AMELOBLASTOMA OF THE MANDIBLE

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INTRODUCTION

Ameloblastoma (previously called Adamantinoma) is an uncommon locally aggressive tumour arising from odontogenic epithelium consisting of cells called ameloblasts forming the inner most layer of enamel organ [1]. Ameloblastoma histologically resembles the embryonic enamel organ but does not differentiate to the point of forming dental hard tissue. It has a high tendency to recur and may metastasize or even undergo malignant transformation. It accounts for only one percent of all oral tumours and behaves as a slowly growing expansile radiolucent tumour, and occurs most commonly in the mandible (80 %), especially at the mandibular angle in molar-ramus area [2].

CASE REPORT

A 29 years old male patient reported in the surgical OPD, PAF Hospital Mianwali in Jan 2005 with history of gradually increasing painless intra-oral lump, 3×4 cm in size, with considerable facial deformity for the last 10-12 months. It involved gums, adjacent buccal mucosa, and mandible. It was foul smelling, friable, fungating mass present in left side of buccal cavity. Left lower molars and premolars were mobile. Tongue was free of growth. Overlying skin was intact. There was no lymphadenopathy and clinical examination of chest and abdomen was unremarkable. X-ray chest, Blood complete picture, urinalysis, and abdominal ultrasound revealed no abnormality. X- ray of the mandible showed multiple radiolucent loculations with soap bubble appearance involving body of mandible. There was marginal sclerosis and erosion of roots of involved teeth. OPG revealed osteolytic lesion involving body of left side of mandible. Incision biopsy of the growth revealed ameloblastoma. He was prepared for surgery giving particular attention to his oral hygiene. Segmental mandibulectomy was carried out by lower lip splitting incision extended up to the angle of mandible. Whole flap was lifted up thus completely exposing left side of mandible. The entire tumour with margin of healthy tissue was excised including the part of mandible bearing the tumour and healthy margin of bone. The healthy rim of mandible was left behind to maintain the continuity. Flap was stitched back (Fig.1,2). Post-operative results were excellent both functionally and cosmetically. Follow-up for the last 11 months has not revealed any sign of recurrence.

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DISCUSSION

This tumour was described for the first time in 1827 by Cassock and then in 1879 Falkson gave first complete histological description of the tumour. Mallasez in 1885 introduced the term adamantinoma, which was later on changed to Ameloblastoma in 1934 by Churchill [1].

Ameloblastoma accounts for one percent of all oral tumours. The mandible is affected in about 80 % of cases, four times more frequently than the maxilla. Rarely it can occur simultaneously in mandible and maxilla1 and even maxilla, skull and groins successively on both sides [2]. Rarely they can pose diagnostic difficulties when they metastasise in kidneys, myocardium, lungs and in long bones as pathological fractures. Both sexes are affected equally. Although the tumour can occur at any age and has been described in the literature in patients as young as 21 months, most patients present in the third or fourth decade of life with the

average age being 27 years. The remaining twenty percent occur in the maxilla with the maxillary tuberosity being the most common site.

Histologically, the ameloblastoma is a neoplasm of odontogenic epithelium with a fibrous background typically arising from tissue resembling the embryonic enamel organ (e.g., ameloblasts). The tumour consists of palisading columnar epithelial cells in the periphery of ameloblastic layer. The nuclei are polarized. There is mature connective tissue in stroma and loosely arranged stellate reticulum inside islands. There is no capsule. These bone tumours are histologically benign but locally destructive, typified by extensive invasion of cancellous bone and occasional cortical disruption with soft tissue extension. An enormous number of histologic variants of ameloblastoma have been described: follicular, pigmented, acanthomatous, plexiform, granular cell, stellate, basaloid, ameloblastic fibroma, and adenoameloblastoma. The histologic pattern of these tumours, however, does not reliably predict their clinical course or serve as a guide to therapeutic decision making. Morphologically, ameloblastomas are classified into four groups; unicystic, solid or multicystic, peripheral, and malignant. Unicystic ameloblastomas are less aggressive than solid ameloblastomas and there is an intraluminal or an intramural proliferation of the cystic lining. Multicystic ameloblastoma has a poorer prognosis than the unicystic lesion due to excessive local infiltration. The peripheral ameloblastoma is the soft tissue version of ameloblastoma. It occurs in the alveolar mucosa and can involve underlying bone secondarily. The malignant ameloblastoma is again rare and it can metastasise to lungs [3], kidneys [4], myocardium [5] and to the long bones [6]. These four subtypes of ameloblastoma are dependent on ameloblastin gene (AMBN), which produce ameloblastin protein in organic matrix of enamel [7]. The matrix metalloproteinases (MMPs) is considered to be an important factor in the local invasiveness of this tumour [8] while Parathyroid Hormone Releasing Protein (PTHrP) plays a significant role in local bone resorption and destructive behaviour. It has significant therapeutic implications, particularly through PTHrP-blocking treatment modalities [9].

Clinically these tumours present with a slowly growing, asymptomatic intraoral swelling usually with facial deformity. Early symptoms are absent in 75% of patients and these tumours are seldom diagnosed in the early stages of development. Pain, numbness, toothache, loose teeth, ill-fitting dentures, malocclusion, ulcerations, draining sinuses, nasal obstruction, or even epistaxis may bring the patient to seek medical advice. Pathological fracture is a very unusual finding on presentation. They are often found incidentally with the use of routine x-rays. Plain radiography, panoramic radiographs (OPG), conventional tomography (CT's), and magnetic resonance imaging (MRI's) are all used as diagnostic aids. Findings may include expansion of cortical plate with scalloped margins, multiloculations or "soap bubble" appearance, and/or root resorption. Due to slow growing tumour margins are usually well defined. MRI's, even though not useful for hard tissue examination, are used to provide information regarding edge definition and tumour consistency. Diagnosis is confirmed by biopsy. The differential diagnosis of a multilocular radiolucency in the jaw includes cherubism, giant cell granuloma, odontogenic myxoma, aneurysmal bone cysts and odontogenic keratocysts, cemento-osseous dysplasia, central giant cell granuloma, central arteriovenous malformation and calcifying epithelial odontogenic tumour (Pindborg Tumour).

TREATMENT

Treatment ranges from conservative therapy to more radical procedures. Conservative therapy includes radiotherapy,

curettage, and enucleation. Radical surgery as defined by Muller and Slootweg is a procedure in which the ameloblastoma is removed with a margin of normal bone [10]. This wide excision is followed by reconstruction of mandible usually by bone grafting. Most investigators believe in resecting at least 1 cm. of normal bone beyond the tumour margin. This means segmental or even hemimandibulectomy in selected cases. Soft tissue borders at the time of resection may also be confirmed by frozen sections to ensure complete tumour removal. Unicystic Ameloblastoma can be treated by enucleation alone. Ameloblastoma has a high recurrence rate if not adequately removed, but local recurrence may occur even in patients who have undergone satisfactory primary surgical treatment. As these tumours recur they become more aggressive [11] and can develop into a lesion that is more aggressive than a sarcoma.

An ameloblastoma is an epithelial tumour similar to a basal cell carcinoma histologically. Therefore, some investigators contend that their radio sensitivities must also be similar. However, radiation therapy is rarely used as a primary treatment. Instead it is used in conjunction with surgery in the management of selected patients with recurrence. Surgery should be followed by radiotherapy in following instances. (1) Mandibular recurrences when the first surgical treatment was adequate, (2) for all recurrences and (3) when soft tissue involvement or positive surgical margins are present after a wide resection. Palliative irradiation has a role in inoperable cases but the role of chemotherapy is not yet established. There have also been reports of sarcomas induced when treating ameloblastomas with radiation.

It is important to note that this case was successfully managed in a peripheral hospital. Ideally such patients should be managed in specialized centres. The patient was a non-entitled poor patient who could not afford expenses of going to another city for his treatment. He was ultimately managed locally as a gratis case.

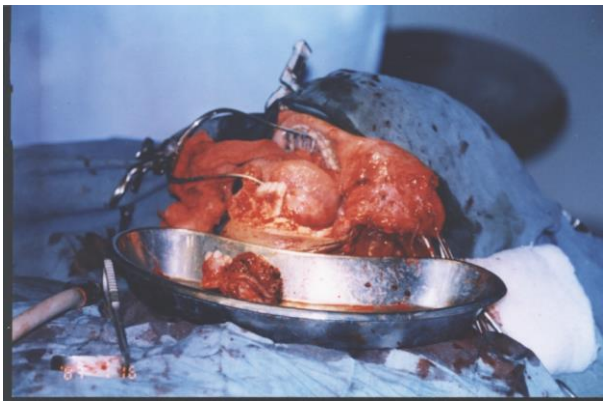

Fig. 1: Segmental mandibulectomy showing extent of mandibular excision.

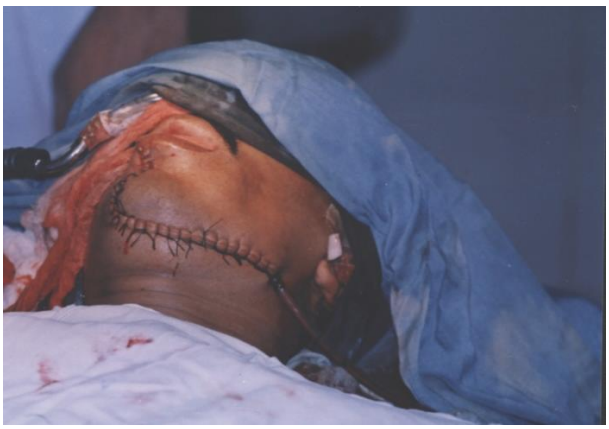

Fig. 2: Closure of skin flap with a drain in position.

CONCLUSION

Ameloblastoma is a rare intra-oral tumour, which is locally invasive and has a high tendency to recur, metastasise and even undergo malignant transformation. They can pose diagnostic difficulties when they metastasise in kidneys, myocardium, lungs and in long bones as pathological fractures. General surgeon working in a peripheral hospital in a military set-up can come across such cases. It is important that the diagnosis is made in time and a well-planned surgical excision is done. Adequate resection is mandatory as the recurrent tumours behave more aggressively. These tumours can recur even after satisfactory primary resection and a life long follow up is required for early detection of recurrence.

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