Case Report

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Classical Gigantiform Ameloblastoma

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Abstract

The ameloblastoma is most common benign odontogenic tumour of ectodermal origin. It represents 1% of all tumours of the jaw. The most common site of occurrence is mandible (75%) involving premolars, molars and angle region. They can also involve maxilla (25%). Ameloblastoma usually occurs in the 4th and 5th decades of life having no sex predilection. It often presents as a slow growing, painless swelling and causing expansion of the cortical bone. It has tendency for local aggressiveness. Ameloblastoma is treated by curettage, enucleation, cryosurgery, marginal, segmental and composite resections depending on the type and nature of ameloblastoma. Recurrence is frequent after inappropriate surgery. Accurate preoperative delineation of the boundaries of the tumour is essential for achieving a complete resection. This case report emphasizes on gigantiform ameloblastoma which resembles ameloblastoma but having more aggressive spread and high morbidity and mortality rate.

Keywords: Odontogenic Tumors; Mandibular Tumors; Ameloblastoma.

Introduction

Gigantiform ameloblastoma are usually benign locally aggressive neoplasm derived from the epithelial odontogenic tissues which are part of the tooth-forming apparatus. They account for about 1-3% of tumours of the jaws. The tumour is more common in the mandible than in the maxilla and shows predilection for various parts of the mandible. It often presents as a slow growing painless swelling and causing expansion of the cortical bone. In its early stages it is confined to the bone but large tumours can break through the cortex and involve the soft tissues [1]. The biologic behaviour cannot be predicted on the basis of morphology and there is often a delay in the diagnosis also because of its slowgrowing nature. Ameloblastoma can develop at any age but its peak incidence is reported in third and fourth decades of life and its incidence is equal in both sexes [2]. The term ameloblastoma was coined by Ivy and Churchill in 1934. Guzack and Broca recorded similar lesions in the literature in 1826 in 1868 respectively. However it was Fallson who describe ameloblastoma thoroughly in 1879. The same type of lesion was described by Malassez in 1885 who called it an adamantinoma. The term giant ameloblastoma should be reserved for lesions that are truly large and cause gross asymmetry and regional dysfunction [2].

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Case Report

A 55 year old patient (Fig.1) reported with presenting complaint of a massive swelling involving anterior region of mandible since 2 years. The swelling was previously small in size which was progressively increasing in size to present form. It was causing significant facial asymmetry and difficulty in chewing food. There was no significant past medical and dental history. The patient had normal gait and posture. He had normal intelligence and well oriented to surroundings. The patient was malnourished and poorly built and generalized pallor noted. The patient had difficulty in eating due to massive nature of lesion (Fig.2a & Fig.2b). On extra-oral examination a massive asymmetrical swelling

of 6x6 cm noted in mandibular anterior region extending till body of mandible in both sides superiorly (Fig.1). The inferior extension of swelling was conical in shape and obliterating the neck anteriorily. It was causing fullness of lower lip and obliteration of mentolabial sulcus (Fig.3). The swelling was non-tender, firm to hard in consistency. There was no sign of fluctuation, reducibility and compressibility noted. The overlying skin over swelling was normal in color and texture. There was no local rise in temperature and no associated pain. On intra-oral examination (Fig.4a) a massive swelling was noted which extending from right mandibular 2nd molar to was left mandibular 3rd molar. There was severe cortical expansion involving both the buccal and lingual cortical plates of mandible. The lingual expansion in left side is very significant causing obliteration of lingual sulcus and elevation of floor of mouth which interferes with tongue movements. In anterior region of mandible the labial cortical expansion was quite massive causing obliteration of labial sulcus (Fig.4b). Intra-orally the swelling was fluctuant in labial sulcus area due to complete cortical degeneration. Many veins were visible on the surface of swelling. It was firm to hard in consistency and was non reducible and non compressible. There was no associated pain. There was no history of bleeding noted. Some teeth were mobile in both sides. The swelling was aspirated with 18 gauze needle from anterior labial sulcus area where it was fluctuant. On aspiration a yellow straw color fluid was obtained (Fig.5 & Fig.6). The patient was further advised for radiological investigation by occlusal radiograph, panoramic radiograph and lateral cephalogram. The occlusal radiograph (Fig.7) shows multilocular radiolucecies with bilateral cortical expansion with massive destruction. The teeth were floating in radiolucencies and have pathological migrations. The panoramic radiographs (Fig.8) show multilocular radiolucencies extending from mesial aspect of right 3rd molar to mesial aspect of left mandibular 2nd molar causing massive destruction of whole length of body of mandible bilaterally. The teeth are pathologically migrated and have root resorption and appear like floating in radiolucencies. The lateral cephalogram (Fig.9) shows the massive bilateral destruction of mandible with floating teeth and root resorption. On the basis of clinic-radiologic findings a provisional diagnosis of ameloblstoma has been made. The patient was advised for incisional biopsy of the lesion. The routine blood investigation has carried out which shows all the values were within normal limit. The incisional biopsy is carried out under local anesthesia (Fig.10a & Fig.10b). The histopathology (Fig.11) showed islands of epithelium that resemble enamel organ in a fibrous connective tissue stroma attached to the basement membrane surrounding the islands are tall columnar cells exhibiting reversed polarity. This is consistent with the diagnosis of a multicystic, follicular ameloblastoma. On the basis of clinical, radiological and histological findings a final

Fig.1: Profile picture of patient showing a massive asymmetrical swelling of 6x6 cm noted in mandibular anterior region extending till body of mandible in both sides superiorly

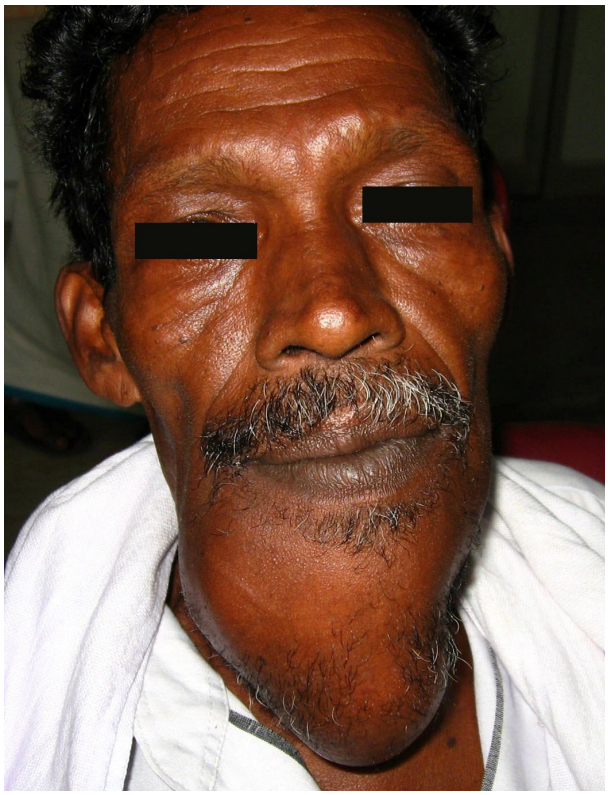


Fig.2a: Left lateral view of patient showing the massive swelling of mandible involving anterior region. The inferior extension of swelling was conical in shape and obliterating the neck anteriorly.

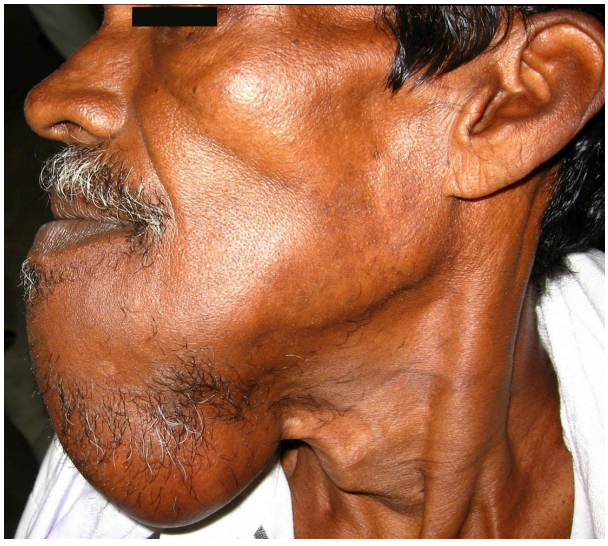


Fig.2b: Right lateral view of patient showing the massive swelling of mandible

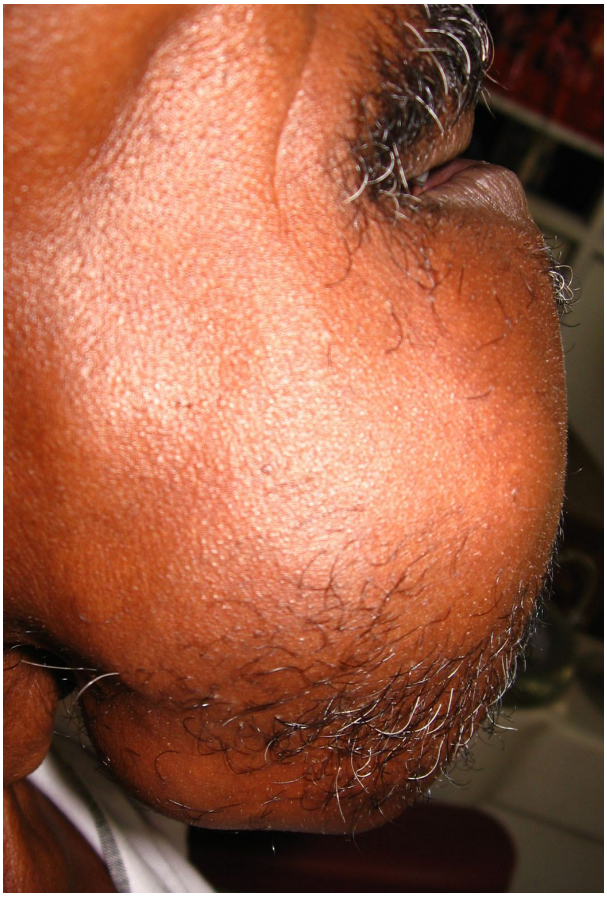


Fig.3: Anterior view of swelling causing fullness of lower lip and obliteration of mentolabial sulcus.

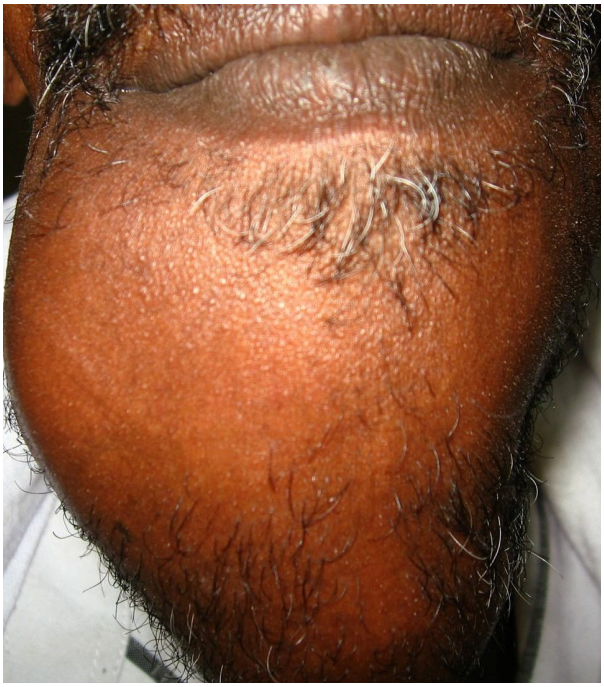


Fig.4a: Intraoral view showing a massive swelling which extends from right mandibular 2nd molar to was left mandibular 3rd molar. The lingual expansion in left side is very significant causing obliteration of lingual sulcus and elevation of floor of mouth.

Fig.4b: Intraoral view showing massive labial cortical expansion in anterior region of mandible causing obliteration of labial sulcus.

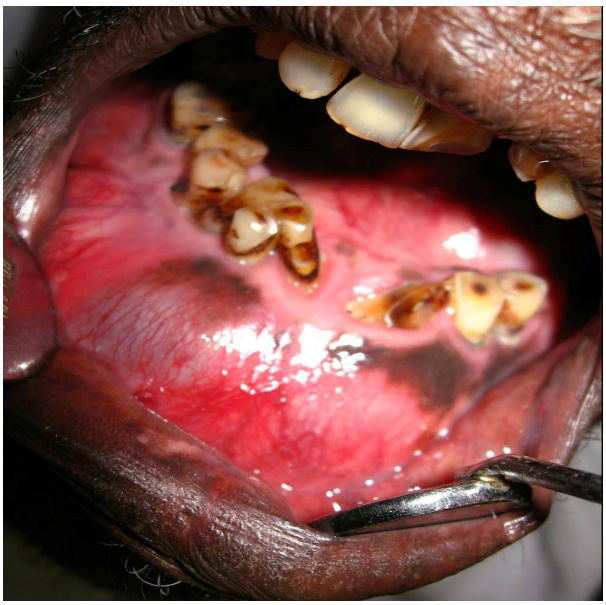


Fig.5: Photograph showing aspiration of swelling with 18 gauge needle with a yellow straw color fluid in syringe.

Fig.6: Yellow straw color fluid in syringe after aspiration.

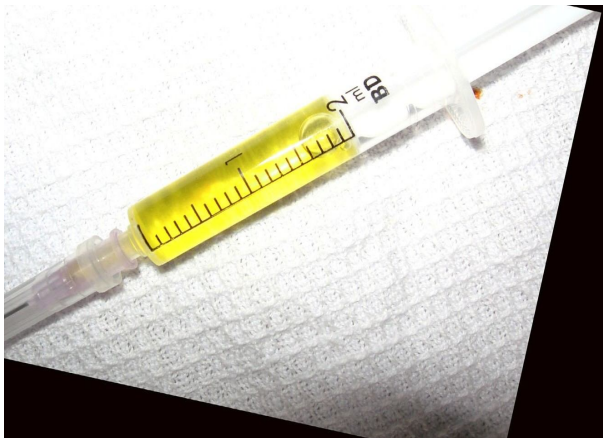


Fig.7: The occlusal radiograph showing multilocular radiolucecies with bilateral cortical expansion with massive destruction. The teeth were floating in radiolucencies and have pathological migrations.

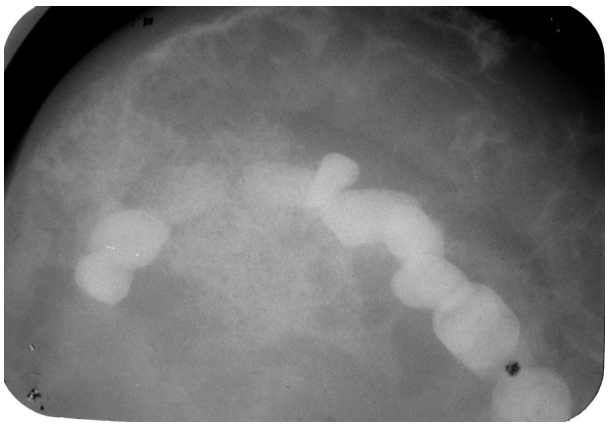


Fig.8: The panoramic radiographs showing multilocular radiolucencies extending from mesial aspect of right 3rd molar to mesial aspect of left mandibular 2nd molar causing massive destruction of whole length of body of mandible bilaterally.

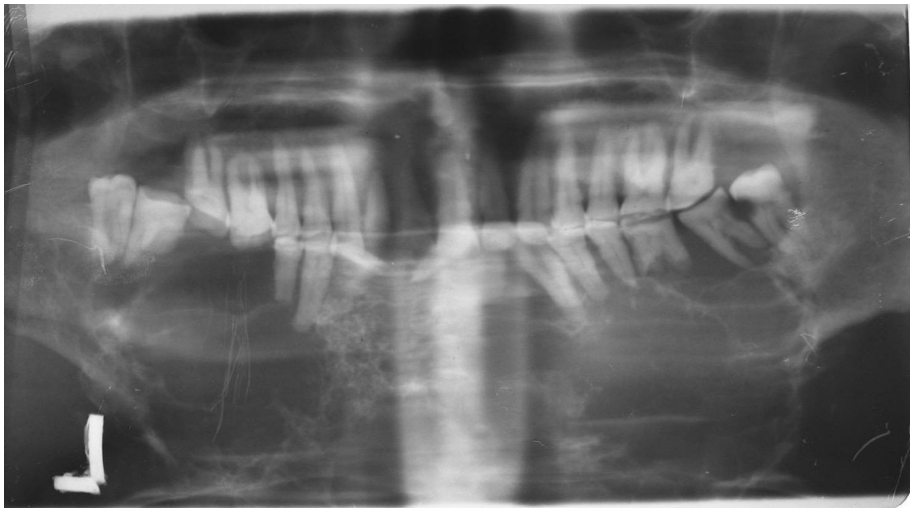


Fig.9: The lateral cephalogram showing the massive bilateral destruction of mandible with floating teeth and root resorption.

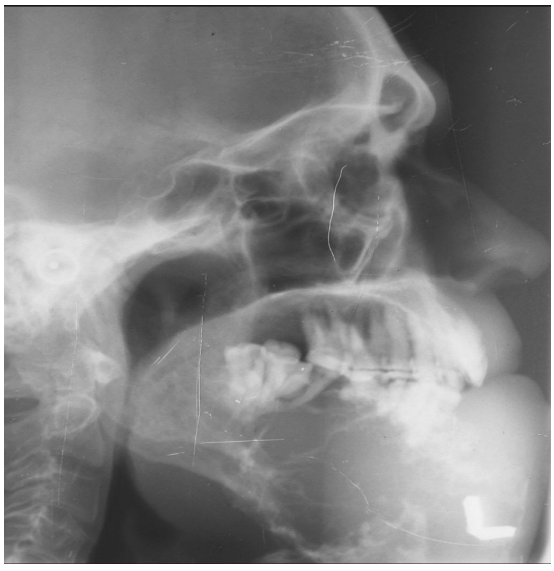


Fig.10a: Photograph showing biopsy procedure.

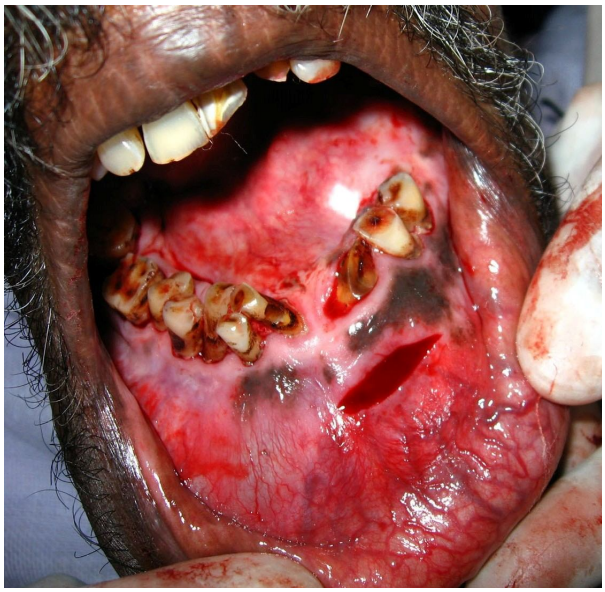


Fig. 10b: Photograph showing sutured biopsied wound

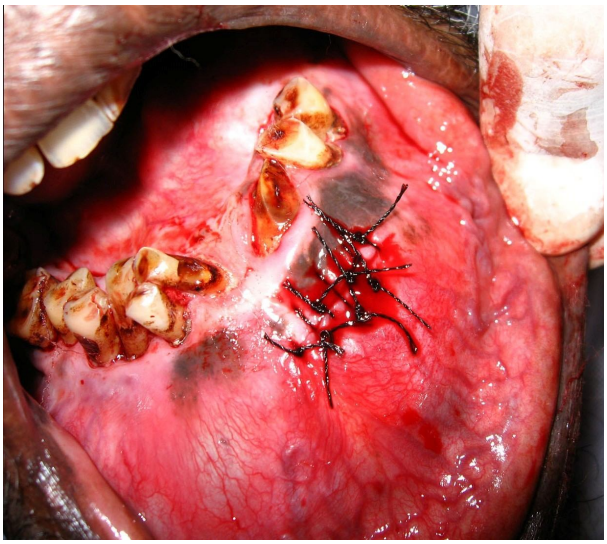
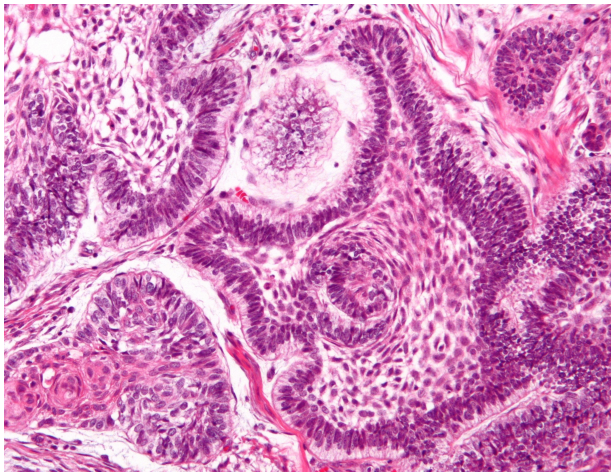


Fig.11: Photomicrograph showed islands of epithelium that resemble enamel organ in a fibrous connective tissue stroma attached to the basement membrane surrounding the islands are tall columnar cells exhibiting reversed polarity.(400x H&E)



diagnosis of ameloblastoma has been reached. The patient was referred to oral surgery where tumour surgical removed by en block resection under general anaesthesia. The patient was followed for 1 year. Till date no recurrence has been reported.

Discussion

Although giant ameloblastoma is rare in developed countries, it can occur in patients who delay treatment because of fear of surgical intervention. It is hypothesized that protein leakage occurs in the cyst that eventually causes the gigantiform ameloblastoma [3]. Our patient was also malnourished because the size of the lesion which impaired his ability to eat. The aetiology of ameloblastoma is unknown. The incidence of ameloblastoma is estimated at 0.5 per million population per year although in some parts in the world, e.g. South Africa a higher incidence has been reported [4, 5]. The molecular and genetic characteristics of ameloblastomas are poorly understood and the origin of the tumour is still unclear. The cloning and characterization of expression of the ameloblastin and amelogenin genes in these tumours suggests that ameloblastoma arise from the odontogenic apparatus or cells that are potentially capable of forming dental tissue. The potential sources for this tumor are the cell rests of the enamel organ (cell rests of Malassez and cell rests of Serre), epithelial odontogenic cysts (dentigerous cysts), basal cells of the surface epithelium of the jaws and heterotrophic epithelium in other parts of the body [6].Clinically the tumour often presents as an asymptomatic swelling of the posterior mandible frequently being associated with an unerupted tooth [7]. Most patients are aged between 30 and 60 years at the time of diagnosis. There is no gender predilection. Multiple presentations are exceedingly rare. It can be expressed as an aggressive odontogenic tumour. It can sometimes cause symptoms such as swelling, dental malocclusion, pain and paresthesia of the affected areas.It spreads by forming pseudopods in marrow spaces without concomitant resorption of the trabecular bone. As a result the margins of the tumour are not clearly seen on radiographs or during surgery and the tumour frequently recurs after inadequate surgical removal[8,9] The appearance of septae on the radiograph usually represents differential resorption of the cortical plate by the tumour and not actual separation of tumour portions [10]. Because of its slow growth, recurrences of ameloblastoma generally present many years and even decades after primary surgery [9]. Malignant transformation can occur in inadequately treated cases [11]. In most cases ameloblastoma has a characteristic but not diagnostic radiographic appearance [8]. The neoplasm usually appears as a unilocular radiolucent area or a multilocular radiolucent area with a honeycomb appearance [8, 11]. Resorption of the adjacent tooth roots is not uncommon [8]. In many cases an unerupted tooth, most often a mandibular third molar is associated with the tumour [11].

Ameloblastoma is divided into 3 clinico-radiologic groups: solid or multicystic, unicystic and peripheral. The solid ameloblastoma is the most common form of the lesion (86%). It has a tendency to be more aggressive than the other types and has a higher incidence of recurrence [12, 13]. Unicystic

ameloblastoma has a large cystic cavity with luminal, intraluminal or mural proliferation of ameloblastic cells. It is a less aggressive variant and it has a low rate of recurrence [13, 14, 15]. The ameloblastoma is a histologically almost always benign odontogenic tumour of the jaw bones. The main tumour cells are columnar resembling preameloblasts of the enamel organ and show reversal of polarity with peripheral palisading. The epithelial components surround a central web-like arrangement of spindle-shaped cells resembling stellate reticulum. However it has a strong tendency to recur after conservative surgical removal [5]. In the 2005 World Health Organization classified the benign ameloblastoma is into solid / multicystic, extra-osseous / peripheral, desmoplastic and unicystic. The solid / multicystic ameloblastoma can histopathologically be divided into a follicular and a plexiform type; the follicular type can be further subdivided into a spindle cell type, an acanthomatous type, a granular type and a basal cell type. The plexiform type contains basal cells arranged in anastomosing strands with an inconspicuous stellate reticulum. The stroma is usually delicate with cyst like degeneration. The unicystic ameloblastoma represents an ameloblastoma variant that on gross examination presents as a cyst. Two histopathological variants are recognized, the luminal variant and the mural variant. The extraosseous type shows the histopathogical cell types and pattern resembles as seen in the solid / multicystic type. In the desmoplastic type the stromal component dominates compressing the odontogenic epithelial components [12].

Lesions Mimicking Ameloblastoma

There are a number of lesions that can be easily misdiagnosed as ameloblastoma. Their proper diagnosis is mandatory because of significant differences in the clinical and biological behaviour of these tumours.

Ameloblastic carcinoma

Ameloblastic carcinoma is a rare malignant odontogenic tumour that shows clear cytological features of malignancy [16]. It occurs at a later age than ameloblastoma and it is locally more aggressive [17]. It is more common in the mandible and shows a high propensity to recur. Histologic examination reveals malignant features that include cellular pleomorphism, nuclear hyperchromatism and high mitotic activity. Some histologic features reminiscent of conventional ameloblastoma, such as nuclear palisading, reverse polarization etc., are also present. About 22% of ameloblastic carcinomas produce metastases having identical histopathological features as those of the malignant primary tumour.

Ameloblastic fibroma

Ameloblastic fibroma is a rare odontogenic tumour that is usually diagnosed in the first decade of life as asymptomatic slowly growing tumour [19]. The posterior mandible is the most common site [20]. Radiographically, this lesion is either unilocular or multilocular radiolucencies associated with an unerupted tooth [21]. This tumour can be misdiagnosed as follicular ameloblastoma. However follicular ameloblastoma lacks the stromal hypercellularity of spindle cells that are characteristic of ameloblastic fibroma. Conservative surgical excision and regular follow-ups are indicated for primary lesions [22, 23].

Ameloblastic fibro-odontoma

Ameloblastic fibro-odontoma is often diagnosed during the first and second decades of life. Ameloblastic fibro-odontoma is usually asymptomatic. It either delays the tooth eruption or may displace adjacent teeth. Ameloblastic fibro-odontoma represents an ameloblastic fibroma with varying amounts of soft and calcified dental tissues [24].

Ameloblastic fibrosarcoma

Ameloblastic fibrosarcoma is a very rare odontogenic malignancy in which the connective tissue shows microscopic features reminiscent of fibrosarcoma [25, 26]. Some of these cases arise de novo but about half of them may arise from a preexisting ameloblastic fibroma [26]. Ameloblastic fibrosarcoma is a locally aggressive tumour and radical resection is indicated. Metastases are not unusual.

Treatment of mandibular ameloblastoma continues to be controversial. It can vary with clinicoradiologic variant, anatomic location, clinical behaviour, age and the general health.

There has been long debate regarding the most appropriate modality of treatment for ameloblastoma. These range from conservative to radical modes of treatment. The conservative modalities include curettage, enucleation and cryosurgery while the radical modalities include marginal, segmental and composite resections. However there is a lack of consensus over the most appropriate treatment modality. The supporters of conservative approach believe that ameloblastoma though a locally invasive tumour is essentially benign in nature so it should be treated as such [27, 28]. Ueno et al suggested that

'excessive resection' of the mandible constituted excessive treatment [27], and Feinberg and Steinberg concluded excessive resection is compatible in young patients [28]. Sammartino et al also advocated for conservative treatment of giant ameloblastoma due to associated low morbidity. However a recurrence rate of 55% to 90% has reported in tumours treated by enucleation or curettage [29].

Conclusion

Ameloblastoma are common odontogenic tumours. They present challenge in both diagnosis and treatment because of diversities in clinico-pathological and radiographic features. This case report emphasized on gigantiform ameloblastoma which resembles ameloblastoma but having more aggressive spread and high morbidity and mortality rate.

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