to optic canal and middle cranial fossa. Being a slow growing radioresistant tumor, radical surgical excision provides a good chance of cure if diagnosis is made early. However with such extensive lesion post operative morbidity compared to slow natural course should be rationally discussed with patient. Post treatment Periodic follow up is necessary to monitor for recurrences.

Compliance with Ethical Standards

Conflict of interest None.

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