International Journal of Otorhinolaryngology and Head and Neck Surgery
Virmani N et al. Int J Otorhinolaryngol Head Neck Surg. 2016 Apr;2(2):91-94
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20160962

Giant multiloculated ameloblastoma of the mandible: a case report and review of literature

Nitish Virmani*, Jyoti Dabholkar

Department of ENT and Head-Neck Surgery, Seth G.S. Medical College and KEM Hospital, Mumbai, Maharashtra, India

Received: 07 January 2016
Accepted: 04 February 2016

***Correspondence:**
Dr. Nitish Virmani,
E-mail: nitish_virmani@yahoo.com

ABSTRACT

Ameloblastomas, in general, are considered benign but locally invasive neoplasms. They present as slow growing, painless swellings and can grow to enormous sizes over the years. While planning the treatment of ameloblastoma, it is important to understand the growth characteristics and removing the full extension of tumor, including the surrounding tissue. Recurrence of ameloblastoma in many cases reflects the inadequacy or failure of the primary surgical procedure. We report a case of giant multiloculated ameloblastoma of the mandible with destruction of the cortical plate and extensive and rapid infiltration of the buccal mucosa. Along with the clinical and imaging features, the importance and method of ruling out malignant ameloblastoma and ameloblastic carcinoma in such a case is discussed.

Keywords: Ameloblastoma, Multiloculated, Ameloblastic carcinoma, Malignant ameloblastoma

INTRODUCTION

Odontogenic tumors comprise a diverse group of lesions with varied clinical behaviour and histological patterns. Although ameloblastoma is the most common odontogenic tumor, it is a relatively rare tumor occurring in the jaws, representing only around one percent of oral tumours. It is a benign but locally invasive tumor and is more common in the mandible. These tumors usually present as slow growing, painless swellings causing expansion and local destruction of cortical bone; they often grow to enormous size over the years without any malignant change. However, it is equally important to rule out malignant ameloblastoma and ameloblastic carcinoma in these patients. WHO has recently made the distinction between these three entities in 2005 classification. We report a case of giant multiloculated ameloblastoma of the mandible with destruction of the cortical plate and extensive and rapid infiltration of the buccal mucosa. Along with the clinical and imaging features, the importance and method of ruling out malignant ameloblastoma and ameloblastic carcinoma in such a case is discussed.

CASE REPORT

A 36 year old male patient presented to our OPD with complaints of a progressively increasing swelling over his right lower jaw for 2 years. There was no associated pain or discharge from the swelling. He did not have any complaints of difficulty in swallowing or breathing. Around 6 months back, he noticed an oral ulcer on the inside of his right cheek which rapidly progressed to involve the entire right buccal mucosa. He did not give history of addictions to either alcohol or tobacco. He had undergone an evaluation in his home country followed by receiving 10 cycles of chemotherapy, the records of which were not available to us for review. He did not

observe any response in terms of either reduction/stabilisation of the tumor and was then referred to our centre. On physical examination, there was a large 15 X 12 X 10 cm swelling over the right mandible extending superiorly to zygoma, and medially to symphysis menti (Figure 1). The surface was lobulated and it was firm too hard on palpation. Overlying skin was normal. Oral cavity revealed an ulceroproliferative growth involving the right buccal mucosa and lower gingiva, extending anteriorly upto the region of canine tooth 1 cm from the oral commissure. Posteriorly, it was extending to involve the anterior tonsillar pillar and tonsillo-lingual sulcus. Inferiorly, the right side of floor of mouth was also involved (Figure 2).

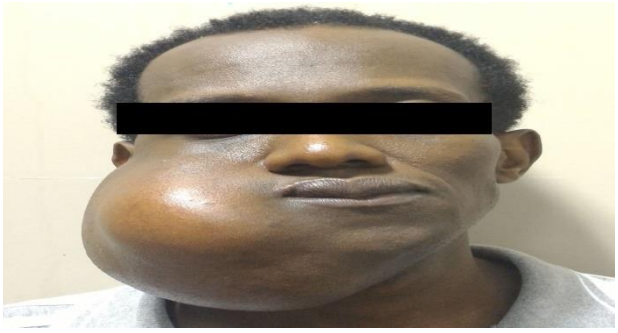

Figure 1: Large, multilobulated swelling over left side of face.

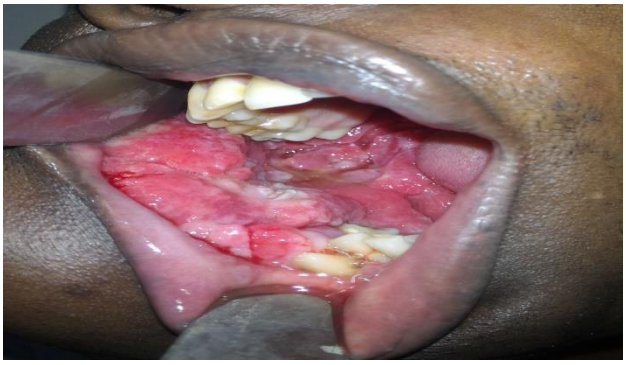

Figure 2: Ulceroproliferative growth involving right buccal mucosa, lower gingiva, anterior tonsillar pillar, tonsillo-lingual sulcus and right side of floor of mouth.

A CT scan of the face and neck revealed a large multiloculated soft tissue density involving the right side of the face causing destruction of the right hemimandible including the mandibular arch (Figure 3). The lesion extended superiorly into the high infratemporal and temporal fossa (along the squamous part of temporal bone). Pterygoid muscles could not be distinctly identified; pterygoid plates, however, appeared intact. Inferiorly, it extended to involve the floor of mouth. There were both solid and cystic areas with scattered calcifications. The lesion did not show any enhancement on administration of contrast. A punch biopsy from the ulceroproliferative oral lesion was performed which was suggestive of ameloblastoma extending to overlying squamous mucosa. Considering the extensive and rapid involvement of oral mucosa, we did a PET-CT to rule out any distant metastatic deposits. It revealed a FDG avid heterogeneously enhancing soft tissue mass with SUV max of 13.32. No lymph node or distant metastatic deposits were noted.

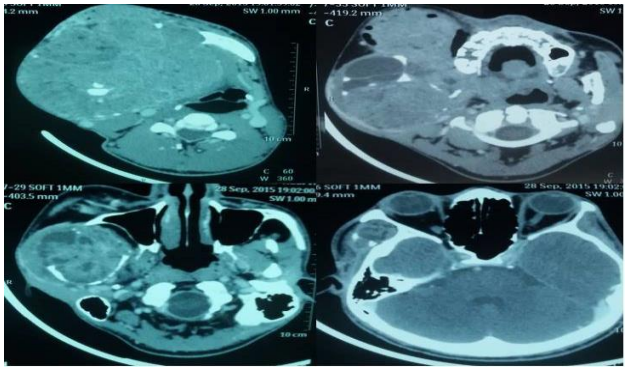

Figure 3: CT scan showing a large multiloculated soft tissue density causing destruction of the right hemimandible including the mandibular arch. Both solid and cystic areas with scattered calcifications. Superior extension into high infratemporal and temporal fossa.

The patient underwent a wide excision of the mass which entailed a segmental resection of the right hemimandible including the mandibular arch with 2 cm margin of healthy bone (Figure 4). Reconstruction of the large mandibular defect (Figure 5) was done using osteotimised vascularised free fibula flap and fixation by titanium plates. Buccal mucosa and floor of mouth defect was reconstructed using the skin paddle of free fibular flap (Figure 6). Tracheostomy was done due to loss of tongue support consequent to resection of mandibular arch. Post-operative period was uneventful without any complications. Oral feeding was successfully resumed on day 10. The patient was decannulated by 2 weeks post-operatively. He achieved a good facial symmetry and proper occlusion (Figure 7). Histopathological report of the resected specimen was suggestive of ameloblastoma with all margins free of tumor.

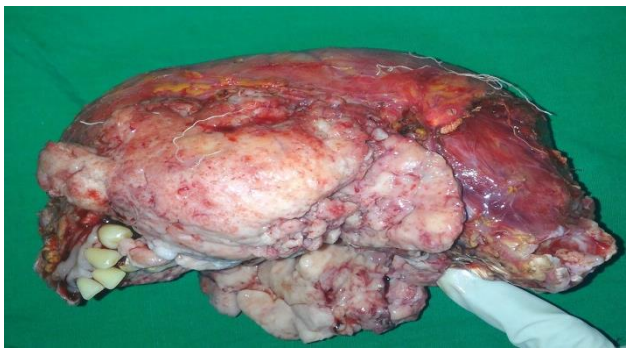

Figure 4: Wide excision of tumor with right hemimandibulectomy.

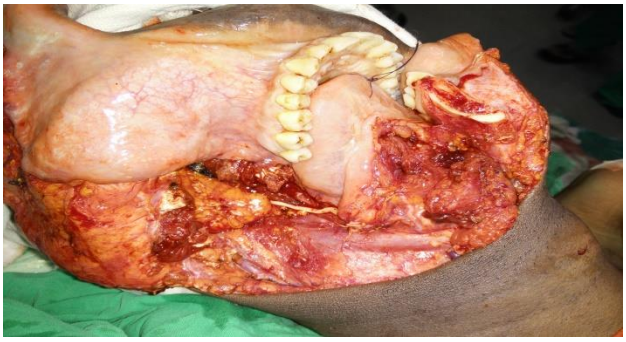

Figure 5: Large bony and soft tissue defect.

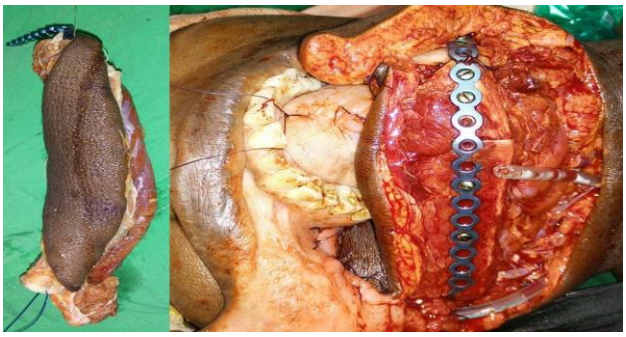

Figure 6: Reconstruction using microvascular free fibular flap.

Figure 7: One month post-operatively.

DISCUSSION

Ameloblastoma, previously known as admantinoma, is a benign but locally aggressive tumor of the jaw that tends to arise from the odontogenic epithelium. The first detailed description of this tumor was published by Falkson in 1879, but the term 'ameloblastoma' was coined by Churchill in 1933.[2] Although it is the most common odontogenic tumor, it represents only around one per cent of oral tumors. Around 80 per cent of ameloblastomas are reported to occur in the mandible.[1]

The current concept is to classify ameloblastoma into solid or multicystic, unicystic, and peripheral types. This classification has direct bearing on the clinic-pathological behaviour of these tumors. While solid or multicystic variants are considered to be locally aggressive, and recur if inadequately excised; the unicystic variant has been identified with a less aggressive behaviour.[3,4]

In the recent WHO classification, a clear distinction was made between ameloblastoma, malignant ameloblastoma and ameloblastic carcinoma. It defines malignant ameloblastoma (MA) as "an ameloblastoma that metastasizes in spite of a benign histological appearance." Ameloblastoma with cytological atypia is defined as ameloblastic carcinoma even if metastasis is absent.[5] MA has been reported as a heterogeneous clinico-pathological entity that consists of tumors with varying histological and clinical behaviors from very aggressive to highly indolent. Ameloblastic carcinomas are more aggressive than most typical ameloblastomas. Perforation of the cortical plate, extension into surrounding soft tissue, numerous recurrent lesions and metastasis, usually to cervical lymph nodes, can be associated with ameloblastic carcinomas.[6]

Our patient presented with quite an aggressive tumor with rapid growth, perforation of the inner cortical plate of the mandible and extension into the overlying buccal mucosa. It is essential to make a distinction between the three entities in such an aggressive case. This necessitated a pre-operative biopsy as well as a PET scan. The biopsy confirmed the benign histological features of ameloblastoma, thus ruling out ameloblastic carcinoma. At the same time, absence of distant metastasis on PET scan ruled out malignant ameloblastoma. Thus, the tumor was confirmed to be an unusually aggressive multicystic variant of ameloblastoma.

While planning the treatment of ameloblastoma, it is important to understand the growth characteristics and removing the full extension of tumor, including the surrounding tissue.

Treatment of ameloblastoma is primarily surgical. The surgical options range from curettage, enucleation and cryosurgery to wide local excision which usually necessitates segmental resection in the mandible. There seems to be a lack of consensus regarding the most appropriate method of surgical removal of ameloblastomas. Proponents of conservative approach argue that ameloblastoma though locally invasive is essentially a benign tumor. Many authors have recommended enucleation with preservation of periosteum which is important for bone regeneration especially in children.[7] However, proponents of radical approach are of the considered view that conservative surgical options such as curettage and enucleation result in unacceptably high recurrence rates; the recurrence rate up to 55–90% have been reported in the literature.[8] Curettage is followed by local recurrence in 90% of mandibular and all maxillary ameloblastomas because of insufficient removal of tumors.[9] Sehdev et al reported recurrence after the conservative approach (curettage) in more than 90% of 92 ameloblastomas.[10] Ameloblastoma has a persistent and slow growth, spreading into marrow

spaces with pseudopods without concomitant resorption of the trabecular bone. As a result, the margins of the tumour are not clearly evident radiographically or grossly during operation, and the lesion frequently recurs after inadequate surgical removal, showing a locally malignant pattern.[11]

At our centre, we recommend a radical approach for all ameloblastomas with a margin of atleast 2 cm of healthy bone. In addition, this particular patient had an unusually aggressive variant of the tumor, all the more reason to perform a wide excision. Because of the giant size of the tumor as well as wide involvement of buccal mucosa and floor of mouth, he had a huge defect of soft tissue and bone post-resection, which required reconstruction by a fibular free flap. Free fibular flap offers lot of advantages in mandibular reconstruction.[12] It allows for transfer of bone, soft tissue and skin in a one-stage procedure using only one donor site, fibula flap allows for a skin paddle up to 25 cm in length and 5 cm in width, and bone up to 25 cm. Blood supply to fibula is both intraosseous and segmental, therefore, multiple osteotomies can be made.[7] With such microvascular reconstructive techniques, very good aesthetic and functional results are obtained post-operatively.

Ameloblastoma is generally considered to be a radioresistant tumor although there is evidence to suggest a palliative role in advanced cases to diminish the volume of tumor. It may reduce the risk of progression and result in long-term local control in such incompletely resectable tumors.[13] However, post-operative adjuvant radiotherapy does not seem to offer any advantage in terms of loco-regional control or overall survival in patients who have undergone complete excision of the tumor with negative pathological margins.

CONCLUSION

Malignant ameloblastoma and ameloblastic carcinoma should be ruled out by a PET scan and biopsy respectively, in tumors associated with rapid growth or extensive soft tissue infiltration or perforation of cortical plate. We recommend radical approach for all ameloblastomas with atleast 2 cm margin of healthy bone. Giant tumors require fibular free flap reconstruction for optimum aesthetic and functional outcome.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Small IA, Waldron CA. Ameloblastoma of the jaws. Oral Surg. 1955;8:281-97.
2. Lucas RB. Pathology of tumours of the oral tissues. 4th edn. Edinburgh: Churchill Livingstone, 1984:31-59.
3. Adeyemo WL, Bamgbose BO, Ladeinde AL, Ogunlewe MO. Surgical management of ameloblastomas: conservative or radical approach? A critical review of the literature. Oral Surg. 2008;1:22-7.
4. Jackson IT, Callan PP, Forte RA. An anatomic classification of maxillary ameloblastoma as an aid to surgical treatment. J Craniomaxillofac Surg. 1996;24:230-6.
5. Sciubba JJ, Eversole LR, Slootweg PJ. Pathology & Genetics of Head and Neck Tumours. LYON: IARC Press; 2005. p. 287.
6. RL Corio, LI Goldblatt, PA Edwards, KS Hartman. Ameloblastic carcinoma: a clinicopathologic study and assessment of eight cases. Oral Surg Oral Med Oral Pathol. 1987;64(5):570-76.
7. Sharma A, Shaikh AM, Deshmukh SV, Dabholkar JP. Radical Management of Giant Ameloblastomas: a case series. Indian Journal of Otolaryngology and Head & Neck Surgery. 2012;64(4):399-401.
8. Ghandhi D, Ayoub AF, Anthony M, MacDonald G, Brocklebank LM, Moos KF. Ameloblastoma: a surgeon's dilemma. J Oral Maxillofac Surg. 2006;64:1010-4.
9. Vanoven BJ, Parker NP, Petruzzelli GJ. Peripheral ameloblastoma of the maxilla: a case report and literature review. Am J Otolaryngol. 2008;29(5):357-60.
10. Sehdev MK, Huvos AG, Strong EW, Gerold FP, Willis GW. Ameloblastoma of maxilla and mandible. Cancer. 1974;33:324-33.
11. Gardner DG, Pecak AMJ. The treatment of ameloblastoma based on pathologic and anatomic principles. Cancer. 1908;46:2514-9.
12. Waldron CA. Ameloblastoma in perspective. J Oral Surg. 1966;24:331-3.
13. Gardner DG. Radiotherapy in the treatment of ameloblastoma. Int J Oral Maxillofac Surg 1988;17:201-5.

Cite this article as: Virmani N, Dabholkar J. Giant multiloculated ameloblastoma of the mandible: a case report and review of literature. Int J Otorhinolaryngol Head Neck Surg 2016;2:91-4.