Giant Myxoid Osteoma of the Frontoethmoidal Region

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ABSTRACT

Although they are relatively rare, osteomas are the most frequent benign neoplasm of the paranasal sinuses. When they are large or symptomatic, surgical intervention through open or endoscopic approaches is required. The association between osteoma and myxoma is rarely addressed in the literature and myxoid degeneration of osteomas is not known. We report a case of a giant frontoethmoidal osseous lesion in a 52 years old man which was completely removed through an open approach. Histological examination of the removed mass revealed an osteoma with a myxoid component.

Keywords: Osteoma, Myxoma, Intraosseous myxoma, Frontal.

INTRODUCTION

An osteoma of the paranasal sinuses is considered large or giant when its size is more than 3 cm. Giant osteomas of the paranasal sinuses are rare but readily extend into the intraorbital or intracranial cavity, causing serious complications. A giant osteoma of the sinuses usually requires surgical resection. The classical method for removing frontal and ethmoid sinus osteomas is still considered to be surgery via an external approach, including bicoronal scalp flap surgery, frontal craniotomy or a lateral rhinotomy. Myxomas of bone are an uncommon benign tumor of connective tissue that is seen almost exclusively in the maxilla and mandible when documented in the head and neck. They are very rarely reported in the frontal sinus, vomer and sphenoid sinus. These tumors are characterized by their generally benign, expansile appearance on radiography, however, locally aggressive imaging characteristics have been described as well as a propensity for local recurrence following excision. Myxoid degeneration of osteomas is not known and the association between myxoid and osteoid content of the same tumor is only reported once. In this work, we report a case of unusual giant myxoid osteoma of the frontoethmoidal region in a 52 years old male.

CASE REPORT

A 52 years old male patient presented with 4 months history of facial deformity, occasional diplopia and diminution of vision. On examination, there was left eye downward and outward proptosis with limitation of upward gaze. CT revealed an ill defined lobulated outline dense sclerotic lesion which harbors lucent areas inside and measures 42 × 33 × 45 mm involving and obliterating both frontoethmoidal recesses more at the left side with intraorbital extraconal extension. This was associated with another soft tissue density mass (mucocele), about 20 × 15 mm completely opacifying left frontal sinus and eroding left superior orbital roof with intraorbital extraconal extension (Fig. 1). There was no optic nerve compression. MRI showed the mass to be ill defined and displaying iso-intense signal at T1 with high signal at T2 with mild heterogenous postcontrast enhancement. There was evidence of minimal intracranial extradural extension. After obtaining an informative consent, the patient was taken to the operating theater where complete resection was performed. Pathological examination revealed an osteoma with a myxoid component.

Fig. 1: Preoperative CT showing osseous lesion with heterogeneity
removal of the mass was done through a wide osteplastic flap technique utilizing a bicoronal incision. The associated left frontal sinus mucocele was drained with aspiration of thick mucus upon opening the left frontal sinus. The mass was found hard bony in consistency and there was some bleeding upon removal. Due to the large size of the mass, it was not possible to be removed except after it was fragmented into three pieces using osteotomes and a cutting burr. There was no insult to the dura or to the orbital contents. After complete removal of the mass, reconstruction of the frontal bone was done using a miniplate and the wound was closed as usual. Postoperative course was uneventful and the patient was discharged 2 days after surgery. Histological examination of the specimen revealed vascularized tumor of spindle to stellate cells with myxoid stroma and mature sclerotic bone islands. It showed marked PAS and Alcian blue staining of the myxoid areas (Figs 2 to 4). Follow-up of the patient revealed complete disappearance of the preoperative proptosis, and follow-up CT revealed complete excision of the mass with no recurrence up to 12 months postoperatively (Fig. 5).

DISCUSSION

Myxomas are benign neoplasms derived from primitive mesenchyme. The histological identification of a benign myxoma of bone relies on the presence of characteristic benign appearing spindle cells in a myxoid stroma with the absence of pleomorphic spindle cells arranged in a lobular pattern more characteristic of entities such as a chondromyxoid fibroma or extensive chondroblasts with myxoid liquefaction more commonly seen in chondrosarcomas. The distinction between fibromyxoma and myxoma may be more difficult and likely relies on the relative degree of fibrous and myxoid stroma components.

The association between myxoma and osteoma of the paranasal sinuses is rarely addressed in the literature. Nilles and Horny reported on a case of a 24 years old male
with an unusual mesenchymal tumor in the frontal sinus. Microscopic and histomorphological aspects demonstrated a benign tumor with biphasic differentiation (a myxoid and an osteoid part). They classified the tumor as fibromyxooosteoma. They suggested that the association of two different tumors was not probable because of the extremely narrow topographic link between soft tissue and osteoid tumor parts. They also argued against a perifocal myxoid reaction of the osteoma because myxoid degeneration of osteomas is unknown and that appropriate immunohistochemical methods were able to show a relatively high proliferation activity of the spindle-shaped cells in the myxoid tissue. Their patient has remained free of any recurrence 2 years after complete removal of the tumor.8

We report a case of a giant myxoid osteoma of the frontoethmoidal region with intracranial and intraorbital extension which, to our knowledge, is the 1st reported in the English literature. Our patient had a rapid rate of growth of the tumor. The CT and MRI appearance of the tumor were characterized by excessive heterogeneity. The tumor was hard in consistency during removal and histological examination showed an osteoid component with an intraosseous myxomatous component. Whether this is a myxoid degeneration or a separate myxoid growth is confusing. Because myxoid degeneration of osteoma is not a known pathological entity, myxoid osteoma seems to be the most appropriate histologic classification of the tumor. The tumor was completely removed and there was no recurrence through the follow-up period. The myxoid content of the tumor would add some properties to the tumor as the rapid rate of growth, the increased vascularity, the tendency to local recurrence and a potential for malignant transformation and hence, conservative management is contraindicated and urgent complete removal and meticulous follow-up are required. Suspecting this myxoid content would contraindicate conservative approach to osteoma management and add some urgency to the surgical intervention.

CONCLUSION

An unusual myxoid component of a giant frontoethmoidal osteoma is reported. This myxoid component would add some properties to the osteoma as a relatively more rapid rate of growth, heterogeneity on imaging, tendency to local recurrence and a potential for malignant transformation and hence, conservative management is contraindicated and urgent complete removal and meticulous follow-up are required.

REFERENCES