Recurrent odontogenic myxoma of maxilla: a diagnostic and operative dilemma

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Abstract Odontogenic myxoma of maxilla is a very rare occurrence. It is a slow growing benign, locally malignant tumor notorious for recurrence. Hereby we present a case of recurrent odontogenic myxoma of the maxilla in a 32-year-old male managed surgically by partial maxillectomy. The diagnostic and operative dilemmas encountered while managing the case are discussed in detail.

Keywords Odontogenic myxoma · Maxilla · Recurrent

Introduction

The odontogenic myxoma is a benign, slow growing locally aggressive neoplasm of ectomesenchymal origin. It occurs almost exclusively in the jaw bones and accounts for 0.2% to 17.7% of odontogenic tumors [1, 2]. It predominantly involves the mandible, and maxillary tumors are known to be more aggressive than tumors involving the mandible [2]. It is prudent to differentiate the tumor from other fibro-osseous lesion as the chance of recurrence in Odontogenic Myxoma is significantly higher than other fibro-osseous lesions which involve the jaw bones [1, 2]. The diagnosis is on the basis of clinical, radiological and histological features [3, 4, 5]. Treatment ranges from simple curettage to radical excision especially in recurrent cases [6].

Case report

A 28-year-old male presented to the otorhinolaryngology outdoor in the year 2004 with a history of swelling in the left side of the face for the last 1-year. The swelling started insidiously and was gradually progressive in nature. On examination, a huge swelling was seen involving the left maxillary region and upper alveolus. It had a smooth surface with a very well defined outline. There was no superficial ulceration, scar, sinus or fistula. The cheek skin and alveolus was not breeched. There was no problem with vision or sensation over the affected area. There was no pain or any tenderness or fluctuation on palpation and the swelling was not adherent to the overlying skin. However it was not mobile and was very firm to bony hard in consistency.

The patient was admitted and subjected to routine investigations. FNAC and CT scan were carried out. FNAC gave a possible diagnosis of fibroosseous lesion of maxilla based on histology and origin of the tumor. CT scan revealed a smooth homogenous mass arising from the anterior wall of left maxilla with slight erosion of outer table

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of maxilla. It had well defined margins with fine interlacing trabeculations but no signs of calcification or ossification within the substance of tumor (Fig. 1).

Depending on these findings, a decision to carry out excision of the mass through a sublabial approach under general anesthesia with orotracheal intubation was taken, expecting a benign mass and easy removal. A smooth pinkish hard mass was easily enucleated from the anterior wall of the maxilla and alveolus and no underlying bone defect and erosion was noted. Biopsy of mass revealed a diagnosis of myxofibroma. Follow up for 6 months was uneventful.

Subsequently in January 2006, the patient came back to us with a complaint of reappearance of the swelling on the same area for the last 6 months. This time it was progressing faster than before. He also complained of loss of two upper left premolars in the last 2 months. On examination, the swelling was involving the left maxillary region and was of similar size as before. There was no obvious palatal bulging but the left upper alveolar margin was irregular and broadened. The mass was also invading the left nasal cavity and presenting as reddish globular mass, which was bony hard on palpation (Fig. 2). Repeat CT scan, revealed similar CT characteristics of the mass as earlier but the extent of the tumor was much more than the previous presentation. The mass was involving the left upper alveolus with obvious signs of erosion. It was also filling the left maxillary antrum with partial erosion of medial wall and extension into left nasal cavity (Fig. 3). Based upon the rapid recurrence and aggressive behaviour of the tumor a decision to undertake a more radical approach was taken and the patient was prepared for partial maxillectomy.

Operative note

Operation was carried out under general anesthesia with orotracheal intubation. A weber fergusson incision used and the alveolar cut was limited only to the affected area

Fig. 1 CT scan showing well circumscribed heterogeneous mass involving anterior wall of left maxilla with erosion of outer table

keeping a margin of safety (Fig. 4). Maxilla including the alveolus with remaining teeth and hard palate was removed en masse (Fig. 5). The orbital plate and a rim of hard palate lateral to septum was preserved as they were uninvolved and also to provide good prosthetic setting. The hard palate was reconstructed with gutta purcha, which was kept in position with stitches, and the cavity was packed with paraffin antibiotic gauze pack subsequently. Ryles tube was inserted for Enteral feeding. Skin sutures were removed on 5th post-operative day and palatal sutures were removed on the 10th postoperative day. Histopathology of excised specimen revealed a diagnosis of odontogenic myxoma (Fig. 6). The patient was referred to dentist for prosthesis and is doing excellent till date.

Discussion

The odontogenic myxoma is a benign, slow growing locally aggressive neoplasm of ectomesenchymal origin.

Fig. 2 Clinical photograph showing bulging of left maxilla with broadening and invasion of alveolus with reddish mass occupying left nostril

Fig. 3 CT scan showing a heterogeneous mass occupying left maxillary antrum and left a nasal cavity with erosion of anterior and medial wall of maxilla

Fig. 4 Per-operative photograph showing exposure of the mass involving the maxilla and left nasal cavity

Fig. 5 Photograph showing specimen removed after partial maxillectomy

This neoplasm occurs almost exclusively in the jaw bones and comprises 0.2% to 17.7% of odontogenic tumors [1]. It may present at any age but is most frequently discovered in the 2nd to 4th decades as in our case. It occurs more frequently in the mandible than in the maxilla with ratio of 3:1. The male to female ratio is 1:1.5. Maxillary odontogenic myxoma, as in our case is less frequent but behaves more aggressively than in the mandible as it spreads through

Fig. 6 Histopathological slide of odontogenic myxoma (10X10)

the maxillary sinus hence explaining the rapid recurrence as seen in our case. The neoplasm probably arises from the primitive mesenchymal structures of a developing tooth including the dental follicle, dental papilla, or periodontal ligament, and is therefore classified as an odontogenic tumor. Displacement of teeth by the tumor was a relatively common finding, but root resorption is rare [1, 2].

Differential diagnosis includes lesions showing typical multilocular radiolucency such as ameloblastoma, central hemangioma, aneurysmal bone cyst, central giant cell granuloma, giant cell lesions of hyperparathyroidism, cherubism, and metastatic tumors in the jaws. For that reason, a pathologist who is not familiar with the histology of a tooth germ can make a mistake very easily. Awareness of potential diagnostic pitfalls and careful evaluation of clinical and radiological data is necessary to narrow the differential diagnosis. Odontogenic myxoma can be invasive locally and has a high recurrence rate ranging from 10% to 33%. Fibrotic odontogenic myxoma is a rare unique variant which is harder and more common in recurrent cases as was seen in our case [1, 2].

The radiological descriptions of the lesion are varied and conflicting. According to various literatures, odontogenic myxoma may be multilocular or unilocular. Expansion of the cortex or distortion of the maxillary sinus wall is common finding. A characteristic feature of odontogenic myxoma on conventional radiographs is “tennis racquet” appearance [3]. Recently, CT and MRI features of this tumor have been demonstrated in many case reports. CT is effective in demonstrating the features of fibro-osseous lesion. It commonly demonstrates fine trabeculations especially in peripheral portion rather than the central portion of the tumor with no inter-lesion calcification. As many other fibro-osseous lesion have similar CT feature,

differential diagnosis based on CT findings is difficult. MRI, on the other hand is more specific in characterizing the tumor. On MRI, the tumor shows intermediate signal intensity on axial T1 weighted image and heterogeneous high signal intensity on T2 weighted image [3, 4].

On histopathology, the tumor consists of rounded, spindled, and stellate cells arranged in a loose, myxoid stroma with few collagen fibrils and delicate fibrous connective tissue as seen in our case. Immunohistochemistry studies suggest that the spindle-shaped cells constituting this neoplasm have a combined fibroblastic and smooth muscle typing, suggesting that it is of myofibroblastic origin. Odontogenic myxoma especially maxillary ones show little encapsulation and often extends into the surrounding soft tissue and intratrabecular space beyond the bony border, so that complete resection is difficult and recurrence is common [5].

Recommended therapy varies from curettage to radical excision. Curettage is associated with a high recurrence rate. Complete surgical removal can be difficult as the lesion is not encapsulated and because the myxomatous tissue infiltrates adjacent bone tissue. The treatment of choice is surgical excision ranging from segmental resection with clear bony margins of up 1.5 cm to prevent recurrence. Reconstruction can be immediate or delayed, and can include an autologous bone graft from the anterior or posterior iliac crest. Fibula free vascular osteocutaneous bone graft and distraction osteogenesis are other reconstructive modalities. Immediate postoperative follow up is weekly for approximately one month, then monthly for the next five months and twice a year for the next five years [6].

Conclusion

Fibrotic variety of Odontogenic myxoma of maxilla, a rare entity is sometimes very difficult to diagnose due to wide spectrum of nonspecific radiological features with no specific histological features on both FNAC as well as biopsy. Unlike other fibro osseous lesions, it is notorious for recurrence and cannot be managed simply by curettage or enucleation particularly in recurrent cases where treatment is often radical excision.

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