Case Report

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Large ameloblastic carcinoma of mandible: Its excision and reconstruction: A rare case report and review of literature

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Abstract:

This article describes a rare case of a large (23x13.4 cm) sized ameloblastic carcinoma of the mandible encountered in a 69 year-old patient and its management. The article also reviews the literature regarding the pathogenesis, classification, diagnosis and management options of Amelobastic Carcinoma and includes an insight into microvascular reconstructive procedures in such situations.

Key words: Ameloblastic carcinoma, Free fibular flap, Mandibular reconstruction, Microvascular anastomosis

Introduction

Ameloblastoma is a tumor of the jaws that arises from odontogenic remnants and represents 1% of all jaw tumors. It is regarded as a "prototypical benign neoplasm" [1]. Ameloblastomas infrequently, however, exhibit malignant clinical and histologic features, which results in the recognition of two related neoplasms: malignant ameloblastoma and ameloblastic carcinoma (AC). The frequency of ameloblastic carcinoma exceeds that of malignant ameloblastoma by a 2:1 ratio [2]. It is important to understand the difference between these tumors.

Malignant ameloblastoma refers to a metastatic but otherwise benign ameloblastoma. Benign secondaries most commonly involve the lung (75-80%) [3,4], but have also been reported in cervical lymph nodes and bone [5-7]. Ameloblastic carcinoma refers to an ameloblastoma that exhibits malignant histopathological features [6]. These features include cytological atypia,

hyperchromatism, basilar hyperplasia, and greater mitotic activity than benign ameloblastomas [8,9]. Metastasis is an irregular finding but if present usually occurs to the lungs [1,10,11].

AC is an exceedingly rare tumor with a poor prognosis. It can develop from a pre-existing ameloblastoma (carcinoma ex ameloblastoma) or autonomously with a histologic resemblance to ameloblastoma (*de novo* ameloblastic carcinoma) [8]. The tumor primarily affects the mandible (2/3 of cases) and has been reported in many age groups with no noted predilection for any race. Some authors have described a slight predominance in males [12]. The typical presentation of the tumor is similar to ameloblastoma but it exhibits more aggressive growth and recurrence. Clinical features include swelling with or without pain, tissue ulceration, paresthesia, and trimus [9]. Radiographic features include uni- or multicystic osteolytic areas with cortical bone expansion and thinning [9].

There is no standard treatment protocol, however, and ameloblastic carcinoma is typically approached like a squamous cell carcinoma [8]. Ameloblastic carcinoma is managed with an aggressive surgical excision and neck dissection. Carbon ion therapy has proven to be of use despite some authors reporting questionable efficacy of radiotherapy for intrabony tumors [6,13].

Case report

A previously healthy 69 year-old patient reported to the department of oral and maxillofacial surgery with a complaint of a progressively increasing swelling in the right side of the face since 71/2 years. He did not complain of any pain or tenderness.

On extra-oral examination, there was a large ovoid swelling measuring 23x13.4cm that was fixed, extending from left molar region crossing the midline to right mandibular angle anteroposteriorly and from the ala-tragus line to the level of the thyroid cartilage superoinferiorly (Fig. 1). Palpation revealed a fibrous, firm, non-tender mass with no signs of fluctuation and normal overlying skin. There were no palpable regional nodes.

Intraoral examination revealed a firm mucogingival swelling involving the whole of the 4th quadrant and the anterior half of the 3rd quadrant. The soft tissue was erythematous and bleeding at several points of ulceration particularly due to trauma from opposing maxillary teeth. The large mass of the 4th quadrant swelling prevented left side tooth contact and also caused displacement of the tongue to the left side (Fig. 2).

A provisional diagnosis of ameloblastoma and a differential diagnosis of primary intraosseous carcinoma were made.

Orthopantomography revealed a large, poorly defined multilocular radiolucency involving the entire body of the mandible extending from right angle, crossing the midline to involve the left molar region with thinning of the lower border of mandible. Resorption of teeth was also present . CT and 3D CT revealed an extensive osteolytic lesion with buccolingual expansion with complete perforation of the cortices and extensive infiltration of the tissue mass into the soft tissue (Fig.3).

Histologically, section revealed numerous epithelial follicles spread out in a scanty connective tissue stroma. The epithelial nests show typical tall columnar peripheral cells with apically placed nuclei and vacuolated cytoplasm. The central cells show stellate reticulum like appearance with some ells showing squamous metaplasia and numerous keratin pearls. A few cells are showing features of dysplasia such as irregular aggregation, cellular and nuclear pleomorphism with nuclear hyperchromasia. (Fig. 4). The pathological diagnosis was ameloblastic carcinoma.

The patient underwent surgery under general anesthesia. The tumor was excised along with a margin of 1.5 cm of healthy bone (Fig. 5). Local lymph nodes were excised and sent for post-op histopathological examination. The mandibular defect was reconstructed using a free fibular flap along with skin paddle and peroneal artery and vein (Fig. 6). Microvascular anastomosis of the peroneal artery and vein to the facial artery and vein was done and the bone was contoured and fixed with mini bone plates. The skin flap was used to cover the intraoral wound (Fig. 7). The anesthesia was reversed and the patient was wheeled out with NTT. Extubation was done the following day. Postoperative healing was uneventful and graft blood supply was adequate as confirmed by a Doppler test performed on the 2nd postoperative day.

Microscopic examination of H and E stained lymph node sections did not show any regional metastasis. Follow-up continued for next 2 1/2 years. (Fig.8,9).

Discussion

The identification and classification of ameloblastic carcinoma has long been a topic of confusion and controversy. AC was widely

recognized in 1971, when the World Health Organization (WHO) classified odontogenic carcinomas into the following groups [14]. a).Malignant ameloblastoma ,b).Primary intraosseous carcinoma(PIOCs), c).Other carcinomas arising from odontogenic epithelium, including those arising from odontogenic cysts. Malignant ameloblastoma was considered as any primary or secondary ameloblastoma exhibiting malignant changes. As per current definition, this type of tumor would be considered as an AC.

In 1982, Elzay proposed a classification of carcinomas that would classify all primary intraosseous carcinomas without salivary gland involvement as PIOCs. He recognized an important histologic differentiation between benign but metastasizing ameloblastomas and histologically malignant ameloblastomas: Type 1: arising from an odontogenic cyst, Type 2: arising from an ameloblastoma: a. Well-differentiated (malignant ameloblastoma), b. Poorly-differentiated (ameloblastic carcinoma), Type 3: arising de novo : a. Nonkeratinizing, b.Keratinizing. Slootweg and Müllerin in 1984 proposed another classification of carcinomas. In this, they differentiated malignant ameloblastoma and ameloblastic carcinoma based on histological features of malignancy: Type 1: PIOC ex odontogenic cyst , Type2:a. Malignant ameloblastoma, b. Ameloblastic carcinoma, arising de novo, ex ameloblastoma or ex odontogenic cyst, Type 3: PIOC arising de novo : a. Nonkeratinizing, b. Keratinizing.

Slootweg and Müller defined ameloblastic carcinoma "as a tumor combining morphologic features of both ameloblastoma and carcinoma, which can arise de novo, ex ameloblastoma, or ex odontogenic cyst." [11,15] This definition is still accepted today.

The neoplasm is described to arise from odontogenic remnants [8]. It resembles the benign neoplasm ameloblastoma both clinically and histologically. As stated, AC may arise from an existing ameloblastoma (carcinoma ex ameloblastoma) or independently (*de novo* AC). In the presence of an existing ameloblastoma, the carcinoma arises when the benign lesion undergoes focal "dedifferentiation," and the aggressive replica overgrows the existing benign ameloblastoma [8]. *De novo* AC is a tumor that demonstrates histologic features of ameloblastoma, but presents several heterogeneous features, particularly cellular atypia and other malignant changes. Both variants may present clinically as a swelling in the jaw with or without ulceration, pain, paresthesia, and trismus. Therefore, aggressive behavior of the tumor with any histologic evidence of malignancy clenches the diagnosis.

Surgical excision with wide margins is generally the preferred method though radiotherapy has been used successfully [13]. Recurrence ranges from 15 to 25% after wide surgical excision [15]. Recurrence rates of 90% have been reported for local curettage without surgical excision. Some authors suggest preoperative radiotherapy in order to decrease tumor size. No extensive studies have been presented which demonstrate the role of chemotherapy in treatment [16]. Treatment in this case consisted of surgical resection with 1.5 cm margins. Considering the duration of the lesion (71/2 years), age of the patient, and lack of clinical & radiological evidence of local lymph node metastasis no attention was given to neck. However it was planned to remove few regional lymph nodes at the time of resection so that should they appear positive, post operative radiotherapy could be given. However since microscopic examination of H and E stained resected lymph node sections did not show any regional metastasis no further treatment was given to patient. Postoperative functional loss and esthetic deformity are two major concerns when excising a tumor of such proportions. Reconstruction of such a large mandibular defect was thus decided to be done with a free fibular flap.

Several methods exist for reconstruction of segmental mandibular defects: nonvascularized bone grafts (NVBGs), titanium reconstructive splints, or free flap transfers that allow the use of vascularized bone. Among the latter, iliac crest, scapula and fibular flaps are most widely used for mandible reconstruction [17]. Free rib or radius are also options but with greater limitations. The use of free fibular flaps offers many solutions to problems encountered in reconstructive mandibular surgery most notably, graft uptake and vascularization. Success rates of 98% have been reported [10].

The fibula is a laterally situated bone of the lower limb that has both endosteal and periosteal vascularization. Vascular pedicle length (up to 8 cm) and vessel caliber (2-3 mm peroneal artery and 3-4 mm peroneal vein) allow for micro anastomosis of the graft to the facial artery and vein to permit immediate perfusion to most of the grafted tissue [17]. The dual endosteal and periosteal vascularity permits multiple segmental osteotomies to be performed on the graft during shaping. This allows for enhanced positioning of bone segments to

facilitate plating as well as precise anatomic contouring. Also, up to 25 cm of bone may be harvested which will suffice for mandibular defects of any size [17]. Other advantages of using a free fibular flab for mandibular reconstruction include decreased resorption, immediate allowance of endosteal implants, low morbidity of the donor site, and the possibility of simultaneous soft and hard tissue reconstruction with the same composite flap [18,19].

Disadvantages include the inability to reproduce adequate height of alveolar bone, inability to reconstruct large soft tissue defects, inability to use in the presence of peripheral vascular disease, and decreased vascularization when a large number of osteotomies are required [10,20].

We had an acceptable functional and esthetic outcome in this particular case with satisfactory graft uptake, uneventful healing, and no immediate or delayed complications. Long-term follow-up is ongoing.

One should be alert for the possibility of local recurrences and distant metastases especially to the lung, bone, or brain [10]. A regular assessment of the chest by periodic imaging is recommended [16].

Conclusion

In conclusion, we have reported the clinical, radiographic, CT, and histologic findings in a rare case of a huge ameloblastic carcinoma of the mandible. A satisfactory surgical approach of excision and reconstruction using a free fibular flap has been summarized.

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References

1. Devenney-Cakir B., Dunfee B., Subramaniam R., et al. Ameloblastic Carcinoma of the Mandible with Metastasis to the Skull and Lung: Advanced Imaging Appearance Including Computed Tomography, Magnetic Resonance Imaging and Positron Emission Tomography Computed Tomography. *Dentomaxillofacial Radiology* 2010; 39(7): 449-453.
2. Regezi JA, Kerr DA, Courtney RM. Odontogenic tumors: analysis of 706 cases. *J Oral Surg* 1978; 36(10):771–8.
3. Henderson JM, Sonnet JR, Schlesinger C, Ord RA: Pulmonary metastasis of ameloblastoma: case report and review of the literature. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 1999; 88(2):170-6.
4. Houston G, Davenport W, Keaton W, Harris S: Malignant (meta- static) ameloblastoma: report of a case. *J Oral Maxillofacial Surgery* 1993; 51(10):1152-5.
5. Kunze E, Donath K, Luhr HG et al. Biology of metastasizing ameloblastoma *Pathol Res Pract* 1985; 180 (5): 526-35
6. Astrid LD Kruse, Roger A Zwahlen and Klaus W Grätz: New classification of maxillary ameloblastic carcinoma based on an evidence-based literature review over the last 60 years. *Head & Neck Oncology* 2009.
7. Laughlin EH: Metastizing ameloblastoma. *Cancer* 1989; 64 (3): 776-80.
8. Rajendran R, Sivapathasundaram B.: Shafer's textbook of Oral pathology. 5th ed. India: Reed-Elsevier India Private Limited; 2006: 417-418.
9. Hye-Jung Yoon,Sam-Pyo Hong,Jae-Il Lee,Sam-Sun Lee,Seong-Doo Hong: Ameloblastic carcinoma: an analysis of 6 cases with review of the literature. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 2009.
10. Jatin P. Shah, Snehal G. Patel : Cancer of the Head and Neck. *Ca-A Cancer Journal for Clinicians* 1995; 45(6):352-368.
11. Slootweg PJ, Müller H: Malignant ameloblastoma or ameloblastic carcinoma. *Oral Surg Oral Med Oral Pathol* 1984; 57:168-176.
12. Lolachi CM, Madan SK, Jacobs: Ameloblastic carcinoma of the maxilla. *JR J Laryngol Otol. 1995; 109(10):1019-22.*
13. Alexandra D Jensen, Swantje Ecker, Malte Ellerbrock, et al.: Carbon Ion therapy for Ameloblastic Carcinoma. *Radiation Oncology* 2011; 6:13.
14. Barnes L, Eveson JW, Reichart P and Sidransky D: WorldHealth Organization Classification of Tumours: Pathology and Genetics Head and Neck Tumours, 2005.
15. Yoon HJ, Hong SP, Lee JI, Lee SS, Hong SD. Ameloblastic carcinoma: an analysis of 6 cases with review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2009; 108(6): 904-13.
16. Diurianne Caroline, Campos França, João Milanez Moreira, Jr, et al.: Ameloblastic carcinoma of the maxilla: A case report. *Oncol Letters* 2012; 4(6): 1297–1300.
17. Dragos Pieptu, Dan Gogalniceanu, Nicolae Getu, et al. : Mandible Reconstruction Using the Free

Osteocutaneous Fibula Flap. *Timisoara Medical Journal* 2005.

18. Elsalanty, Genecov: Bone Grafts in Craniofacial Surgery. *Craniomaxillofacial Trauma Reconstr* 2009; 2(3): 125–134.
19. Srinivas R Ponnam, Gautam Srivastava, Sudhakar G: Ameloblastic Carcinoma with Diverse Histological Features: A Case Report. *International Journal of Oral & Maxillofacial Pathology* 2012; 3(1):60-64
20. Ferri J, Piot B, Ruhin B, Mercier J: Advantages and limitations of the fibula free flap in mandibular reconstruction. *J Oral Maxillofac Surg*. 1997; 55(5): 440-8; discussion 448-9

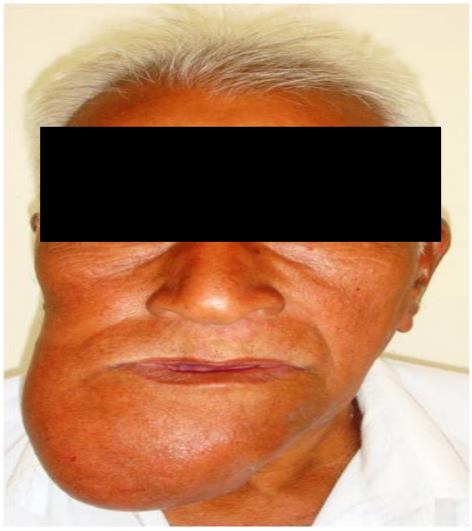

Figure 1: Extra Oral View

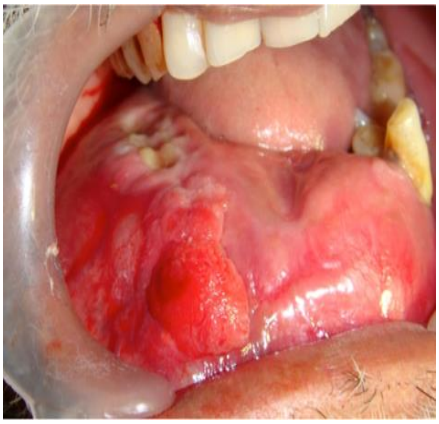

Figure 2: Intra Oral Examination

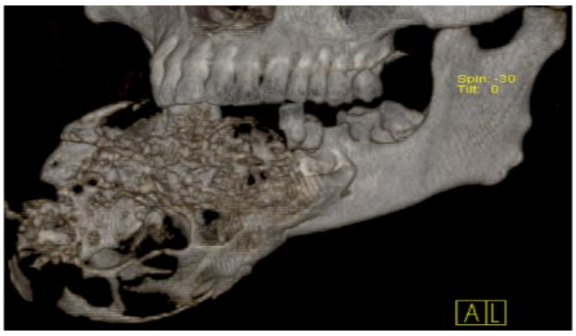

Figure 3: CT SCAN

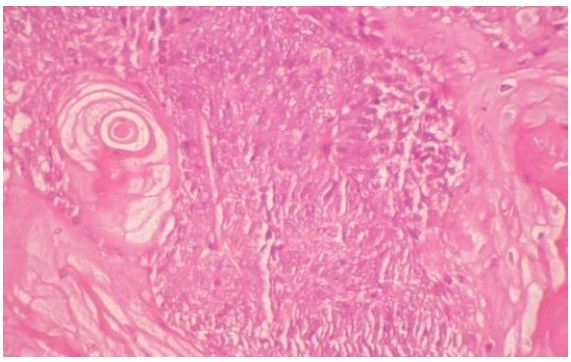

Figure 4: HP Slide showing Keratin Pearls

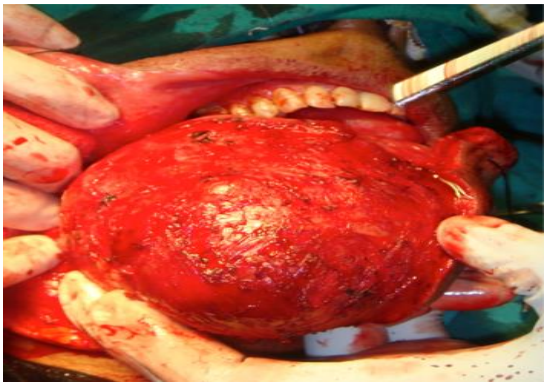

Figure 5: Tumor Resection

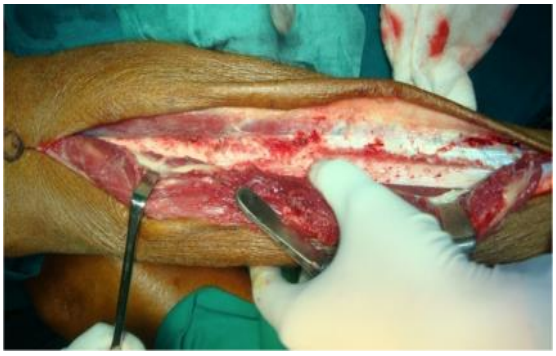

Figure 6: Fibular Flap Harvesting

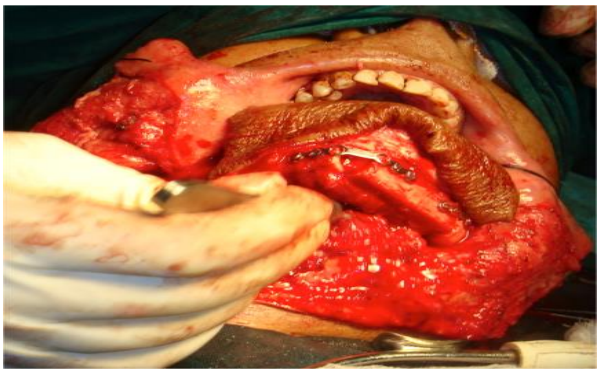

Figure 7: Fixation

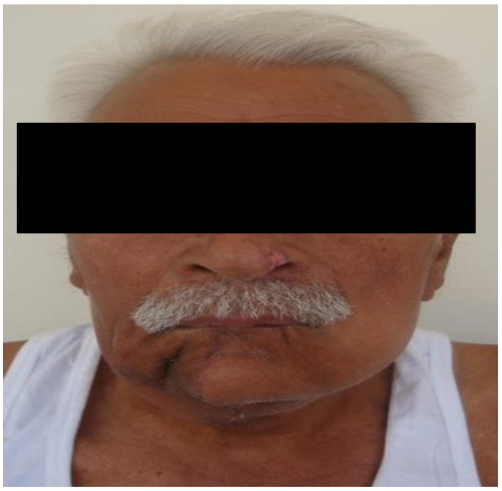

Figure 8: Post Op Frontal View

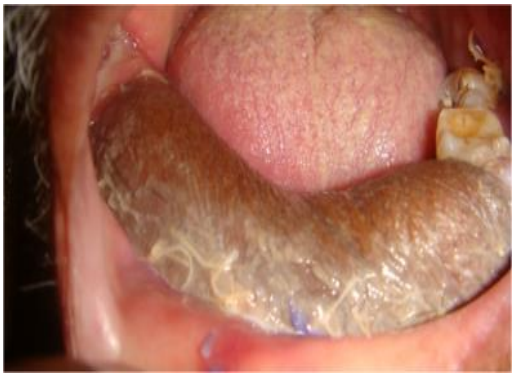

Figure 9: Intra Oral