CASE REPORT

An Unusual Case of Capacious Ameloblastic Carcinoma

Keerthi M[1], Shyam Prasad Reddy D[2], Madhusudan Rao T[3], Ramanand P[4]

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[1,2,3]Senior Lecturer
[4]Post Graduate
Department of Oral & Maxillofacial Pathology,
Kamineni Institute of Dental Sciences,
Sreepuram, Narketpally, Nalgonda Dist. A.P.

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Email for correspondence:
keerthikrishnamds@yahoo.com

ABSTRACT:

Ameloblastic carcinoma, a rare malignant lesion with characteristic histologic features and behaviour, dictate a more radical surgical approach than does a simple ameloblastoma. Clinically, it is more aggressive than most typical ameloblastomas with extensive local destruction, perforation of the cortical plate, extending into surrounding soft tissues, numerous recurrent lesions, and metastasis usually to cervical lymph nodes. Alternatively Ameloblastic carcinoma represents a malignant tumour that bears a histologic resemblance to an ameloblastoma. Here we present a case report of ameloblastic carcinoma in a 69year old male patient where a longstanding ameloblastoma of the mandible has turned into carcinoma.

Key words: Ameloblastic carcinoma, malignant odontogenic tumour.

INTRODUCTION

Ameloblastoma is locally invasive benign tumour of odontogenic origin with a high chance of local recurrence. It will therefore become progressively destructive if left untreated. The invasive ameloblastoma that occur centrally in either jaw will usually present as an asymptomatic expansion. Malignant ameloblastoma distinguishes from ameloblastoma by the presence of metastases.[1] It shows the histopathological features of ameloblastoma, both in the primary tumour and in the metastatic deposits. Whereas the ameloblastic carcinoma has cytologic features of malignancy not only in a primary tumour but also in a recurrence or in any metastatic deposits. These lesions are locally aggressive, but metastasis does not necessarily occur.

Ameloblastic carcinomas seem to be more common compared to malignant ameloblastoma in the ratio 2:1. Ameloblastic carcinomas have been reported to metastasize to the lungs and to distant sites. Male to female ratio is almost 1.4:1. Lesions are located in the mandible and maxilla in 66% and 34% of the patients, respectively.[2]

Case report:

A 69-year old male patient presented to the Kamineni Institute of Dental Sciences with a chief complaint of swelling on the right side of the face for the past 7years and he had difficulty in normal functioning of the oral cavity. As the swelling increased in size, the teeth in the associated region became mobile and exfoliated. There was no history of pain, ulceration or discharge associated with the swelling. The patient's history revealed that swelling was initially noticed as a small nodule, which gradually increased to the present size. The patient's medical history and family history was non-contributory.

Extra orally a single nodular swelling measuring about 23 x 13.4cms in size was present, roughly oval in shape. The swelling extended anteriorly from left side of the corner of the mouth to the middle of the right side of the mandible posteriorly. Superiorly it extended to the level of line joining the corner of the mouth to tragus of the ear and inferiorly extended up to 2cms below the chin. The skin over the swelling was normal. The chin was slightly deviated to the right side (Figure 1). On palpation, the swelling was fibrous hard in consistency, non tender, non fluctuant. The regional lymphnodes were not palpable.

Intraoral examination revealed a single nodular growth extending from mandibular left lateral incisor to mandibular right 3rd molar. The growth enclosed the entire tooth surfaces in the right side of the mandible. In right mandibular anterior tooth area the colour of the growth was reddish when compared to the rest (Figure 2).

The patient was advised CT scan where it revealed an extensive osteolytic lesion involving both bucco-lingual cortical plates with complete perforation of the cortices and infiltration of the tumor mass in to the surrounding soft tissue (Figure 3). Based on the radiographic investigations a provisional diagnosis of ameloblastoma / carcinoma of the mandible were made. An incisional biopsy was made and was sent for histopathological examination.

The histopathology revealed numerous epithelial follicles spread out in a scanty connective tissue stroma (Figure 4). The epithelial nests showed typical peripheral tall columnar cells with apically placed nuclei and vacuolated cytoplasm. The central cells showed stellate reticulum like appearance with some cells showing squamous metaplasia and numerous keratin pearls (Figure 5). A few cells showed features of dysplasia such as cellular and nuclear pleomorphism with nuclear hyperchromatism (Figure 5). Abnormal mitotic figures were also evident. Based on the above histological features a diagnosis of Ameloblastic Carcinoma was made.

Discussion:

Ameloblastic carcinoma is a rare malignant lesion with characteristic histological features and behaviour that dictate a more radical surgical approach than does a simple ameloblastoma. Clinically, ameloblastic carcinoma is more aggressive than most typical ameloblastomas with extensive local destruction, perforation of the cortical plate, extension into surrounding soft tissues, numerous recurrent lesions, and metastasis, usually to cervical lymph nodes.[2]

Naagai divided the malignant tumours with features of ameloblastoma into two groups: 1) malignant ameloblastoma 2) ameloblastic carcinoma. Malignant amelobastoma is an ameloblastoma with a benign and typical histological pattern which metastasize. Ameloblastic carcinoma is any ameloblastoma in which there is primary or recurrent evidence of malignancy regardless of metastasis.[3]

Ameloblastic carcinoma is defined as a malignant epithelial odontogenic tumour that histologically has retained the features of ameloblastic differentiation yet also exhibits cytological features of malignancy.[1] The term ameloblastic carcinoma was introduced by Shafer. Corio et al in 1987 reported 8 cases of ameloblastic carcinoma and redefined the term ameloblastic

carcinoma to refer to any ameloblastoma in which there is histologic evidence of malignancy in the primary or the recurrent tumour regardless of whether it has metastasized.[4]

Ameloblastic carcinoma mainly affects elderly individuals, but the age range is 15 to 84 years. The male:female ratio is 1.4:1. Approximately 2/3 of Ameloblastic Carcinoma involves the mandible.[5] The most commonly involved area is the posterior portion of the mandible. Involvement of the maxilla by ameloblastic carcinoma seems to be less frequent than that of the mandible. Common clinical signs and symptoms of ameloblastic carcinoma include swelling, pain, trismus and dysphonia. Rapid tumor growth and nerve paresthesia may also occur.[6]

The pathogenesis of ameloblastic carcinomas is not clear. They may originate ex ameloblastoma or ex odontogenic cyst, and both central (intraosseous) and peripheral (extraosseous) variants have been described. An ameloblastic carcinoma originating from the gingival or alveolar mucosa epithelium is very rare. Peripheral Ameloblastic carcinomas may arise de novo and as dedifferentiated Ameloblastic carcinomas from pre existing benign peripheral ameloblastomas.[7] According to Avon et al Ameloblastic carcinoma is thought to originate from dental embryonic remnants, possibly from the rests of dental lamina, a developing enamel organ or the epithelial lining of an odontogenic cyst.[8]

Histomorphologically, two different Ameloblastic carcinoma entities may be recognized. One is characterized by lesions that initially demonstrate the morphology of a Solid Multiystic Ameloblastoma but dedifferentiate over time. Dedifferentiation may occur spontaneously or be related to surgical procedures that become necessary due to recurrences of the primary tumor or therapeutic radiation. The second entity comprises of those Ameloblastic Carcinomas that have malignant cytologic features de novo.[7]

Radiographically Ameloblastic carcinoma can be radiolucent, either unilocular or multilocular, which generally has a honeycomb appearance with tooth root resorption. It will often have distinct borders, slight marginal sclerosis without periosteal new bone formation, loss of lamina dura, resorption of the tooth apex and tooth displacement. CT demonstrates the aggressive nature of the tumour including the presence of significant cortical and tooth root destruction. MRI images reinforce the aggressive nature of the tumour, demonstrating large solid components, heterogeneous enhancement, cystic degeneration and extraosseous extension.[9]

Histologically Ameloblastic Carcinoma is composed of islands and cords of ameloblastomatous odontogenic epithelium in an infiltrative pattern with in a stroma of mature fibrous connective tissue. The epithelium may reveal a single outer layer of Ameloblastic cells of columnar to cuboidal shape which exhibit a tendency for palisading and reverse nuclear polarization; peripheral palisading and polarization are not always clearly evident. The stellate reticulum within epithelial islands is often condensed presenting a less orderly pattern. The characteristic differentiating features are nuclear enlargement, nuclear hyperchromatism, mild pleomorphism, an increased nuclear cytoplasmic ratio, and increased mitotic activity with abnormal forms of mitoses. Mitotic figures may attain a count of 2 to 5 per high-power field. In some cases, individual cell keratinization and keratin pearl formation may be seen. Necrosis and dystrophic calcifications also may be observed in some of the epithelial islands. The connective tissue is usually composed of mature collagen fibres with occasional inflammatory cells, haemorrhage, and/or hemosiderin pigment.[7] According to Elzay and Slootweg & Muller, the term ameloblastic carcinoma should be used to designate lesions that exhibit histologic features of both ameloblastoma and squamous cell carcinoma.[11, 12] Lau et al studied two cases of ameloblastic carcinoma immunohistochemically for the presence of cytokeratins and showed that the amelobtastomatous areas of the Ameloblastic

carcinomas reacted strongly with antibodies directed against cvtokeratins CAM 5.2 and AEl and AE3.[14]

Treatment

The rarity and unusual biologic behaviour of Ameloblastic carcinomas make it difficult to develop effective treatment protocols. However, the clinical course of these tumours is aggressive with extensive local destruction. The treatment of choice is radical surgery with neck dissection.[13] Some authors have recommended preoperative irradiation to decrease tumour size, but this seems to be of only limited value.[15] Meticulous follow-up is essential because of common recurrence and metastasis to the lung and regional lymph nodes.[4] The survival rate of ameloblastoma if metastasis occurs ranges from 3 months to 5 years. [16] Alexandra et al in 2011, first time, treated Ameloblastic carcinoma by radiotherapy with carbon ions. It showed complete remission at 6 weeks post completion of radiotherapy with only mild side effects.[17] In the present case complete resection of the tumour with at least 1.5cm of uninvolved bone is done (Figure 6). Reconstruction of the defect was done by microvascular fibula free one flap. Fibula bone flap along with skin paddle & paroneal artery and vein were harvested. The length of the vascular pedicle was approximately 3cm. Graft was then shaped to fit into the defect area and fixed with mini bone plates. The patient was advised to undergo Radiotherapy/Chemotherapy.

Conclusion

It is reasonable to assume that this case illustrates the malignant portion in the spectrum of ameloblastomas. It is possible that ameloblastoma shows a variety of histologic and biologic behaviours ranging from benignity to frank malignancy. Cases of ameloblastoma should thus be studied carefully, correlating their histological pattern with biologic behaviour to detect subtle changes in histology that may predict aggressive behaviour.

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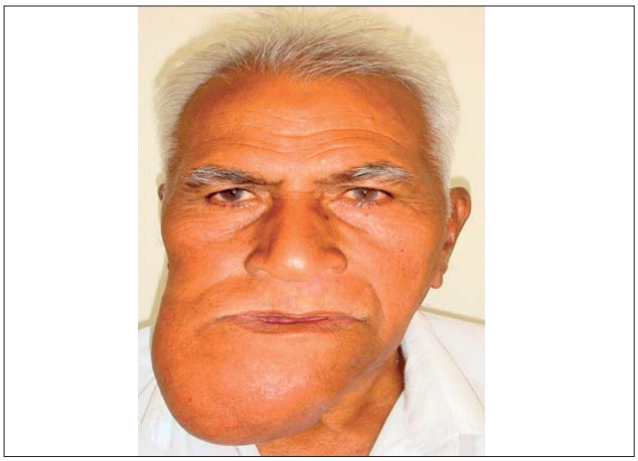

Figure 1: Extra oral view showing swelling on the right side of the face, roughly oval in shape. The skin over the swelling was normal. The chin was slightly deviated to the right side.

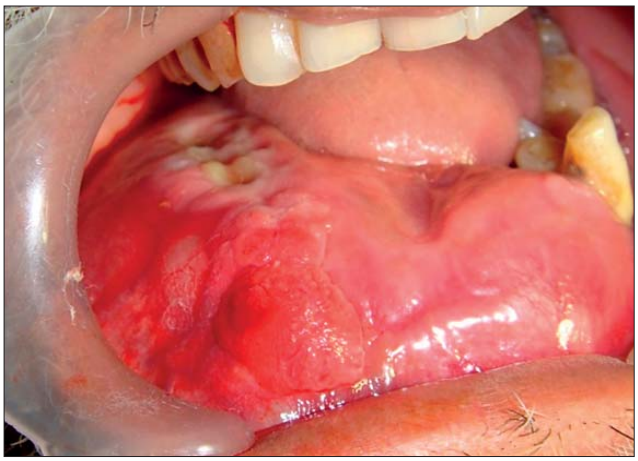

Figure 2: Intraoral examination revealed a single nodular growth extending from mandibular left lateral incisor to mandibular right 3rd molar. The growth enclosed the entire tooth surfaces in the right side of the mandible.

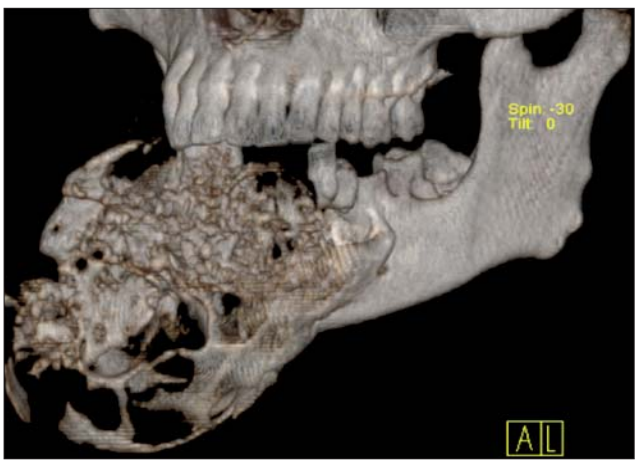

Figure 3: 3D CT Scan revealed an extensive osteolytic lesion involving both bucco-lingual cortical plates with complete perforation of the cortices and infiltration of the tumor mass in to the surrounding soft tissue.

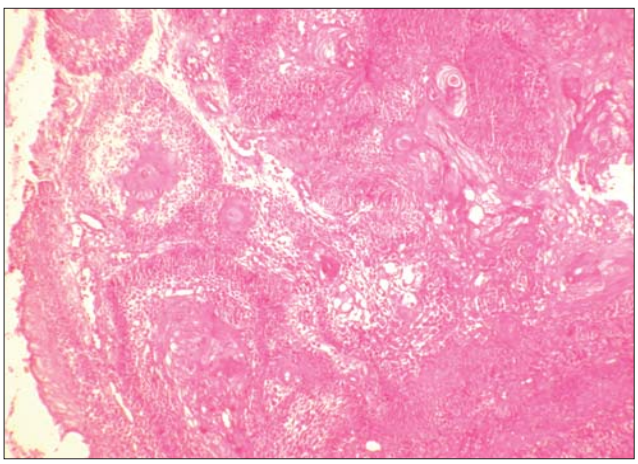

Figure 4: Section shows numerous ameloblastic follicles spread out in a scanty connective tissue stroma. The epithelial nests showed typical peripheral tall columnar cells with apically placed nuclei and vacuolated cytoplasm.

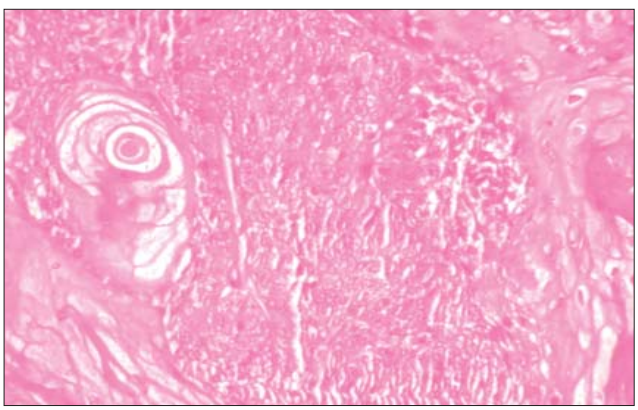

Figure 5: Section shows stellate reticulum like appearance with some cells showing squamous metaplasia and numerous keratin pearls. A few cells showed features of dysplasia such as cellular and nuclear pleomorphism with nuclear hyperchromatism.

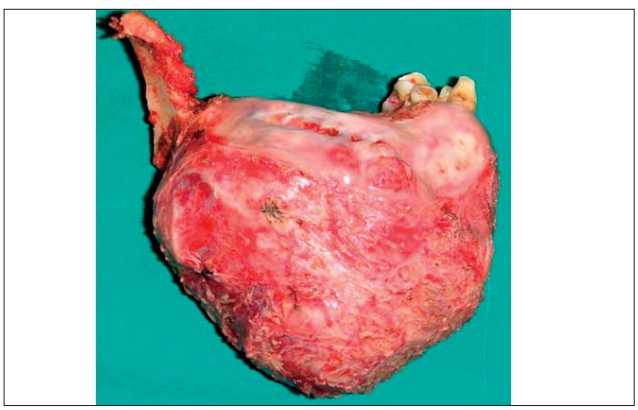

Figure 6: Complete resection of the tumour with at least 1.5cm of uninvolved bone.