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Case Report

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A Rare Tumour, a Rare Site and a Rare Presentation - "The Nasal Apricot"; Seeing Ameloblastic Carcinoma in a New Light

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ABSTRACT

Ameloblastic Carcinoma (AC) is a rare malignant tumor that usually arises from the bones of the jaw. Out of approximately 120 reported cases in literature to date, only about 5 cases have been reported at extra-gnathic sites. The mandible is the most common site for the development of ameloblastic carcinoma. Less frequently, the maxilla is the primary tumor site. In one reported case, the primary site was the anterior skull base. We report a rare presentation of ameloblastic carcinoma primarily arising from the nasal cavity, which, as far as we know is the first of its kind as reported in literature.

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Introduction

Ameloblastic carcinoma is usually an odontogenic tumour, and the pattern of epithelial growth is similar to developing tooth germ cells [1].

They are often aggressive and nasty, metastasise to other areas of the body, especially the lungs, bone, brain, and liver.In the 2005 WHO classification of odontogenic tumors,ameloblastic carcinomas were divided into three categories [2].

(a) Primary Type: The primary type demonstrates malignancy in the primary tumour with characteristics of ameloblastoma and cytologic atypia.
(b) Secondary Type (Dedifferentiated) Intraosseous
(c) Secondary Type (Dedifferentiated) Peripheral

The secondary type comprises of malignant variations, which originate in a previously existing ameloblastoma, irrespective of the presence or absence of metastasis. The secondary type of ameloblastic carcinoma can be further classified into two subtypes. The intraosseous type occurs within a pre-existing benign intraosseous ameloblastoma, and the peripheral type occurs within a pre-existing benign peripheral ameloblastoma.

These tumours are presently classified under 'ameloblastic carcinoma' based on the morphologic continuum and analogous behavior between these entities have reported that ameloblastic carcinoma possess unique histopathological features [3]. At the early stages of malignancy or dedifferentiation, epithelial tumour nests and islands surrounded by a layer of stellate basaloid cells are observed in the mesenchymal tissue. Recognised that these cells exhibit malignant features, such as cellular pleomorphism, mitoses, focal necrosis, perineurial invasion and nuclear hyperchromatism [4]. Besides, ameloblastic carcinoma display histological features of ameloblastoma and carcinoma.

The exact cause of AC is still unknown, but researchers speculate genetic and immunological abnormalities, environmental factors like UV rays, chemicals, ionising radiation, diet, stress etc [5].

A few individuals with ameloblastic carcinoma remain asymptomatic [6]. Symptoms that may occur include

- Progressive pain and swelling of the jaw.
- Bleeding and headaches may also occur.
- inability to open the mouth (trismus)
- dysphonia, a voice disorder characterized by hoarseness, weakness,
- tingling or numbness (paresthesia), and, in rare cases,
- Voice loss (my note: disruption of the voice because of laryngeal involvement (vocal cords) would be quite unusual but dysphagia, trouble eating, can be a problem if the tumor is large enough to cause obstruction or malocclusion).
- Nasal discharge and blockage of the nasal passages may occur if the tumor involves the sinuses of the maxilla.
- The size of the tumor may result in dental abnormalities such as causing the upper and lower teeth to fail to meet properly (malocclusion).
- The most common course of the disease is persistent recurrence with local spread.

This case report is presented due to the rarity of this disease and its site of origin.

Case Report

A 60-year-old male arrived at the otolaryngology department on 4th May 2022 with complaints of five episodes of nasal bleeding from the left nostril, on and off for the past 2 months. He stated symptoms of left sided nasal obstruction along with foreign body sensation in the left nostril. There was no pain, dyspnea, or olfactory complaint at the time of presentation. He had received a course of antibiotics prior to presentation.

Physical examination demonstrated obliteration of nasomaxillary groove on the left side, flaring of the left ala and multiple reddish pedunculated polyploidal mass of tissue protruding through the left nasal cavity (Figure.1), which was firm in consistency and did not bleed on touch. Cranial nerves were clinically intact.

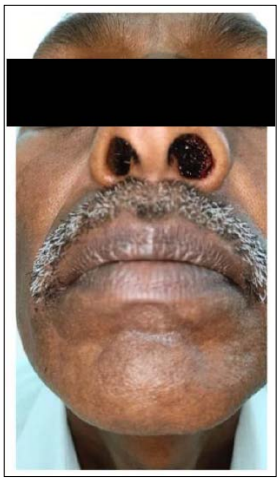

Figure 1: Polyploidal mass protruding through the left nasal cavity

A rigid nasal endoscopy guided biopsy was taken and sent for histopathological examination, which revealed areas of haemorrhage with scattered sheets and nests of malignant epithelial cells with moderate eosinophilic cytoplasm and hyper-chromatic pleomorphic nuclei suggestive of malignancy. Pathological analysis of the biopsy was suggestive of sinonasal carcinoma. Diagnostic work up proceeded with a non-contrast CT imaging which showed a polyploidal growth of size 4.5*3*2 cm arising from the left nasal septum with features of bilateral maxillary, ethmoid and sphenoid sinusitis (Figure. 2,3).

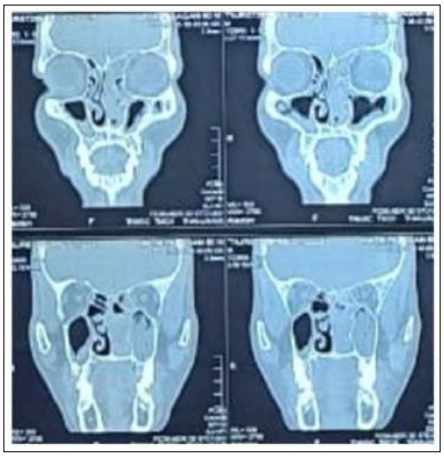

Figure 2: CT (coronal view); showing hyper dense lesion arising in left nasal cavity with features of sinusitis. The figure shows no evidence of bony erosion.

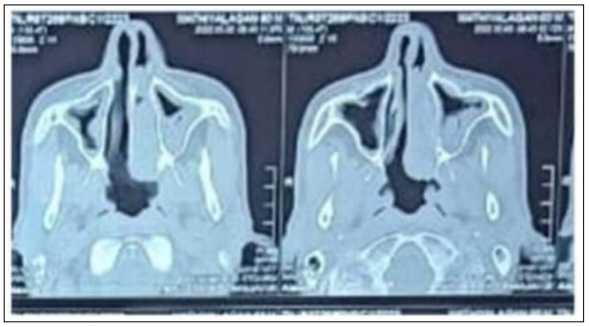

Figure 3: CT (axial view); showing hyper dense lesion in left nasal cavity of dimension 4.4*3*2 cm

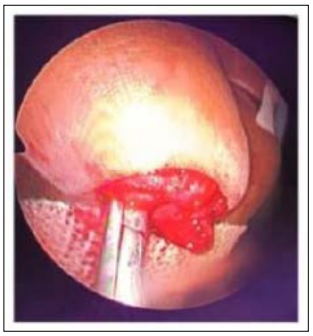

Figure 4: DNE BIOPSY

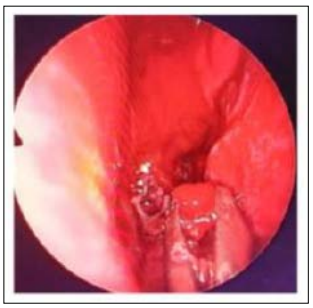

Figure 5: Microdebrider assisted Endoscopic Excision

A surgery involving debulking of the tumor with posterior septectomy and functional endoscopic sinus surgery of left nasal cavity was performed. The tumors was found to arise from the posterior end of nasal cavity and was removed into to (Figure. 4, 5).

About 3ml of mucopurulent discharge was also drained from the frontal sinus.

The tissue was then sent for histopathological examination and the results were consistent with Ameloblastic Carcinoma (Figure. 6, 7, 8).

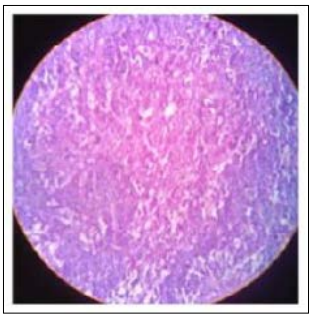

Figure 6: Scanner view shows a neoplasm composed of islands of epithelial cells with peripheral palisaded columnar epithelium

Immunohistochemistry was positive for Ki67 which is indicative of Ameloblastic Carcinoma.

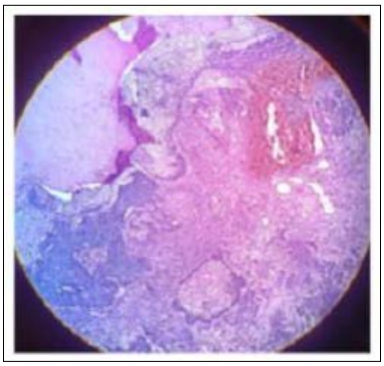

Figure 7: 10x view shows Peripheral palisaded columnar epithelium exhibiting reversal of polarity and stellar reticular stroma. Some islands show squamous Metaplasia

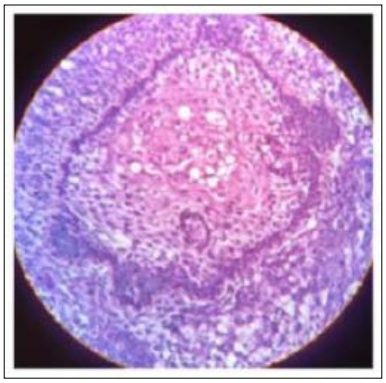

Figure 8: 40x high power view shows Areas in the islands composed of epithelial cells replaced by solid sheets of neoplastic cells exhibiting prominent nuclear atypia.

Discussion

The incidence of AC is difficult to ascertain because of the minuscule number of cases reported in literature, a fraction of which involve Extragnathic sites.

A primary AC arising from the nasal cavity, more accurately the nasal septum would typically be mistaken for a sinonasal carcinoma, and the diagnosis can be confirmed by imaging, histopathological examination and immunohistochemistry [7].

Studies have shown that ameloblastic carcinoma is particularly sensitive to FDG uptake on a PET scan and till date remains the best mode of diagnosing AC [8].

This patient's diagnostic work up consisted of a CT, histopathology and immunohistochemistry which was consistent with AC. There is no proven or established consensus on treatment of AC due to the low occurrence of this disease, although wide local excision is the basis of therapy for the primary tumor.

Usually, some combination of chemotherapy and radiation is used in the treatment of local extension and metastases. In rare or uncommon cases, neoadjuvant radiation or chemotherapy is adopted to shrink a tumor prior to resection.

Radiation therapy in itself can be effective but is reserved specifically for local recurrences or cases where the primary tumor may not be surgically resected.

In cases of systemic metastases, chemotherapy may be suggested, although studies have demonstrated mixed results. Targeted therapy focused at genomic aberrations is experimental.

Conclusion

AC is a rare odontogenic malignancy that is often discovered late in its course and therefore carries a poor prognosis.

Due to its low occurrence or incidence, there is little existing knowledge in its diagnosis and management.

Most primary neoplasms are found in the mandible or maxilla; but have since then discovered that they are not the only sites from which Ameloblastic carcinoma can originate.

The existing information regarding AC is limited due to the rarity of the disease and the prognosis for this patient is yet to be established. Follow up will be done with sessions of chemotherapy and through this paper, clinicians can become aware to suspect the occurrence of this disease in extra gnathic sites and intervene early and provide better care for the patient.

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