Aneurysmal bone cyst of the maxilla

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ABSTRACT

An aneurysmal bone cyst is a rare bone lesion. Its origin and precise nature remain unknown. It is seen as a locally-destructive, rapidly expandable, benign multicystic mass. We report a 17-year-old boy with an aneurysmal bone cyst of the maxilla, with extensive local involvement and bony destruction that was treated surgically. There was no recurrence noted after four years of follow-up.

Keywords: aneurysmal bone cyst, bone tumour, maxilla

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INTRODUCTION

An aneurysmal bone cyst (ABC) is an uncommon, osteolytic, benign, cystic solitary localised bone lesion, which expands rapidly and usually appears in the second decade of life. It is characterised by the replacement of the bone by spongy fibro-osseous tissue, and is a locally destructive and multicystic lesion filled with blood.[1] ABC may occur in any bone in the body. However, only 2% of the cases are found in the head and neck, and only 22 cases of ABC located in the maxilla have been reported.[1,2] The pathogenesis of ABC is still in doubt, and some authorities have suggested that its development is the result of a haemodynamic alteration or an arteriovenous malformation.[1] The diagnosis of ABC can only be confirmed by a histopathological examination. Clinical examination, preoperative radiological investigation and incisional biopsy are nonspecific and inconclusive.[3] We report a rare case of ABC of the maxilla with extensive local involvement and bony destruction that was treated surgically.

CASE REPORT

A 17-year-old boy presented with a swelling of the left cheek which increased in size for four months, and was associated with a left cheek dull pain, left nasal obstruction and loosening in his left upper teeth. The physical examination showed a 10-cm mass, which was cystic in nature, on the left side of his face, and a firm mass on the left buccal sulcus and hard palate. Direct endoscopy showed a polypoidal mass in the left nose arising from its lateral wall. Fine-needle aspiration cytology was performed and yielded a brownish haemorrhagic fluid; however, the cytological examination was inconclusive for a diagnosis.

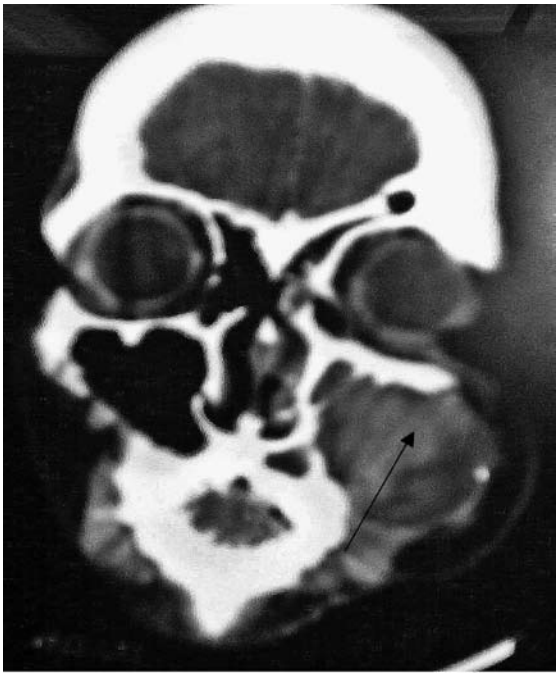

Fig. 1 Coronal CT image of the paranasal sinuses shows a large soft tissue mass within the left maxillary sinus (arrow) extending superiorly into the left ethmoidal sinuses and inferiorly into the ipsilateral superior alveolar process. Medially, the mass extends into the nasal cavity and is in close contact with the bony nasal septum.

Computed tomography (CT) of the paranasal sinuses showed a large soft tissue mass within the left maxillary sinus extending superiorly into the left ethmoidal sinuses and inferiorly into the ipsilateral superior alveolar process. Medially, the mass extended into the nasal cavity and was in close contact with the bony nasal septum. Laterally, it extended into the ipsilateral infratemporal fossa. There was an extension of the tumour in the subcutaneous region with multiple ring-enhancing lesions. The bony wall and superior alveolar process of the left maxillary sinus was eroded (Fig. 1). Angiography showed a high vascularity of the tumour with the main supply from the left internal maxillary artery. Preoperative embolisation of the left internal maxillary artery was done. He underwent left maxillectomy, ethmoidectomy and sphenoidotomy, where the tumour was fully excised macroscopically (Fig. 2). The surgery was uneventful.

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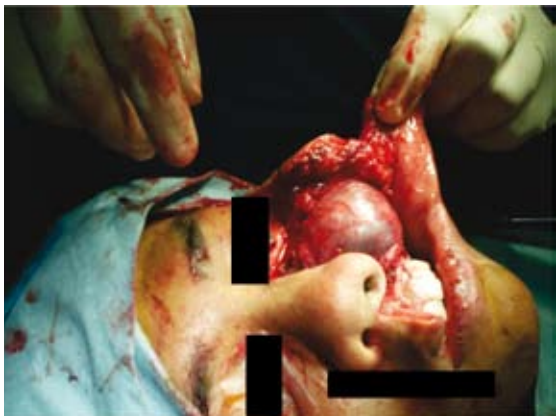

Fig. 2 Intraoperative photograph shows the macroscopic appearance of the tumour.

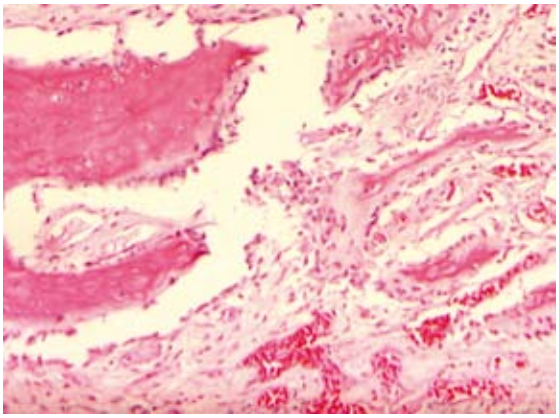

Fig. 3 Photomicrograph of the cyst wall shows the trabeculae of osteoid and immature bone against a background of congested fibrovascular connective tissues (Haematoxylin & eosin, × 100).

The histopathological examination revealed a severely haemorrhagic fibrovascular cyst wall without any neoplastic bony elements. In areas, trabeculae of reactive osteoid were present, as well as some haemosiderin-laden macrophages and scattered multinucleated giant cells. The features were consistent with an ABC (Fig. 3). He was well when discharged three days postoperatively. A repeat CT was done six months later and no recurrence was noted. Currently, there is no evidence of recurrence after four years of follow-up.

DISCUSSION

ABC is a rare bony lesion. Its origin and precise nature remain unknown.[4] It is a locally-destructive, rapidly-expanding, benign multicystic mass. Only 2% of ABCs are found in the head and neck, with 66% of these being located in the mandible.[3] From the literature review, only 22 cases of maxillary ABCs have been reported so far.[1,2] There is no gender or laterality preference of ABC of the maxilla. Generally, 80% of the cases are teenagers who present with complaints of a slight tender swelling mass on the face, with progressive enlargement within a week to three years.[1] Other clinical signs and symptoms basically depend on the extension of the lesion, e.g. nasal symptoms with extension nasally, eye symptoms when there is orbital extension, and recent mobility or migration of the teeth, as in our case, when the tooth bed is involved.

Preoperative diagnosis of the aneurysmal bone cyst is difficult due to its similarity to other lesions. The radiographical appearance is not characteristic; it may only be suggestive but not diagnostic. CT often shows an osteolytic and expandable multinuclear mass with surrounding bone shell and displaced surrounding normal structures. The classic blow-out appearance has been described. A clearly-defined body wall (bony septa) imparting a soap bubble or honeycomb appearance has been reported.[5] There is no obvious fluid level noted on the CT of our patient.

Fine-needle aspiration cytology and incisional biopsy are both unable to conclude the definite tissue diagnosis of the mass preoperatively. Aspiration of a dark red or brownish haemorrhage fluid favours the diagnosis of ABC. Bataineh did not report any cases being diagnosed from an incisional biopsy. The definitive diagnosis was made only after macroscopical examination of the surgical specimen in his study.[4] It is important to examine the whole tissue to exclude an associated additional pathological process. Histologically, the ABC is characterised by large blood-filled spaces which do not have an endothelial lining. Instead, the cyst wall and septa are made up of fibroblasts, myofibroblasts, histiocytes, congested vessels, osteoblasts, osteoid, bone and degenerated calcifying fibromyxoid tissue.[6] The cyst wall also shows these findings.

ABC is associated with other entities, including giant cell tumour, giant cell reparative granuloma, infant hamartoma, fibrous dysplasia, osteosarcoma, haemangioma and telangiectatic osteosarcoma in 32%–50% of the cases.[7] Surgery is the main treatment of ABC of the maxilla. However, due to the vascularity of the mass, preoperative angiographical examination with embolisation of the feeding vessels as close to the lesion as possible, has been advocated. This will reduce the complication of excessive bleeding during the surgery. The principle of surgery is to fully excise the tumour. Curettage with enucleation is the first choice of treatment and depends on the extent of the extension of the tumour locally. Irradiation of the ABC is not advisable due to a high risk of sarcomatous change.[2]

ABC has a high recurrence rate, which varies from 26% to 56% in most cases, within the first year after treatment.[1] Recurrence seems to occur most frequently within the first year of the initial treatment. It is usually due to incomplete or inadequate removal of the tumour, which may in turn be due to the underexposure of the surgical field or a difficulty in accessing the extension of the tumour. Therefore, regular follow-up of the patient is recommended, with periodic imaging guidance until complete osseous repair and remodelling of the affected area have taken place.

In conclusion, an ABC of the maxilla is a rare bony lesion. Clinical examination, radiological investigation, fine-needle aspiration cytology and incisional biopsy preoperatively are crucial for the diagnosis, before the planning for definite treatment. Adequate exposure of the surgical field with the aim of total excision of the tumour is the main treatment of ABC of the maxilla. Preoperative angiography with embolisation of the feeding vessels may reduce the haemorrhage intraoperatively. A careful histopathological examination of all the fragments to determine the existence of associated pathological findings, is necessary. Regular follow-up with image guidance is important to detect the recurrence of the disease.

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