Case Report

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Dentistry

CHONDROSARCOMA OF THE ZYGOMATICO-TEMPORAL REGION: A REPORT AND REVIEW OF LITERATURE

Dr. Vivek Saxena	Consultant, MDS oral and maxillofacial surgery, Delhi, India.
Dr. V Gopalakrishnan	Consultant, MDS oral and maxillofacial surgery, Delhi, India.
Dr. Rangarajan	Consultant, MDS oral and maxillofacial surgery, Delhi, India.
Dr. Vikram Singh	Reader, Consultant oncopathologist, MD (Path), DNB (Path), MNAMS, DM (histopathology), Pune, India.
Amolika Choube*	Consultant, MDS Oral Pathology, Microbiology and Forensic Odontology, Bikaner, Rajasthan, India. *Corresponding Author

ABSTRACT Chondrosarcoma in the maxillofacial region has been chiefly reported in nasal septum, anterior maxilla, mandibular ramus and body, and temporo-mandibular joint. Reports of chondrosarcoma in temporal bones, primarily involving petrous apex and mastoid, extending to the skull base have been published. A case of a 61-year-old male with a slow-growing bone mass in the preauricular region is described in this report. This is the first description of chondrosarcoma in the zygomatic process of temporal bone. Also, a brief review of the histological grading and management options is compiled hereby.

KEYWORDS : Chondrosarcoma, Temporal bone, Malignancy, Sarcoma, Surgical excision

INTRODUCTION

Chondrosarcoma, following myeloma and osteosarcoma, is the third most commonly encountered primary malignancy of bone (Limaiem et al., 2022). Chondrosarcoma most commonly occurs in the long bones of the body and is rarely encountered in the head and neck region, representing approximately 0.1% of all head and neck neoplasms (Coca-Pelaz et al., 2014; Iro & Slimani, 2021). Chondrosarcoma in the maxillofacial region has been reported mostly in areas with pre-existing cartilage (nasal septum, anterior maxilla, mandibular ramus and body, and temporo-mandibular joint). Reports of chondrosarcoma of the temporal bones have been documented, which are found mostly in the petrous apex and mastoid process where endochondral ossification occurs (Coltrera et al., 1986; Yagisawa et al., 2007). The occurrence of chondrosarcoma in the zygomatic process of temporal bone is rare. In this case report, we present a case of grade I chondrosarcoma in the zygomatic process of temporal bone in the maxillofacial region.

CASE REPORT

A 61-year-old male patient reported to the department of oral and maxillofacial surgery, with the chief complaint of swelling in the right side of face in front of the ear along with reduced mouth opening for two years. Extraoral swelling was present extending from the zygomatic prominence to the preauricular region antero-posteriorly and extending to a few centimetres inferior to the zygomatic arch superio-inferiorly. The swelling was bony hard in consistency, with smooth surface, and no abnormality was detected with the overlying skin. The swelling was tender to palpation, non-fluctuant, non-compressible, and presented with no visible and palpable pulsations. There was no regional lymphadenopathy. On the basis of slow progression of the swelling and the absence of cortical perforation, a clinical diagnosis of benign tumor of osteoarticular complex of the right temporomandibular joint was considered. Non-contrast computed tomography (NCCT) of facial bones was carried out, which revealed a well-defined sclerotic osseous sessile outgrowth arising from the zygomatic process of the right temporal bone. No cartilaginous cap and lytic component were seen within, however, cortical margins showed irregular outline, which needed ruling out of malignancy by way of histopathological examination. (Figure 1)

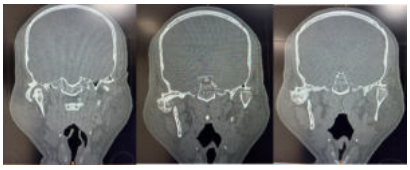

Figure 1: Sectional NCCT images showing sclerotic sessile outgrowth arising from zygomatic process of the temporal bone

An excisional biopsy was planned for the patient under general anaesthesia. A surgical model was constructed by 3D printing based on NCCT face. The zygomatic process of temporal bone and the condyle was exposed by Alkayat-Bramley incision. The growth on the zygomatic process of temporal bone was excised along with soft tissues associated with the temporomandibular joint (figure 2). The hard and soft tissues were submitted for histopathological examination. Histopathological examination of tissues from the temporomandibular joint revealed fibro-collagenous tissue. Sections from the hard tissue from zygomatic process of temporal bone showed cartilaginous and bony fragments. The cartilaginous fragments show varying sized lobules with increased cellularity of chondrocytes, with many showing nuclear atypia, binucleation, and multinucleation. Infiltration into bone, intertrabecular spaces, and surrounding soft tissue was also identified. Tumor cells was only focally positive for S-100 protein on immunohistochemical analysis (figure 3). The overall features were suggestive of grade I chondrosarcoma on clinico-pathological correlation. The patient is kept on follow-up by way of NCCT to check for any residual tumor and recurrences.

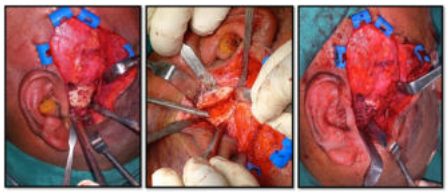

Figure 2: Intra-operative images showing Al-Kayat–Bramley approach for surgical exposure to the zygomatic process of temporal bone

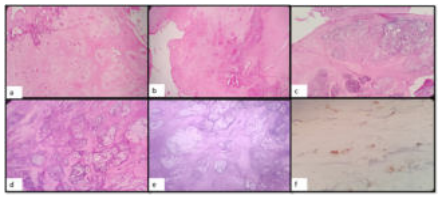

Figure 3: (a,b) Cartilagenous fragments show increased cellularity of chondrocytes under low power view (4x). (c,d,e) Higher magnification reveals varying sized lobules of cartilage tissue permeating bone, intertrabecular spaces, and surrounding soft tissues (10x). (f) Tumour cells focally positive for S-100 protein on immunohistochemical analysis (10x).

DISCUSSION

Chondrosarcomas are malignant hyaline cartilaginous tumors. They

usually arise in long bones, accounting for about 10 - 20 percent of all primary malignant neoplasms (Thorkildsen et al., 2019). Head and neck chondrosarcomas constitute 1 to 12% of those affecting the body, with the larynx, mandible, nasal cavity, and maxilla being most frequently affected. Chondrosarcomas have been reported in skull base (Konovalov et al., 2020). It has been suggested that cartilaginous precursors in the anterior part of the maxilla (nasal and septal cartilages) and in the posterior aspect of the mandible (Meckel's cartilage) are responsible for chondrosarcomas of the maxillo-facial region (Marx & Diane, 2003). Temporal bone chondrosarcoma is rare, and though cases occurring in the petrous apex, mastoid, and tympanic part of temporal bone have been described, no case of chondrosarcoma in the zygomatic process of temporal bone has been described to date (Zhang et al., 2009).

Depending on the clinical, imaging, and histopathological features, chondrosarcoma of bone can be categorized as chondrosarcoma of small bones, conventional chondrosarcoma, secondary chondrosarcoma, juxtacortical chondrosarcoma, clear cell chondrosarcoma, mesenchymal chondrosarcoma, and dedifferentiated chondrosarcoma (Inwards & Oliviera, 2013). Conventional chondrosarcomas, based on the basis of cellularity, frequency of mitoses, and nuclear atypia observed on histopathological examination, can be categorized into grades I, II, and III (Gallego et al., 2009).

Clinical features may vary depending on the subtype of chondrosarcoma. Chondrosarcomas of the jaws and skull bones are more common in men with patients presenting mostly in 4th to 5th decades of life (Karadwal & Chatterjee, 2018). Grade I chondrosarcoma is characterized by slow growth, and thus patients are usually asymptomatic or may present with pain and/or swelling. However, patients with skull base involvement may present with neurological deficits (Bovee et al., 2020). Large lesions close to the axial skeleton associated with pain are more likely to be chondrosarcomas, whereas lesions involving the small bones of appendicular skeleton are likely to be benign enchondromas (Inwards & Oliviera, 2013).

Radiographically, chondrosarcomas appear more aggressive and destructive than enchondromas. Poorly defined borders showing cortical expansion and destruction are most suggestive of chondrosarcoma (Huang et al., 2010). Other typical features are periosteal reaction, endosteal scalloping, and cortical thickening, expansion, and destruction. Magnetic resonance imaging is useful to evaluate of the extent of marrow and soft tissue involvement (Inwards & Oliviera, 2013).

Histopathologically, grade I chondrosarcomas can be difficult to differentiate from enchondromas. Increased cellularity and pleomorphic chondrocytes are typical of chondrosarcomas. Chondrocytes evade or permeate pre-existing normal bone. Binucleate and multinucleate cells may be found along with mucoid or myxoid change in matrix. These features are important to differentiate enchondromas from grade I chondrosarcomas (Inwards & Oliviera, 2013). While mitotic figures are typically absent or rare in grade I chondrosarcomas, grade II chondrosarcomas usually show less than two mitoses per 10 high-power fields. Also, cellularity and atypical features are more in grade II chondrosarcomas and stroma appears more myxoid. Grade III chondrosarcomas present with greater cellularity and nuclear atypia, and show higher mitotic activity (more than two mitotic figures per 10 high-power fields). Spindling of cells and less differentiated forms are also indicative of grade III chondrosarcoma (Evans et al., 1977; Bertoni et al., 2002). Grade I chondrosarcomas are locally aggressive and do not usually present with distant metastasis. Histological features like grade, mitotic count, tumor necrosis, and myxoid matrix are usually predictive of risk of recurrences and distant metastasis (Evans et al., 1977).

Chondrosarcomas are considered to be resistant to chemotherapy and radiation, thus surgical excision is the mainstay of treatment (Sundaresan et al., 2004; Takaishi et al., 1996). Intralesional curettage, burring, and application of hydrogen peroxide as adjuvant to surgery have been proposed (Leerapun et al., 2007). Wide local excision is treatment of choice for grade I chondrosarcomas, while wide en bloc excision is satisfactory for grades II and III chondrosarcoma (Gelderblom et al., 2008). Two to three centimetres of surgical clear margin is important for favourable prognosis and for a decreased risk of recurrence, even when the cortex is intact (Carlson et al., 2004). Cryosurgery has been suggested for the management of grade 1 chondrosarcoma. Results obtained from the combined approach of cryosurgery and intralesional excision are comparable, and better, to those of marginal excision and wide excision of grade 1 chondrosarcoma (Veth et al., 2005). The use of cryosurgery can be associated with complications such as infection, neuropathy, and embolism. Infection is the most common associated risk in the head and neck region, especially when cryosurgery is combined with reconstructive procedures (Sammartino et al., 2008). In cases of residual, surgically inaccessible, and histologically identified high-risk tumors (grades II and III), radiation therapy is indicated to prevent local recurrences (Suit et al., 1982; Raza et al., 2017). Patients with high grade and dedifferentiated chondrosarcomas and may benefit from chemotherapy (Raza et al., 2017). Distant metastasis for grade I chondrosarcoma is less than 10 percent. Five-year survival rate corresponds to the histologic grade, 89% for grade 1 chondrosarcoma and 53% for grades II and III (Gelderblom et al., 2008).

CONCLUSION

This is the first case report of a chondrosarcoma arising in the zygomatic process of temporal bone. Chondrosarcoma in temporomandibular joint has been reported in literature, the clinical presentation of which, similar to the present case, is usually slow-growing swelling in the preauricular region with decreased mouth opening. In this case as well, the presentation indicated towards a benign chondrogenic or osteogenic neoplasm of the temporomandibular joint. NCCT face and 3D printed model were very helpful in localizing the origin of the neoplasm (from the zygomatic process of temporal bone instead of the temporomandibular joint). Histopathological examination was, however, suggestive of a grade I chondrosarcoma. The patient is currently on follow-up through clinical examination and NCCT face to ascertain the extent of residual tumor and to follow- up for recurrences.

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