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Case Report

Pathology Section

Rare Case of Maxillary Ameloblastoma: A Challenging Entity

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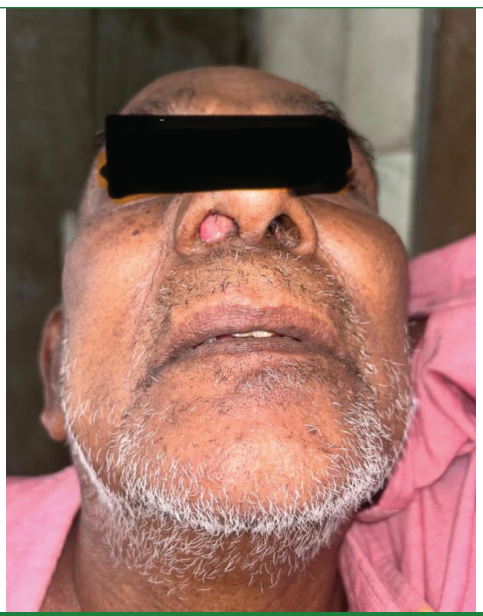
ABSTRACT

Maxillary ameloblastoma is a rare benign, locally aggressive extragnathic sinonasal tract epithelial odontogenic neoplasm that arises from remnants of odontogenic epithelium, the lining of odontogenic cysts, basal cells of the oral mucosa, rests of the dental lamina, or heterotopic embryonic organ epithelium. It has an unusual incidence of 0.5 cases per million person-years worldwide, with no gender predilection, and is most commonly found in individuals in their 6th to 7th decades of life. In the present case report, the authors presented a case of a 77-year-old male who presented with a history of right-sided nasal obstruction for 15 days. Anterior rhinoscopic examination revealed a soft-tissue nasal mass covering the entire nasal cavity, protruding as a nasal polyp, while posterior rhinoscopy examination was found to be normal. A Computed Tomography (CT) scan of the paranasal sinuses showed evidence of a soft-tissue density mass with internal hyperdensities noted in the right maxillary sinus. The patient underwent endoscopic medial maxillectomy, and gross examination revealed multiple grey-brown soft-tissue fragments with haemorrhagic areas. Microscopic analysis showed a neoplasm arranged in cords lined by columnar epithelial cells exhibiting reversal of polarity, with a central stellate reticulum. No bony fragments, atypical mitosis, or necrosis were identified. The diagnosis was established through histological analysis. Although maxillary ameloblastoma is a benign tumour, it requires aggressive treatment to prevent recurrence and local invasion. Early diagnosis using histopathological and radiological methods is crucial for optimal treatment planning and follow-up of the patient. The present case is presented due to its rarity and the diagnostic challenges it poses, including the lack of definite clinical presentation and radiological dilemmas. It also aimed to examine the key features of ameloblastoma in the maxillary region, emphasising their clinical, radiological and histological characteristics.

Keywords: Epithelial odontogenic neoplasm, Nasal mass, Stellate reticulum

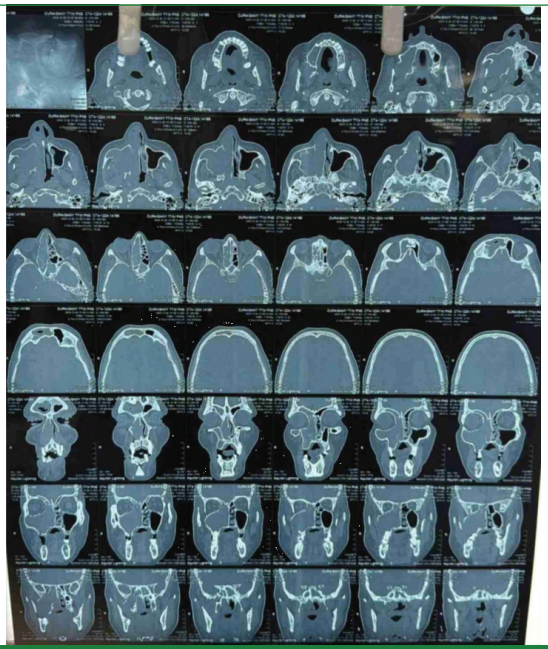
CASE REPORT

A 77-year-old male came to the Outpatient Department (OPD) with the complaint of nasal obstruction for 15 days. The patient was apparently normal until 15 days ago, when he developed right-sided nasal obstruction that was insidious in onset, gradually worsening to severe obstruction over the past five days. The patient had no history of epistaxis, nasal discharge, or postnasal drip, and there was no history of similar complaints in the past. On local examination, swelling of the right nasolabial fold was noted, and anterior rhinoscopy revealed a nasal mass that covered the entire nasal cavity and protruded as a polyp, as shown in [Table/Fig-1]. The posterior rhinoscopic examination was found to be normal.



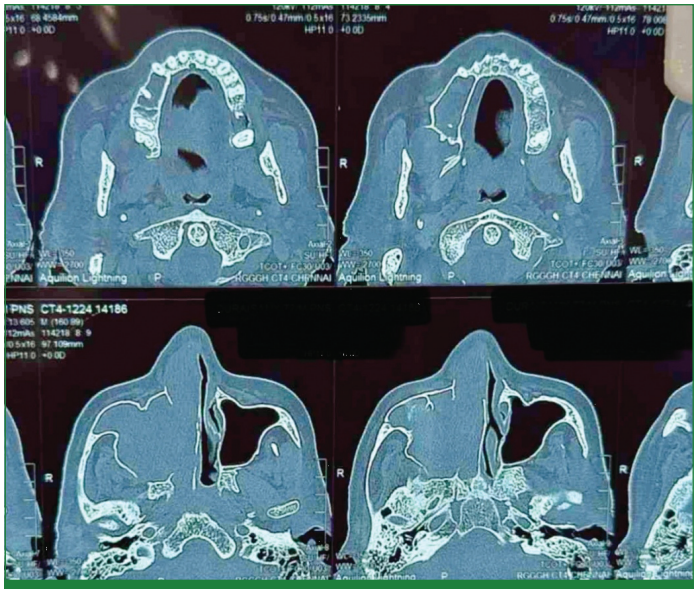
[Table/Fig-1]: A patient with a right-side nasal polyp.

Laboratory investigations, including complete blood count, liver function tests, renal function tests, serum electrolyte profile, lipid profile and coagulation profile, were all within normal ranges. A CT scan of the paranasal sinuses showed evidence of a soft-tissue density mass with internal hyperdensities noted in the right maxillary sinus with expansion of the ostium and extension into the right nasal cavity. It was also seen involving the right ethmoid sinus, frontal sinus, and sphenoid sinus. The mass caused bony erosion of the right turbinate, medial wall and posterolateral wall of the maxilla, extending into the retromaxillary space, as shown in [Table/Fig-2]. The radiological diagnosis indicated a sinonasal mass



[Table/Fig-2]: Evidence of soft-tissue density mass in right maxillary sinus.

with a possibility of invasive fungal sinusitis. Additionally, polypoidal mucosal thickening was observed in the left maxillary sinus, as seen in [Table/Fig-3]. Bony erosion of the medial wall of the orbit with intraorbital extension was also noted. The final impression was a possibility of a right sinonasal mass suggestive of invasive fungal sinusitis.

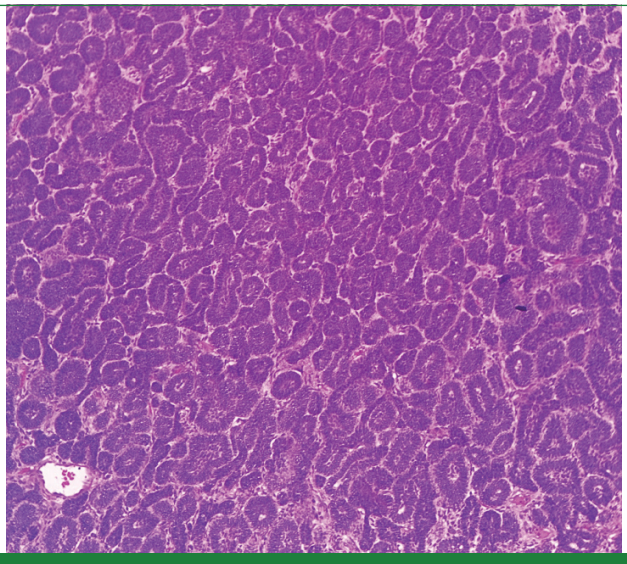


[Table/Fig-3]: Evidence of bony erosion of medial wall of orbit.

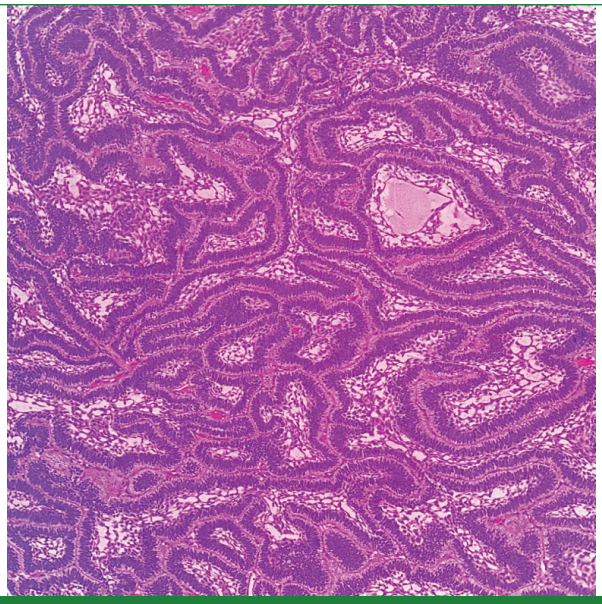
The clinical diagnosis of invasive fungal sinusitis or inverted papilloma was made, and the patient was subsequently proceeded to endoscopic medial maxillectomy.

Gross examination of the specimen revealed multiple grey-brown soft-tissue fragments, the largest measuring 3.5×1.8×0.9 cm and the smallest measuring 0.2 cm in diameter. No bony fragments were found. The external surface appeared congested, while the cut surface displayed grey-black and haemorrhagic areas.

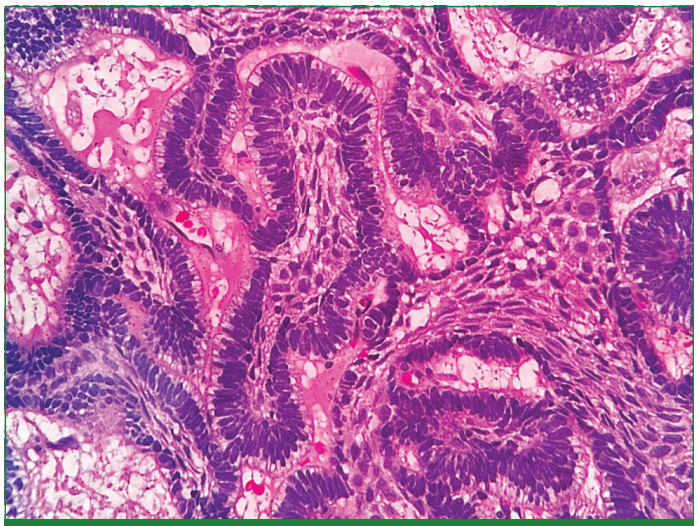
Microscopic examination revealed respiratory epithelium with an underlying neoplasm arranged in nests, follicles and cords of columnar epithelial cells exhibiting reversal of polarity and central stellate reticulum with vessels. A tubular pattern with nuclear crowding and pseudostratification was also observed, as shown in [Table/Fig-4-7]. The stroma displayed scattered lymphoplasmacytic infiltrate, histiocytes with foamy cytoplasm, cholesterol clefts and congested blood vessels. No bony fragments, atypical mitoses, or necrosis were identified in the multiple sections studied. Fungal stains were not performed, as the morphology favoured the classical diagnosis.



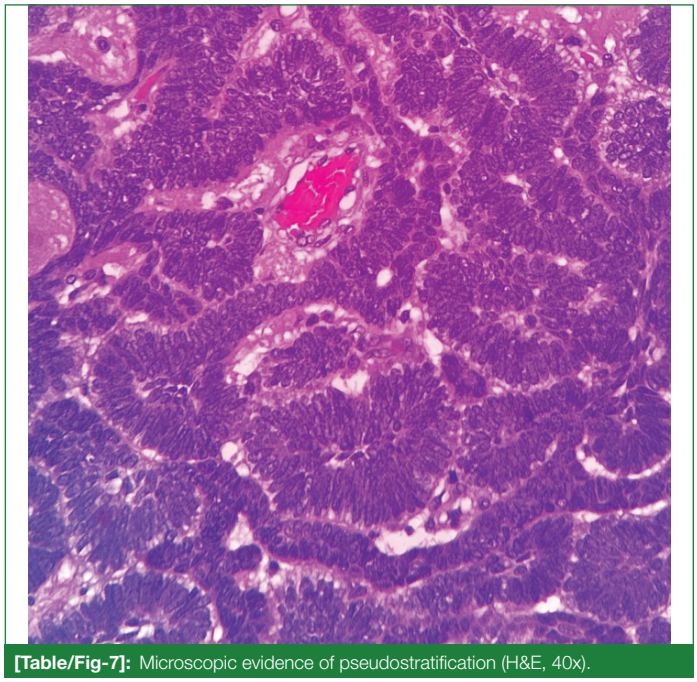
[Table/Fig-4]: Microscopic evidence of follicular and nested arrangement of cells (H&E, 4x).



[Table/Fig-5]: Microscopic evidence of arrangement of cells in anastomosing cords pattern (H&E, 10x).



[Table/Fig-6]: Microscopic evidence of stellate reticulum and reversal of polarity (H&E, 10x).



[Table/Fig-7]: Microscopic evidence of pseudostratification (H&E, 40x).

Based on clinical, radiological and histomorphological findings, the case was diagnosed as a benign odontogenic neoplasm-ameloblastoma of the maxillary sinus. After the procedure, the

patient was treated with appropriate antibiotics, and postoperative care was provided, with follow-up by the clinician.

DISCUSSION

The term 'ameloblastoma' was first described by Cusack JW in 1827 [1]. Later, it was called 'adamantinoma' by Malassez L in 1885; this term is currently used for a bone tumour by Fisher in 1913 [2]. Ameloblastoma is derived from the English word 'amel', which means enamel, and the Greek word 'blastos', which means germ [3]. It arises from the odontogenic epithelium, primarily from enamel that has not undergone differentiation into hard tissue formation [4]. The incidence is 0.5 cases per million worldwide, although there is an increase in incidence in South Africa [5]. There is no gender predilection for this disease.

Ameloblastoma is an unusual benign entity that is locally aggressive and has a tendency to recur. It arises from the odontogenic epithelium, including rests of dental lamina, a developing enamel organ, the lining of odontogenic cysts and the basal layer of oral mucosa. It is the second most common odontogenic neoplasm, comprising 9-20% of all odontogenic tumours [6]. It most commonly occurs in the mandible (80%), with only 20% occurring in the maxilla [7].

Three subtypes of ameloblastoma include conventional ameloblastoma, which is the most common [8] an intraosseous lesion with predominantly solid areas that indicate a locally aggressive behaviour; unicystic ameloblastoma, which is an intraosseous lesion with cystic areas, luminal projections; and peripheral ameloblastoma, which is extraosseous in the soft tissues of the gingiva and the retromolar area, generally presenting a good prognosis.

Microscopic analysis reveals that the neoplasm is arranged in islands and cords of odontogenic epithelium. These consist of columnar cells with hyperchromatic nuclei in the basal layer, exhibiting peripheral palisading. The cells show reverse polarity away from the basement membrane, as seen in the Vickers-Gorlin change and subnuclear vacuolisation. Similar to normal odontogenesis, the suprabasal cells recapitulate the stellate reticulum.

Histologically, multiple patterns of ameloblastoma can be found. The follicular pattern includes islands of odontogenic epithelium exhibiting peripheral palisading, embedded in fibrous connective tissue. The second pattern, known as the plexiform pattern, shows cords and sheets of anastomosing odontogenic epithelial cells, where the classic features of ameloblastoma may not be apparent. The acanthomatous pattern exhibits squamous metaplasia and keratinisation of stellate reticulum-like cells. The granular pattern shows eosinophilic granular cytoplasm, with islands of hyperchromatic basaloid cells found in the basal cell or basaloid pattern. The stroma is composed of dense cellular fibrous connective tissue and is collagenised, with compressed and angulated islands of neoplastic cells in the desmoplastic pattern, which is more common in the maxillary area [9]. Less common patterns include clear cell and papilliferous types.

Recent studies of ameloblastoma have shown genetic mutations, predominantly the V-Raf Murine Sarcoma Viral Oncogene Homolog B Valine is replaced by Glutamic acid (E) (BRAFV600E) mutation in Mitogen-activated Protein Kinase (MAPK). Other less common mutations include Kirsten Rat Sarcoma (KRAS), Neuroblastoma Rat Sarcoma (NRAS), Harvey Rat Sarcoma (HRAS) and Fibroblast Growth Factor 2 (FGF2) [10].

Immunohistochemistry has a limited role in the diagnosis of ameloblastoma, as histomorphology plays a pivotal role in resolving diagnostic dilemmas. Immunohistochemistry markers of odontogenic epithelium, like Cytokeratin 19 (CK19), highlight the ameloblastoma cells, including those in the acanthomatous pattern [11]. Additionally, stellate reticulum will be displayed by CK13 and calretinin [12].

The differential diagnosis of maxillary ameloblastoma includes inverted papilloma, odontogenic fibromas, sinonasal non keratinising squamous cell carcinoma, adenoid cyst carcinoma and craniopharyngioma [13], as illustrated in [Table/Fig-8].

Differential diagnosis	Age and site of presentation	Microscopic features
Inverted papilloma	50-60 years, lateral nasal wall, nasal septum, paranasal sinus polyp	Prominent downward endophytic growth of round to elongated interconnected epithelial nests with smooth outer contour. Squamous, transitional or respiratory epithelium.
Ameloblastic fibromas	20 years,mandible	Small islands and cords of markedly attenuated ameloblastic epithelium. Two cells thick with dense collagenous stroma, occasional stellate reticulum.
Sinonasal non keratinising squamous cell carcinoma	50-70 years, maxillary sinus, nasal cavity	Squamous differentiation with minimal keratinisation. Minimal stromal desmoplasia.
Adenoid cystic carcinoma	50-70 years, maxillary sinus, ethmoid sinus, pharynx and salivary gland	Solid, cribriform, tubular pattern. Hyperchromatic angulated nuclei of ductal cells with flattened myoepithelial cells.
Craniopharyngioma	Bimodal 5-15 years and 45-60 years, pituitary gland, nasopharynx, sinonasal tract	Cords, lobules, nodular whorls of well differentiated squamous epithelium bordered by palisading columnar epithelium with stellate reticulum, wet keratin.

[Table/Fig-8]: Differential diagnosis of maxillary ameloblastoma [13].

Similar studies with different clinical and radiological presentations are summarised in [Table/Fig-9] [14-16]. In a study by Vasan V et al., a 60-year-old patient presented with gradually progressive facial swelling, accompanied by radiological evidence of a maxillary cystic lesion an unusual presentation with similar morphology [14]. Similarly, in a study by Dwivedi N et al., a 30-year-old patient presented with swelling on the left upper face, which was diagnosed as a unicystic lesion of the maxillary antrum based on morphology alone, comparable to the present study [15].

Articles	Age/Gender	Clinical features	Radiological findings
Vasan V et al., [14]	60 years/ male	Left-side face swelling gradually increased in size	Maxillary bone shows cystic lesion.
Dwivedi N et al., [15]	30 years/ male	Swelling in the left upper face and discharge on pressing the face	Unicystic lesion in maxillary antrum.
Karp J et al., [16]	64 years/ male	History of recurrent allergic fungal sinusitis	Unicystic mass in maxillary sinus
Current study	77 years/ male	Nasal obstruction	Mass with evidence of bony erosion

[Table/Fig-9]: Similar articles published in literature [14-16].

In another case reported by Karp J et al., a 64-year-old male with a recurrent history of allergic fungal sinusitis and no other complaints exhibited findings parallel to the present study [16]. Additionally, a case report by Chebil A et al., described a 54-year-old female who presented with painless ulceration of the alveolar process of the maxilla, showing the morphology of ameloblastoma that was comparable to the present study [17].

Recent studies have approached the treatment of ameloblastoma using BRAF inhibitors due to the associated mutations and their prognostic effects on management [18].

CONCLUSION(S)

Maxillary ameloblastoma, an uncommon disease, is noteworthy due to its rarity and the diagnostic challenges it presents. Its locally aggressive behaviour and propensity for recurrence often affect the patient's quality of life. The disease requires a multidisciplinary approach for evaluation and management, including clinical

assessment, radiological imaging, histopathological evaluation and treatment. Identifying the complexity of the condition at an early stage and adapting the appropriate surgical treatment is necessary to achieve the best outcomes for patients.

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