AMELOBLASTIC CARCINOMA: A CASE WITH CERVICAL NODE AND PULMONARY METASTASES

SP Khoo, ST Ong. Ameloblastic Carcinoma: A case with cervical node and pulmonary metastases, Annals Dent Univ Malaya 1998; 5: 49-52

ABSTRACT

Odontogenic carcinomas of the jaws are subclassified into malignant ameloblastoma, ameloblastic carcinoma and primary intraosseous carcinoma arising from within the bone. These may arise from residual islands of epithelium derived from dental lamina or epithelial lining of dental cysts. Ameloblastic carcinoma is extremely rare. An aggressive case of ameloblastic carcinoma occurnng in a 59-year-old Malay man is presented. Wide excision of the primary lesion with radical neck dissection was carried out. He developed lung metastasis 4 months post-operatively. Despite chemotherapy upon discovery of lung metastasis, he expired 7 months following the initial diagnosis.

Keywords: Ameloblastic carcinoma, aggressive lesion, neck nodes metastases, pulmonary metastasis, malignant ameloblastoma.

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INTRODUCTION

Ameloblastoma is considered the most common epithelial odontogenic neoplasm, representing about 1% of all oral tumours with 80% occurring in the mandible and 20% in the maxilla (1,2). The behaviour of ameloblastomas is essentially persistent, locally invasive but not malignant.

The current WHO classification of odontogenic carcinomas into three types namely, malignant ameloblastoma, primary intraosseous carcinoma, malignant variants of other odontogenic tumours and malignant changes occurring in odontogenic cysts remains unchanged from its original classification in 1972 (3,4). Elzay and coworkers (5) have proposed a subclassification of these malignant tumours and this in turn has been modified by Slootweg and coworkers (6). In this classification the term "ameloblastic carcinoma" is used to describe the tumour which demonstrates histological evidence of malignant transformation of the ameloblastoma-like epithelial component in the primary tumour whether or not it has metastasized.

Ameloblastic carcinoma generally occurs in the mandible affecting the average age group of 30 - 33 years with the lung being the commonest site for metastasis (6,7). The following report is a case of ameloblastic carcinoma with lung metastasis which developed 4 months after the initial diagnosis.

Case report:

A 59-year-old Malay man came to the Department of Oral & Maxillofacial Surgery with a complaint of swelling in the lower alveolus. He had first noticed the swelling about one month previously. The swelling had rapidly increased in size and had became painful for the past one week. There was no difficulty in eating and no noticeable weight loss.

His significant medical history included ischaemic heart disease and myocardial infarction two years previously. He had been taking antihypertensive medications which include Isordil, Adalat, Betaloc and aspirin.

Clinical examination revealed a fairly fit man with no other signs and symptoms elsewhere. There was a firm, indurated and ulcerated swelling on the right side of the chin (Figure 1). There was anaesthesia on the right mental region. Two right upper cervical lymph nodes were palpable.

Intra-oral examination showed a completely edentulous mouth. An ulcerated and proliferative lesion was

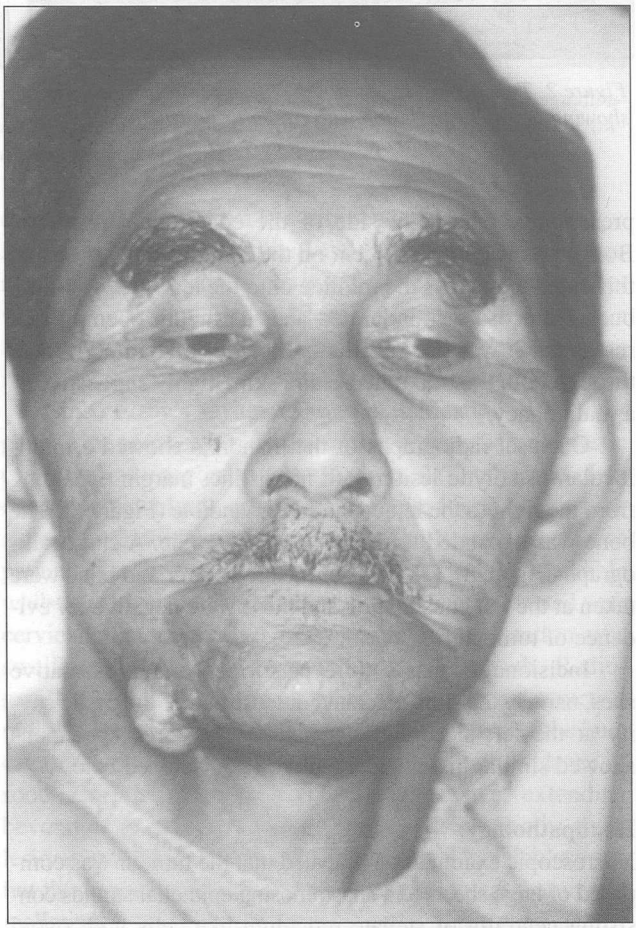

Figure 1 Extra-oral appearance of the lesion on presentation

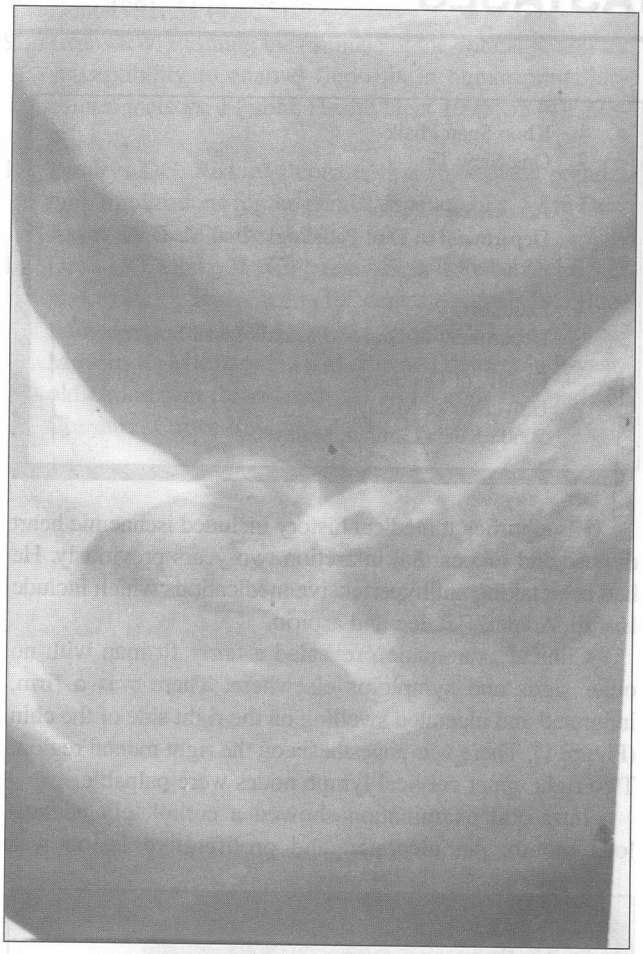

Figure 2 True occlusal view of the anterior portion of the lesion showing an osteolytic lesion with irregular destruction of the lingual and buccal cortices.

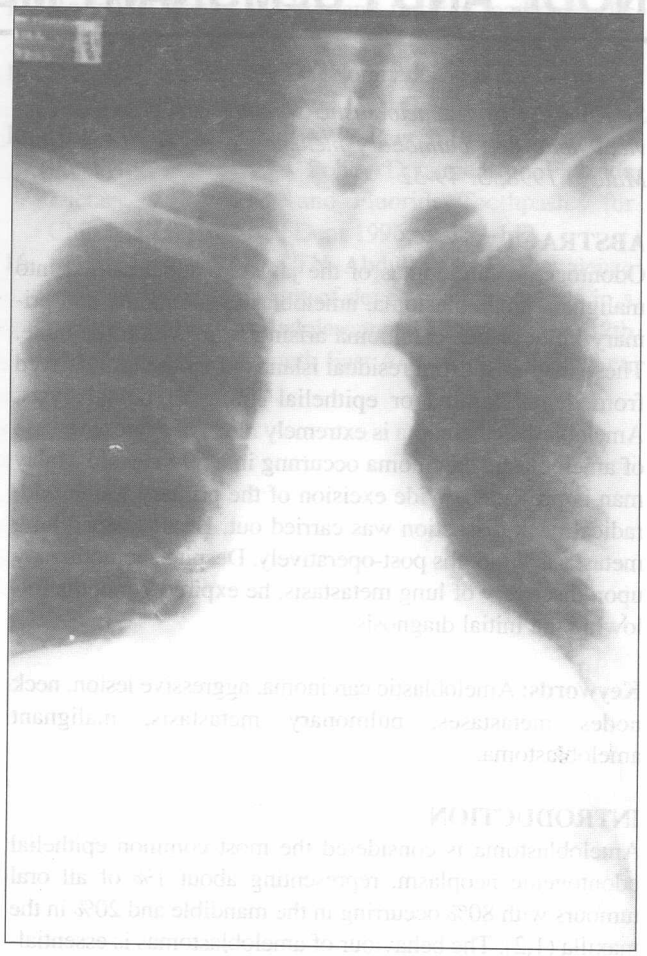

Figure 3 Pre-operative chest radiograph appearance.

present over the right side of the edentulous mandible. Bony expansion was evident on the right buccal region and this extended across the midline onto the left side. The right buccal mucosa was indurated and the mouth opening was restricted. It was obvious that a bony lesion had perforated through both the oral mucosa and skin, highly suggestive of a malignancy.

Occlusal radiographs of the mandible showed a multilocular, osteolytic lesion with no distinct margin extending from the right to the left edentulous mandible (Figure 2). The bone was expanded to the left of the midline. A chest radiograph (Figure 3), CT scans of the brain, liver and bone were taken at the frst consultation and these were negative for evidence of tumour.

Incisional biopsies were performed at representative sites, namely the skin, ulcerated mucosa and of the cystic cavity at the left side of the mandible. All three specimens showed similar histological features as discussed below.

Histopathology:

Microscopic examination revealed that the tumour was composed of large sheets and anastomosing epithelial strands consisting centrally of stellate reticulum-like cells with ovoid nuclei. Preameloblast-like cells were bordering the epithelial strands and the large sheets. The intervening stroma was slightly oedematous. In areas cellular and nuclear pleomorphism was evident. In such areas, the periphery of cell nests exhibited a columnar morphology. These cells contained pleomorphic nuclei with mitotic figures. Squamous metaplasia together with infiltrating well-differentiated squamous cell carcinoma islands were present in these areas. The tumour infiltrated diffusely into the surrounding soft tissues. (Figure 4).

Treatment:

Surgical excision with primary reconstruction of the mandibular defect was the mode of treatment chosen for this patient. Segmental resection from angle to angle of the mandible with wide excision of the involved mucosa and skin was carried out. A frozen section of the two right deep cervical lymph nodes showed infiltration by tumour. On the basis of this fnding, a right radical neck dissection was performed. A frozen section of the cervical lymph nodes showed tumour involvement and hence these were removed in the neck dissection. A 20cm length of fibula graft with the overlying skin was harvested from the left leg. The fibula graft was fixed to the remaining mandibular rami using miniplates. The vessels were anastomosed to the left facial artery and vein while the skin was used to provide both intra-oral cover and reconstruction of skin defect. The grafted site healed

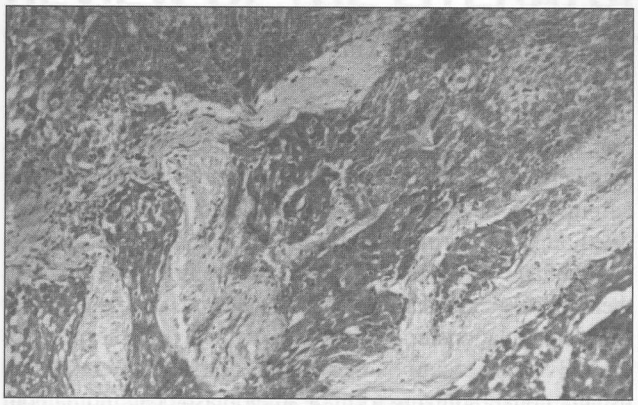

Figure 4 Photomicrograph showing islands of ameloblastic carcinoma infiltrating adjacent fibrous connective tissue stroma. Dedifferentiation within areas of conventional ameloblastoma is evident (H & E X 200).

without fistulation and there was satisfactory oral seal despite obvious facial defect. He was able to take soft diet and speech was acceptable.

Four months post-operatively, he was admitted for acute shortness of breath due to pneumonia. A chest radiograph taken showed patchy lung deposits (Figure 7), most probably representing secondaries from malignant ameloblastoma of the mandible. The patient refused a biopsy of the lung lesion. He was treated by the physician in another hospital but the patient passed away two months later. There was also no consent for an autopsy.

DISCUSSION

Carcinomas derived from ameloblastomas have been given many designations such as malignant ameloblastoma (1), metastatic carcinoma (6) and primary intra-alveolar epidermoid carcinoma (8). In the most recent classification of odontogenic tumours by the World Health Organisation, malignant ameloblastoma is clearly defined as "a neoplasm in which the pattern of an ameloblastoma and cytological features of malignancy are shown by the primary growth in the jaws and/or by any metastatic growth"(3).

Slootweg and Muller (6) subclassifed 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 ex 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.

As a result of their subclassification, malignant ameloblastoma and ameloblastic carcinoma were distinguished from each other. The former is a term which should be reserved for those lesions that, in spite of a seemingly innocuous histology, have given rise to metastatic growth. The latter however, is a term for lesions that combine features

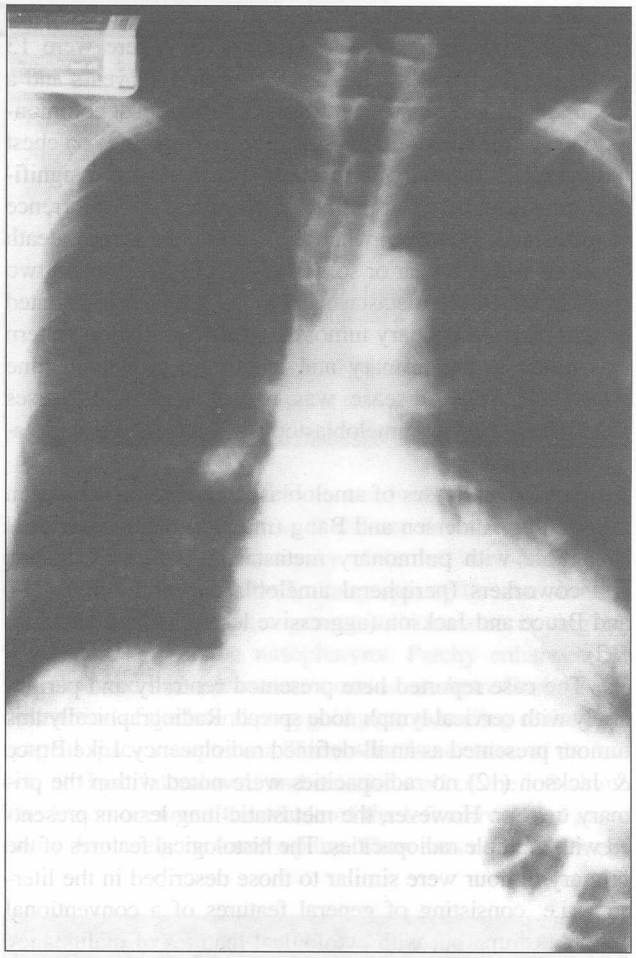

Figure 5 Chest radiograph 4 months post-resection of the mandibular lesion. Bilateral pleural effusion with hazy opacities (arrowheads), possibly representing metastases.

of ameloblastoma with a less-differentiated histomorphology. Accordingly hence, this case could be described as malignant ameloblastoma according to the WHO classification (3) but would fit into Slootweg and Muller's (6) classification as a type 2B odontogenic carcinoma (although evidence of lung metastases could not be obtained in this patient).

Since Elzay's review (5) and classification of odontogenic carcinoma, other workers have reported arneloblastic carcinoma as a separate entity. Corio et al (7) reviewed eight cases of ameloblastic carcinoma and found four cases which appear to arise from a cyst lining. The mean age ofthese patients was 30.1 years. Seven cases involved the mandible whilst one involved the maxilla. Only one case presented with cervical metastases. The most common presentation was swelling, followed by pain and/or rapid growth. Radiographicallv, 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 various cytological features of malignancy. The five year survival rate was not presented by these authors.

Slootweg and Muller studied all cases in the literature reported as malignant ameloblastoma (6). There were 13 males and 10 females with ages between 4-75 years and a mean age of 33.5 years. Metastasis occurred most commonly to the lung. These lesions appear as radiopacities on chest radiographs. Generally, it appeared that there was a significant time span between initial treatment and the occurrence of metastasis. However, when the disease had spread, death occurred within a year or so. Histologically, they found two cases in which the metastasis exhibited a less differentiated pattern than the primary tumour. A dedifferentiation pattern was noted in the primary and metastatic growth in nine cases. Metastatic disease was not present in 14 cases although the primary ameloblastoma had undergone anaplastic transformation.

Subsequent cases of ameloblastic carcinoma have been reported by Andersen and Bang (maxilla) (9), Dorner et al (mandible with pulmonary metastasis) (10), McClatchey and coworkers (peripheral ameloblastic carcinoma) (11), and Bruce and Jackson (aggressive lesion in the mandible) (12).

The case reported here presented centrally and peripherally with cervical lymph node spread. Radiographically this tumour presented as an ill-defiined radiolucency. Like Bruce & Jackson (12) no radiopacities were noted within the primary tumour. However, the metastatic lung lesions presented with multiple radiopacities. The histological features of the primary tumour were similar to those described in the literature i.e. consisting of general features of a conventional ameloblastoma but with cytological features of malignancy ie. ameloblastic carcinoma.

The clinical course of ameloblastic carcinoma is typically aggressive with extensive local destruction. Local recurrences are common and so are metastases to the neck and lung (1, 6,7). The clinical course of this case was more aggressive than the cases described in the literature. The patient possibly had lung metastasis three months post-operatively and from the point of initial diagnosis the patient survived only seven months despite radical surgery and chemotherapy. Corio and coworkers (7) reported three of seven cases which had recurrences within a year. Slootweg and Muller (6) noted that cases which showed dedifferentiation in both the primary and metastatic growth died with two years after metastasis. More recently, Bruce & Jackson(12) reported an aggressive case with early metastasis to the left lung.

There is ample evidence in the literature that so-called "benign" ameloblastoma can dedifferentiate with time and multiple inadequate surgical procedures are associated with metastasis(l2). The length of time taken for dedifferentiation of the tumour is unknown. The usefulness of radical neck surgery in the management of ameloblastic carcinoma remains debatable. Radiotherapy appears to be of limited value for ameloblastic carcinomas and chemotherapy is yet unproven (12). From a clinical viewpoint, ameloblastic carcinoma is an extremely aggressive neoplasm and any mode of treatment chosen should be aimed at preventing recurrences.

Histogenetically, ameloblastic carcinoma like other odontogenic carcinomas, may arise de novo from remnants of the dental lamina or from odontogenic cysts and may be located centrally in the jaws or peripherally in the surrounding mucosa (3,13). However, little is known of the relationship between the different histologic patterns of odontogenic carcinomas and their prognosis. Corio et al (7) stated: "Although the primary intra-alveolar carcinoma and the ameloblastic carcinoma exhibit some clinical differences, their histological features are similar enough to suggest a histogenetic relationship. It is possible then that the primary intra-alveolar carcinoma may represent a less differentiated, usually nonkeratinizing form of ameloblastic carcinoma both lesions being derived basically from odontogenic epithelial remnants".

In conclusion therefore, due to the rarity of this tumour and paucity of knowledge of its relationship between the histological patterns and its clinical course, it is rational to treat these lesions aggressively with wide excision.

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