

Up-to date review and case report

Ameloblastic carcinoma of the mandible: A case report and review of the literature

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(Received 31 December 2015, accepted 26 September 2016)

Key words:
ameloblastic
carcinoma / mandible /
surgical treatment

Abstract – Aim: The purpose of this paper was to report an additional case of ameloblastic carcinoma of the left hemi-mandible and to review and analyze clinical, radiological and histopathologic features, treatment modalities and prognosis. **Presentation of case:** A 70 year old male patient presented to Maxillofacial Department for a painful swelling of the left cheek. An ameloblastic carcinoma of the left hemi-mandible was diagnosed. He underwent radical surgical intervention comprising hemimandibulectomy, supraomohyoid neck dissection, and fibula free-flap reconstruction. Postoperative radiotherapy was employed. **Discussion:** Ameloblastic carcinoma of the mandible is a rare aggressive tumor with a poor prognosis. It can be seen in different ages. The most common symptom is a rapidly progressing painful swelling. It is defined as a malignant epithelial odontogenic tumor that retains the features of ameloblastic differentiation with cytologic features of malignancy. Non therapeutic consensus is established. However, wide surgical excision with radiotherapy is the most common treatment. **Conclusion:** Diagnosis of ameloblastic carcinoma at early stage and close periodic screening for metastasis are necessary to improve patient prognosis.

Introduction

Ameloblastic carcinoma (AC) is extremely rare, aggressive malignant epithelial odontogenic tumor with a poor prognosis. It can arise de novo or in a preexisting benign ameloblastoma [1].

Differentiating ameloblastic carcinoma from ameloblastoma and malignant ameloblastoma in a patient presenting with a suspicious jaw tumor is a challenge due to overlapping clinical features, inconclusive cytology/biopsy reports, different management approaches.

Metastasis occurred most commonly to the lung [2]. There are no guidelines for the treatment for AC owing to the rarity of this disease.

We report a case of ameloblastic carcinoma of mandible in 70 year old patient with difficult differential diagnosis and unusual outcome.

Observation

A 70-year-old man came to the Department of Maxillofacial Surgery, with a complaint of swelling in the left cheek that he first noticed 2 months previously. He hadn't prior history. Clinical examination revealed a painless mass with asymmetry in the left body region of the mandible. There was no sign of trismus or anaesthesia. Intra-oral examination showed a partially edentulous mouth. Teeth 31, 32, 33 and 34 were vital and immobile. An ulcerated lesion was observed in the left retro-molar region (Fig. 1). Examination of the neck showed no lymphadenopathy.

Panorex wasn't done. CT scan revealed an expansile mixed radiolucency measuring 60 × 60 mm in the left ramus, mandibular angle and body of the mandible perforating the buccal and lingual cortices with soft tissue extension (Figs. 2 and 3). PET-scan wasn't available.

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Fig. 1. Intraoral photograph showing the appearance of the lesion.

An incisional biopsy was made. Microscopically, the lesion consisted of a tumor mass that exhibited islands and cords of odontogenic epithelium with palisading columnar cells at the periphery of the islands. The histopathologic and immunohistochemical features brought us to the diagnosis of ameloblastic carcinoma (Fig. 4).

The patient underwent radical resection which included a left hemimandibulectomy up to the parasymphseal region (Fig. 5) and prophylactic supraomohyoid neck dissection in order to prevent delayed metastases. Surgical margins were clear. The surgical defect was reconstructed immediately with a fibula osteocutaneous free flap (Fig. 6). The patient received 60 Gray of radiotherapy postoperatively. The 4-year postoperative follow-up revealed neither local nor distant metastasis.

Discussion

Carcinomas derived from ameloblastomas have been given many designations such as malignant ameloblastoma, metastatic carcinoma and primary intra-alveolar epidermoid carcinoma.

Slootweg and Muller [1] subclassified odontogenic carcinomas into three categories as they felt that these tumours exhibit considerable differences in biological behaviour and histomorphology. The sub-classification is as follows:

Type 1. Primary intraosseous carcinoma arising from odontogenic cyst.

Type 2.

A. Malignant ameloblastoma

B. Ameloblastic carcinoma, arising de novo, ex ameloblastoma or odontogenic cyst.

Type 3. Primary intraosseous carcinoma arising de novo

A. Nonkeratinizing

B. Keratinizing.

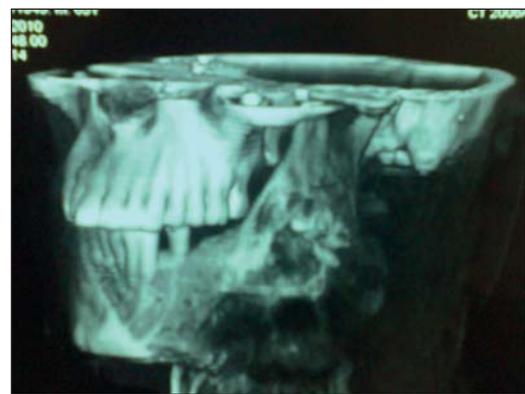


Fig. 2 and 3. CT Scan: osteolytic mass of the left mandibular ramus with hyperostotic border and cortical bone expansion.

According to the World Health Organization's (WHO) classification [3], ameloblastic carcinoma is in the group C of odontogenic carcinomas.

The consensus was to use the term ameloblastic carcinoma for those tumours with histological evidence of malignancy in the primary, recurrent, or metastatic tumour regardless of whether there is metastasis or not while malignant ameloblastoma is reserved for metastasizing ameloblastomas which exhibit benign histological features both in the primary and metastatic lesion [4, 5].

According to Benlyazid et al. in 2007 a total of 67 cases of ameloblastic carcinoma have been reported in the literature including one case reported by them [6]. According to Sciubba et al, [7] mean age was 30.5 years, the male to female ratio was 1.5 to 1, and the most commonly involved area is the posterior portion of the mandible.

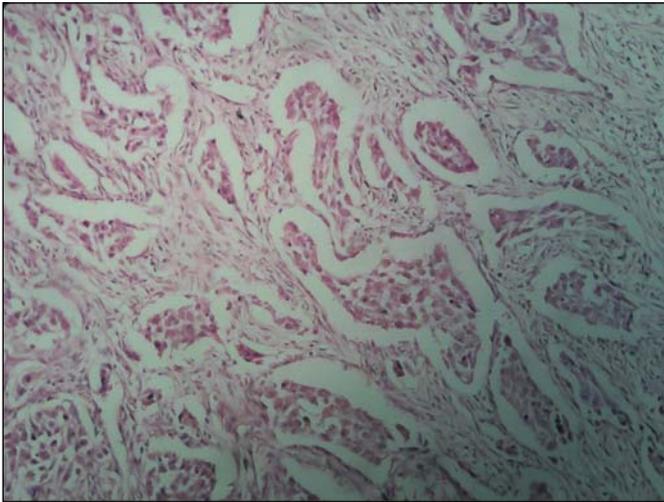


Fig. 4. Ameloblastic carcinoma exhibiting a follicular growth pattern with stellate reticulum-like structure or central necrosis. Peripheral columnar cells are palisaded in the follicular islands. (Hematoxylin-eosin stain; original magnification x100).



Fig. 5. Surgical specimen.

Majority of ameloblastic carcinoma originates *de novo*. Our case would fit into this category because the patient had no contributing medical history.

The remaining are malignant transformation of an ameloblastoma [8].

The clinical symptoms of ameloblastic carcinoma are more aggressive than ameloblastoma. Features distinct from ameloblastoma are swelling with rapid growth, perforation of the cortex, pain, tooth mobility, a nonhealing extraction site, ulcer or fistula, facial asymmetry, trismus and paresthesia [2].

The diagnosis and extent of these odontogenic tumours are facilitated by both a panorex and computerized axial



Fig. 6. Intraoperative view with mandibular reconstruction by fibula free flap.

tomography. Radiographically, most cases presented as an ill-defined destructive radiolucency with focal radiopacities. Perforation of the buccal and lingual plates also occurred together with evidence of root resorption. The lesions were aggressive, extending beyond bone into adjacent soft tissue.

Histologically, these lesions exhibited features of conventional ameloblastoma but the epithelium displayed cytologic atypia and an increased mitotic index [6]. Other features of malignancy include hyperchromatism, large, necrosis, and neural and vascular invasion. However, when ameloblastic carcinomas arise *de novo*, diagnosis is not as simple because differential diagnosis must include primary intraosseous squamous cell carcinoma, metastatic carcinoma to the jaw, central high-grade mucoepidermoid carcinoma, and bony invasion of carcinoma originating from the adjacent soft tissue.

The term ameloblastic carcinoma can be applied to our case, which showed focal histologic evidence of malignant disease.

The clinical course is reported as typically aggressive, with extensive local destruction. Regional nodal metastases and/or hematogenous dissemination may occur. Seventy five percent of distant metastases arise in the lungs, while the remaining lesions occur predominantly in the bones. The median survival after development of distant metastases is 2 years [9].

Because of the rarity of large clinical series and long-term follow-up, there is no consensus on treatment of ameloblastic carcinoma. Wide local excision is the treatment of choice. Monobloc excision of the tumour with 1 to 2cm of normal bone margin is the safest surgical approach to ensure disease-free survival. This method has resulted in local recurrence rates of less than 15% [10]. Cervical lymph node dissection should be considered even if there was no obvious lymphadenopathy.

Studies demonstrate a very high success rate, with good-to-excellent functional and aesthetic results using osseous free flaps for primary mandible reconstruction. The fibula donor site should be the first choice for most cases, particularly those with anterior or large bony defects requiring multiple osteotomies. Use of alternative donor sites (radius and scapula) is best reserved for cases with large soft-tissue and minimal bone requirements [11].

Radiotherapy has been suggested to decrease tumour size before surgery and to improve local control when surgical margins are close or microscopically positive [12].

Experience with chemotherapy is minimal in the treatment of ameloblastoma and is largely limited to isolated cases. However, in the setting of metastatic disease, Ramadas found the use of cisplatin, adriamycin, and cyclophosphamide to be beneficial [13].

Long-term follow-up is mandatory to detect the late recurrence, metastasis or regional lymph nodes. Therefore, diagnosis at an early stage, close periodic screening for metastasis and adequate treatment are necessary to improve patient prognosis.

Conflicts of interest

The authors declare that they have no conflicts of interest in relation to this article.

Authors contributions

The authors alone are responsible for writing the manuscript.

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