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Case Report

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A Rare Case of Ameloblastic Carcinoma Presenting as a Nasal Mass

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Abstract

The ameloblastic carcinoma is a rare, malignant odontogenic tumour that can behave aggressively with both local tissue invasion and distant metastases. Although the mandible is most commonly affected, others areas of the maxillofacial skeleton can be infrequently involved. This case report describes a rare case of an ameloblastic carcinoma affecting the nasal region in a 34 year-old female who presented with a longstanding swelling of the left nasal cavity. Treatment involved total rhinectomy and post-operative chemo radiotherapy, however 18 months later she developed pulmonary metastasis.

Keywords: Ameloblastic carcinoma, Malignant odontogenic tumour

Abbreviations: AC: Ameloblastic Carcinoma; WHO: The World Health Organisation; MRI: Magnetic Resonance Imaging; SCC: Squamous Cell Carcinoma; CT: Computed Tomography

Introduction

The ameloblastic carcinoma (AC) is a true oral malignancy with classic features of cancer including cytological atypia, recurrence, and metastatic spread into both regional and distant sites [1]. It is a rare odontogenic tumour that may arise de novo or from a pre-existing ameloblastoma. Chaisuparat and colleagues [2] showed that it is the most common malignant odontogenic tumour (29%), and can make up 10.5% of all ameloblastomas [2]. It is more likely to affect an older age of patients (mean 50 years) compared to the solid-multicystic or unicystic benign ameloblastomas. Males are affected more than females at a ratio of approximately 2.5:1 [2], with the mandible being affected more than the maxilla. The World Health Organisation (WHO) [3] has further sub classified the AC into 3 subtypes:

- Ameloblastic carcinoma – primary type
- Ameloblastic carcinoma: secondary type (dedifferentiated) intraosseous
- Ameloblastic carcinoma: secondary type (dedifferentiated) peripheral

The diagnosis of AC-secondary type (dedifferentiated) is made when a previous diagnosis of a 'typical' ameloblastoma undergoes carcinomatous change. This includes both 'typical' intraosseous and peripheral (extraosseous) benign ameloblastomas [3]. Some consider that the gingival 'basal cell carcinoma' may in fact be a dedifferentiated AC - peripheral type, and further affirms the suggestion that the cutaneous basal cell carcinoma

and intraoral ameloblastoma could be related [4].

Here we discuss the case of an AC primary type in the nasal region in a 34 year-old female. To our knowledge, this location has not been described previously in the literature.

Case Presentation

A 34 year-old lady was referred to the Maxillofacial Surgery, Head and Neck Department with an unusual longstanding swelling on the left side of her nose (Figure 1). This mass had gradually increased in size over 2 years and became more uncomfortable. There were no underlying medical issues, with no history of smoking or excessive alcohol intake. Examination revealed a firm swelling approximately 2 cm in diameter, of the left lateral nasal cartilage with distortion of the alar and erythema and crusting of the overlying skin. Initial soft tissue biopsies from a different institution suggested inflammatory change, but no malignancy was identified. 9 months later she developed a left level I neck mass which was mobile, but firm to palpation.

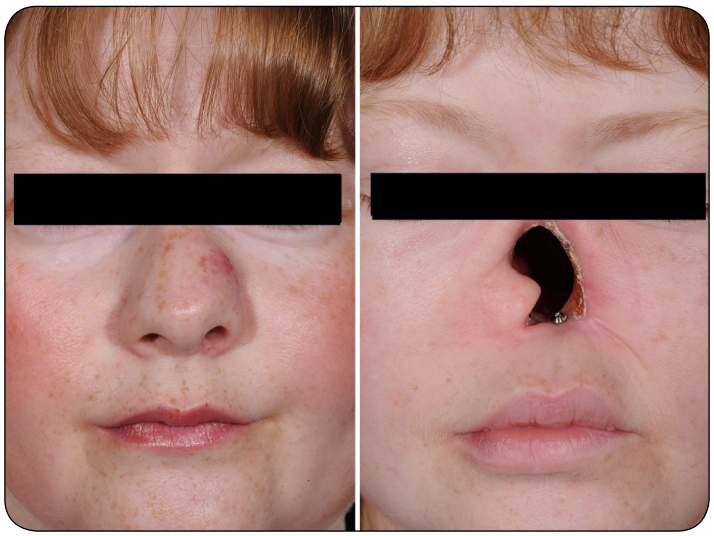

Figure 1: Pre-operative and 1 year post-operative photographs.

Magnetic resonance imaging (MRI) was used to identify the size and extent of the lesion (Figure 2). This showed a 2 cm diameter soft tissue mass on the left side of her nose in the subcutaneous tissue extending anteriorly to the skin. A left level 1B pathological lymph node mass measuring 28 mm with central necrosis and evidence of extracapsular spread.

This was confirmed on ultrasound scan and cytology of a fine needle aspiration from the node demonstrated a necrotic undifferentiated malignant process. A further incisional biopsy of the nasal mass was undertaken and histopathology suggested moderately differentiated squamous cell carcinoma (SCC). A computed tomography (CT) scan of the chest was used to complete her staging, and showed no evidence of pulmonary metastasis. She was staged cT3N2bM0 SCC of the left nasal cavity with extra capsular spread in left level 1 b neck node.

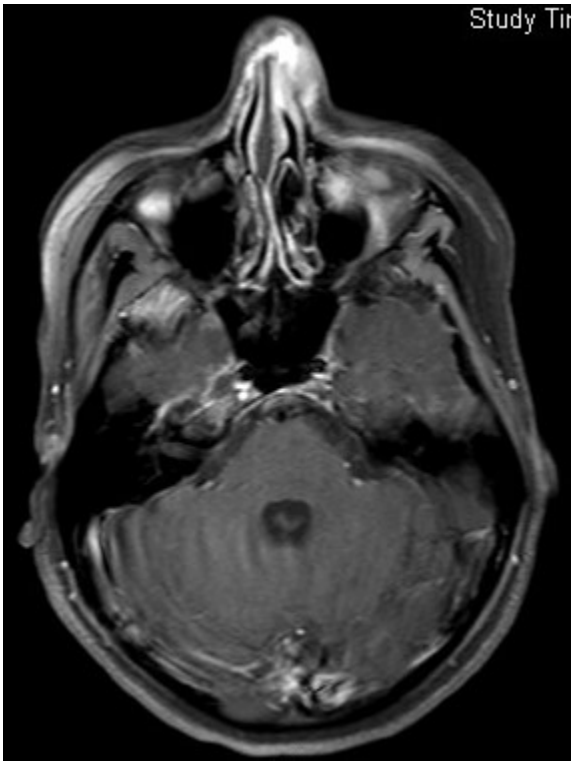

Figure 2: MRI scan showing left nasal mass.

After a multidisciplinary team discussion, she underwent a total rhinectomy, bilateral level I-IV selective neck dissection and left total parotidectomy due to the close proximity of nodal neck metastasis to the parotid and due to the potential for metastasis to the parotid.

Histological examination of the primary lesion demonstrated an ameloblastic carcinoma (AC) with areas of squamous differentiation (Figure 3). The left neck dissection showed a solitary 3 cm level I extracapsular AC metastatic lymph node, and no nodes were involved in the right neck dissection specimen. Initial margins were positive and she therefore underwent further re-excision and the margins were later confirmed to be clear. She had a staged reconstruction with the placement of titanium implants for a prosthetic rehabilitation. Post-operative chemo-radiotherapy with 60 Gray in 30 fractions and 2 cycles of Cisplatin at 100 mg/m^2 and 20% dose reduction for the second cycle due to the severity of the symptoms,

to the nasal region and left neck was completed. She made a good recovery and was followed up monthly, with surveillance scans at 6 and 12 months post-operatively showing no signs of recurrence.

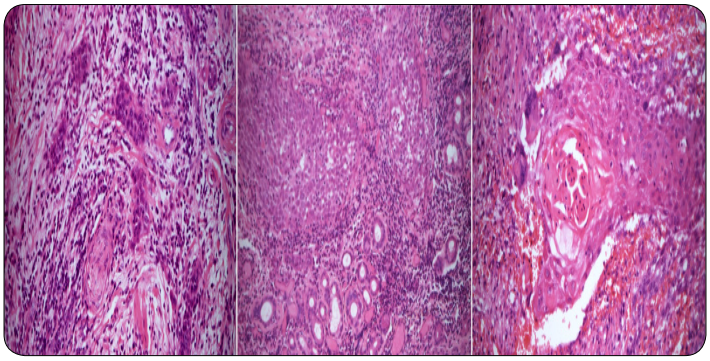

Figure 3: Histological images.

18 months following her initial surgery, she reported a chronic cough, shortness of breath and haemoptysis. Unfortunately, a chest CT scan showed left malignant pleural effusion with collapse of her lung, particularly affecting the upper lobe. Cytology of the pleural aspirate identified metastatic malignant cells with appearances similar to the primary ameloblastic carcinoma. She had a left intercostal chest drain placed prior to undergoing palliative chemotherapy with Carboplatin AUC 5 and Capecitabine at a 70% dose. Although she did not have evidence of loco-regional recurrence, she continued to suffer from shortness of breath, chest pain and a recurrent left sided pleural effusion and subsequently required an indwelling pleural catheter, 20 months after her initial surgery. A follow-up CT scan showed left lower lobe consolidation, right sided pleural effusion, bone metastases to the spine and pelvis, and a possibility of liver metastasis. She sadly passed away 23 months after her initial surgery.

Discussion

Ameloblastic carcinomas (AC) account for less than 2% of all odontogenic tumours, and are one of the two malignant variants of the more common benign ameloblastoma. Although the AC has both true clinical and histological features consistent with a carcinoma, the other malignant variant (the malignant ameloblastoma) has benign cytological appearances but can metastasise to regional or distant sites [3]. The majority of AC occur de novo, however they can also arise from pre-existing ameloblastomas.

Embryological development of the sinonasal tract is closely related to the development of odontogenic apparatus. The oral cavity and sinonasal tract are separated during intrauterine development. Due to the close relation between the development of these structures, an odontogenic tumour such as the AC, can present in the nasal structures [5]. This may occur if the odontogenic epithelium becomes trapped in the sinonasal mucosa or if the sinonasal cells develop odontogenic potential. The presence of ectopic teeth in the nasal cavity may also lead to the development of odontogenic tumours in the nasal cavity [5], although this was not exhibited in this case and there was no involvement of the maxilla.

Histological features include cellular atypia, hyperchromatism, and hypercellularity, in combination with the 'typical' ameloblastoma patterns and cellular features. Unlike the 'malignant' ameloblastoma subtype, metastasis is not required for the diagnosis of an AC to be made [5]. Areas of tumour may undergo necrosis or show perineural invasion, and overall the lesion may show a high proliferation index compared to benign ameloblastomas [6]. Although these features maybe present, a true microscopic distinction from a benign ameloblastoma is difficult and the diagnosis maybe more subjective [6] than the diagnoses of other oral malignancies.

The commonest signs of AC include pain, swelling, trismus, rapid growth and dysphonia. Hall et al. in 2007 suggested that the AC may produce a distinct set of signs of symptoms than benign ameloblastomas including rapid growth, perforation of cortical bone, pain, and paresthesia [7]. Other common findings are non-resolving swelling, tooth mobility, and ulceration from direct tumour extension [2]. The presence of these features can be correlated with radiological and histological findings to confirm the diagnosis. Approximately 50% of the lesions are well-defined radiolucencies, and they may take a unilocular or multilocular appearance. A minority may cause dental root resorption and they can be related to impacted teeth [2].

The progression of this aggressive tumour with extensive local destruction and metastatic spread results in a poor long-term prognosis, however treatment modalities for such patients are controversial as there is no clear consensus on the best approach to manage this condition [5].

Radical surgical intervention is necessary to avoid local recurrence, but this is dependent on the size and location of the lesion as well as patient factors (e.g. medical co-morbidities). Neck dissection should be strongly considered even in the absence of nodal involvement, especially if free-flap reconstruction is planned. The reported use of

radiotherapy and chemotherapy as an adjuvant treatment modality is conflicting [6], and their effectiveness is not well documented in the literature due to the rare incidence of AC. Radiotherapy can assist in preventing loco-regional recurrence, especially in the presence of close or positive margins [8], however the role of chemotherapy is more unclear. Novel approaches include the use of carbon ion therapy [9], Gamma Knife stereotactic radio surgery [10] and BRAF V600E targeted therapies [11], although they are yet to reach international acceptance.

Vigilant post-operative surveillance is required for the early detection of recurrence and metastatic spread, however distant metastases can occur even in the absence of loco-regional disease.

The most common site for distant metastases is the lung, followed by bone, liver and brain [5]. Any signs or symptoms of lesions affecting these areas on routine follow-up should be investigated thoroughly and appropriate multi-disciplinary treatment should be undertaken.

Conclusion

AC is a rare odontogenic tumour with a high potential for recurrence and metastasis. The present study reports the case of a 34 year old female with an extremely rare nasal AC that was managed by radical surgery and adjuvant chemo-radiotherapy. Despite close follow up, distant metastasis to the lung was detected. Additional studies and case reports can assist in expanding knowledge about this tumour and assist in the treatment and management strategies of such cases.

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Declarations

Ethics approval was not required and consent was obtained for publication.

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