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Case Report
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Chondromyxoid fibroma of the zygoma: a case report

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Abstract. Chondromyxoid fibroma is a rare benign tumour of chondral origin. It usually involves the long bones of the lower extremity, whilst involvement of craniofacial skeleton is extremely unusual. The second case of chondromyxoid fibroma of the zygoma described in literature is presented and the surgical resection of the lesion with tumour-free margins as the key factor for avoiding local recurrence of this tumour is emphasised.

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Chondromyxoid fibroma is an unusually benign tumour of cartilagineous origin, and one of the less common tumours of bone, comprising less than 1% of bone tumours[3]. The neoplasm was first described in 1948 by Jaffe and Lichtenstein as a separate entity from chondrosarcoma or benign chondroblastoma, having slightly different plain film characteristics and a benign clinical course, so that treatment and prognosis are different[4]. It is histologically characterized by varying proportions of lobular, spindle-shaped, or stellate cells in a myxoid background. Chondromyxoid fibroma occurs most frequently in the bones of the lower extremity, most often the proximal tibia[8].

Craniofacial bones, instead, are involved in only 2% of cases[1]. In particular, the neoplasm occurs in the mandible, whilst only one case of chondromyxoid fibroma of the zygoma has been reported in literature[2]. The peak age incidence is in second and third decades of life with no predominance between sexes[6].

Case report

A 51-year-old man was referred to the Department of Oral and Maxillofacial Surgery, University ''Federico II'' of Naples with a large and painless swelling of the right zygomatic region in October 2002. The patient had noticed this slowly growing expansile mass 6 months before.

CT scan showed a 3 cm × 3 cm osteolytic lesion in the right body of zygoma with lobulated and well-demarcated margins (Fig. 1A). Expansion of cortical bone and partial destruction at the lateral wall of the orbit was present. Radiological findings were consistent with a benign bone tumour.

The tumour was approached using a Weber–Ferguson incision with Lynch extension and it was resected widely enough to include a rim of normal bone (Fig. 1B). Surgical defect of zygomatic arch was reconstructed using a calvarial bone graft fixed with microplates and screws (Fig. 1C). Post-operative clinical course was uncomplicated. Finally, at follow-up after 2 years, a CT scan did not show any sign of recurrence (Fig. 1D).

The specimen was sent for pathological examination and diagnosed as a chondromyxoid fibroma. Grossly, the tumour was solid and yellowish-white, well demarcated from the surrounding bone and thins the cortex.

Microscopically, it was composed of hypocellular lobules with a myxochondroid appearance, separated by bands of hypercellular tissue composed of stellate and spindle-shaped cells. Focal coarse calcification was seen. Mitotic figures and necrosis were absent (Fig. 2A and B). It is extremely important to distinguish chondromyxoid fibroma from chondrosarcoma because surgical management of this aggressive tumour is different.

Discussion

Chondromyxoid fibroma is a rare benign bone tumour arising from cartilage-form-

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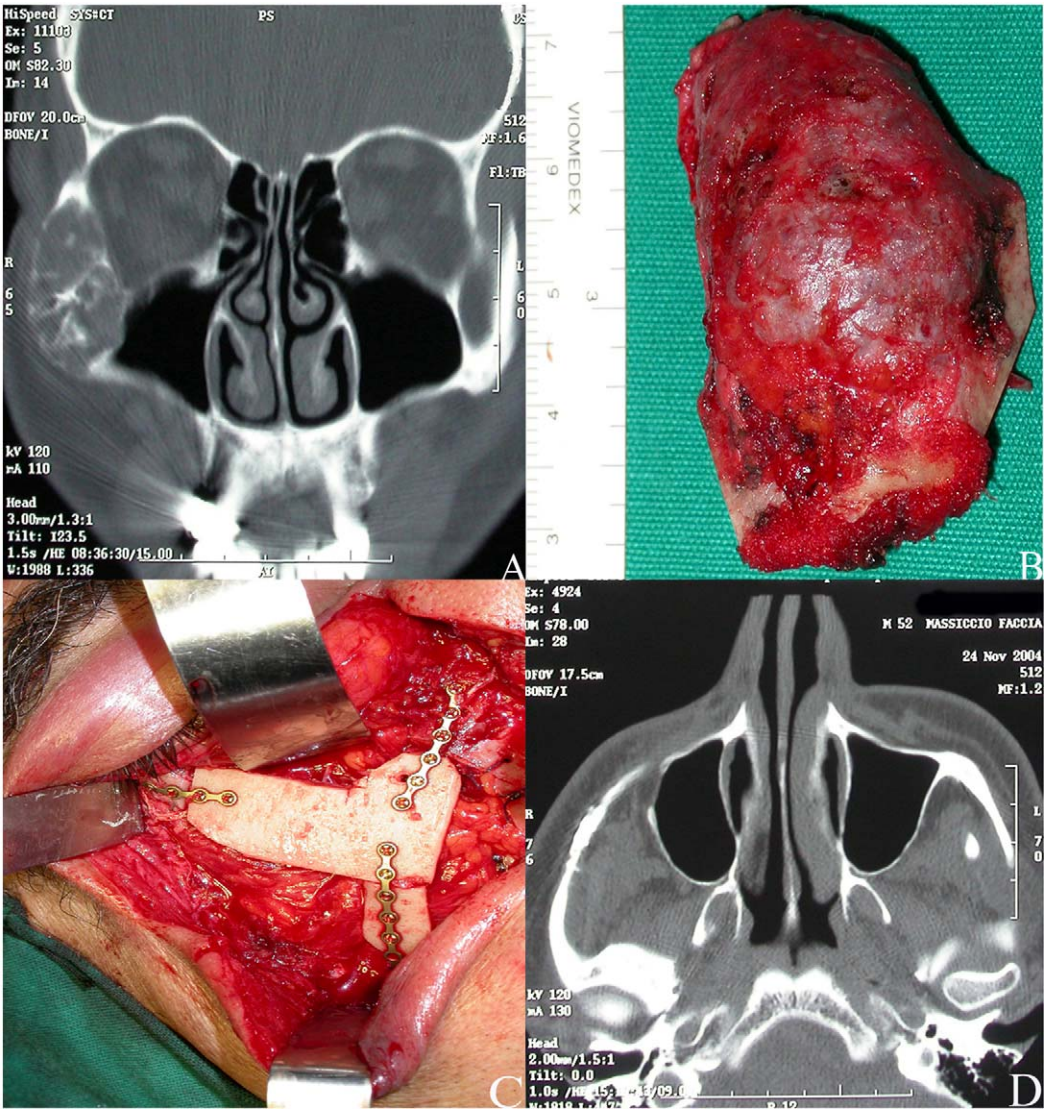

Fig. 1. (A) Coronal CT scan of chondromyxoid fibroma of the zygoma. There is an osteolytic lesion in the right body of zygoma with lobulated and well-demarcated margins. Expansion of cortical bone and partial destruction at the lateral wall of the orbit is present. (B) Surgical specimen. Tumour was resected widely enough to include a rim of normal bone. (C) Surgical defect was reconstructed using a calvarial bone graft fixed with microplates and screws. (D) Post-operative CT scan after 2 years showed no signs of recurrence.

ing mesenchymal tissue, accounting for less than 1% of all bone tumors[3]. It generally affects long bones of lower extremity. The occurrence of this tumour in the skull is extremely rare. In the jaws, the neoplasm typically occurs in the mandible[6]. Initial symptoms are pain or a slowly growing expansile mass. Histologically, the classic features include a lobular pattern with stellate or spindle-shaped cells in a myxoid background. The differential diagnosis includes chondroblastoma as well as chondrosarcoma. Surgical treatment varied from conservative curettage to block resection.

When this kind of tumour is localized in the facial skeleton, many authors recommend curettage followed by a strict follow-up for avoiding cosmetic and functional sequelae of bone resection[5]. Recurrences are not uncommon, especially when curettage is the method of treatment. Recurrence rates of 25% after curettage have been reported[6]. A complete resection including a rim of normal bone because of the possibility of recurrence is recommended. Malignant transformation to chondrosarcoma has been reported in literature, but it is likely a misdiagnosis, especially when the tumour occurs in an unusual location or in older patients[7]. Radiotherapy is not recommended because of the potential for sarcomatous conversion[3]. Chondromyxoid fibroma is benign tumour, but recurrence is possible especially when treated by curettage alone. In this case, the resection with tumour-free margin was completely curative.

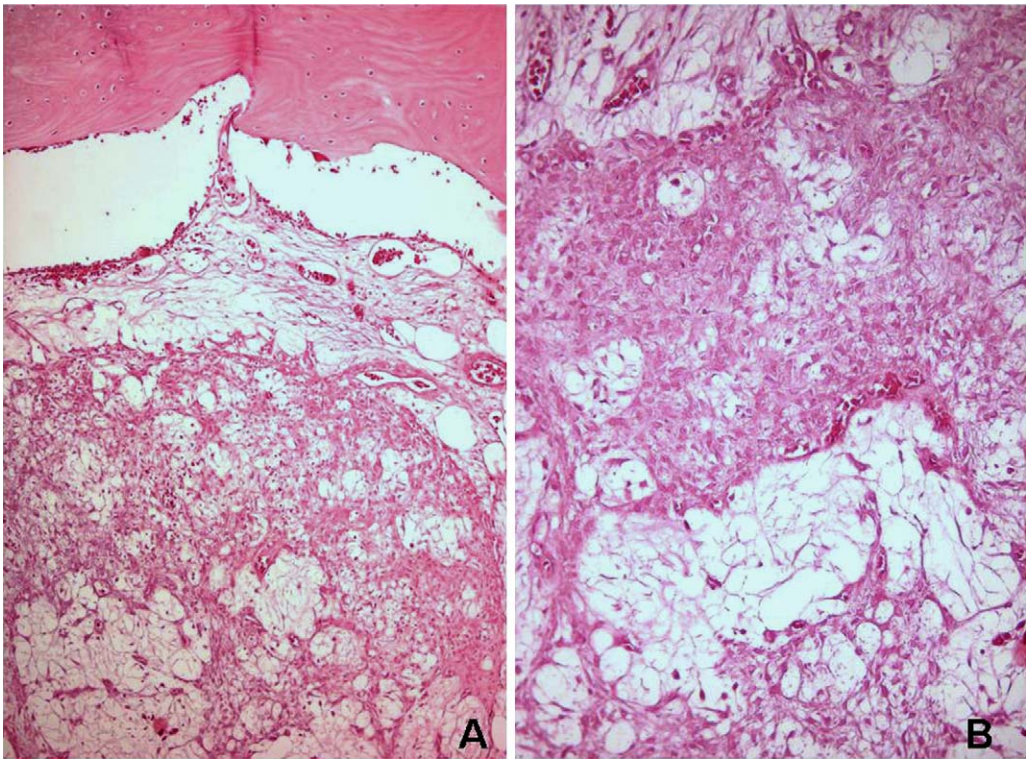

Fig. 2. (A) Well-circumscribed tumour with a lobulated appearance (Hematoxylin and Eosin ×106). (B) Myxochondroid hypocellular lobules alternate with more cellular foci (Hematoxylin and Eosin ×250).

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