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Primary sinonasal ameloblastoma – a rare cause  
of unilateral nasal obstruction

Примарни синоназални амелобластом – редак узрок  
једностране носне опструкције

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## Primary sinonasal ameloblastoma – a rare cause of unilateral nasal obstruction

### Примарни синоназални амелобластом – редак узрок једностране носне опструкције

#### SUMMARY

**Introduction** Ameloblastomas are rare, locally invasive benign jaw tumours originating from odontogenic epithelium and their presence in sinonasal tract is usually due to their spread from gnathic region of the maxilla. Primary sinonasal ameloblastomas are extremely rare, with only a handful of cases being reported so far. The objective of this article was to describe a patient with a primary ameloblastoma of the right maxillary sinus and nasal cavity.

**Case outline** We report a case of a 67-year-old male patient with a year-long history of progressive unilateral nasal obstruction. Clinical and CT examination revealed a mass in the right maxillary sinus and right nasal cavity. After in-office biopsy in local anaesthesia which suggested the diagnosis of ameloblastoma, the patient underwent complete removal of the mass by medial partial maxillectomy. Histopathologic analysis confirmed the diagnosis of ameloblastoma.

**Conclusion** Primary sinonasal ameloblastoma is clinically and radiographically similar to the more common pathology of this particular area and should be included in the differential diagnosis of unilateral nasal obstruction. The treatment of choice is complete surgical resection. Due to the rarity of the disease and small number of cases described so far in the literature, there is still no consensus regarding the optimal surgical technique.

**Keywords:** ameloblastoma, sinonasal tumour, paranasal sinuses, maxillectomy

#### САЖЕТАК

**Увод** Амелобластоми су ретки, локално инвазивни бенигни тумори вилица који потичу од одонтогеног епитела, а њихова појава у пределу носа и синуса је најчешће последица ширења тумора из загрижајног предела горње вилице. Примарни амелобластоми носно-синусне регије су изузетно ретки и досада је описано свега неколико случајева. Циљ овог чланка је био да прикажемо случај пацијента са примарним амелобластомом десног максиларног синуса и носне шупљине.

**Приказ болесника** Приказујемо случај мушкарца старог 67 година са прогресивном једностраном носном опструкцијом. Клинички налаз и компјутеризована томографија су указали на експанзивну промену у десном максиларном синусу и у десној носној шупљини. Након биопсије у локалној анестезији која је указала на амелобластом, учињена је парцијална медијална максилектомија и ресекција тумора. Хистопатолошка анализа је потврдила дијагнозу амелобластома.

**Закључак** Примарни амелобластом носно-синусне регије је клинички и радиолошки сличан чешћој патологији ове регије и требало би га укључити у диференцијалну дијагнозу једностране носне опструкције. Лечење избора је комплетна хируршка ресекција. Обзиром на реткост овог обољења, као и на мали број до сада описаних случајева, тренутно не постоји сагласност о оптималној хируршкој техници.

**Кључне речи:** амелобластом, носно-синусни тумор, параназалне шупљине, максилектомија

#### INTRODUCTION

Ameloblastomas are rare, locally invasive benign tumours originating from odontogenic epithelium [1]. They consist about one percent of all jaw tumours, with mandible more frequently affected than maxilla [2]. Primary sinonasal ameloblastomas without involvement of the maxillary alveolus are extremely rare. In this article we describe the case of sinonasal ameloblastoma located in the right maxillary sinus and in the right nasal cavity, which was initially biopsied in ambulatory setting and subsequently treated by partial medial maxillectomy.

## CASE REPORT

A 67 year-old male patient came to our department with a year-long history of progressive, right-sided nasal obstruction. Relevant medical history included hypertension and glaucoma, for which he was taking regular medications. There was no history of prior sinonasal disease, allergies or tobacco use. Anterior rhinoscopy revealed a solitary polypoid lesion, originating from the middle meatus (*meatus nasi medius*) and partially involving the common nasal meatus (*meatus nasi communis*) on the right side. The surface of the lesion didn't have the typical "glassy" appearance of nasal polyp and was somewhat more firm on palpation. A CT of the nose and paranasal sinuses demonstrated mixed-density mass, completely opacifying the right maxillary sinus, with displacement and partial destruction of its medial wall and propagation in the nasal cavity (Figure 1). An in-office biopsy under local anesthesia was performed and initial histopathologic examination suggested a diagnosis of ameloblastoma. The patient then underwent right partial medial maxillectomy through the modified Weber-Ferguson incision (Figure 2). During the operation the solid and well-defined lesion completely filling the right maxillary sinus was noted, along with its intranasal component which was observed on initial clinical examination. The lesion was removed in en-bloc fashion, together with the medial wall of the maxillary sinus, including the inferior nasal concha. The structures removed resulted in the creation of wide antrostomy, which would further facilitate the postoperative examinations using rigid Hopkins telescopes. The definitive diagnosis of ameloblastoma was confirmed by histopathology (Figure 3). The postoperative period was uneventful and the patient was discharged four days following surgery. The patient is currently on a regular endoscopic follow-up, which has shown no recurrence of the disease 10 months after surgery.

## DISCUSSION

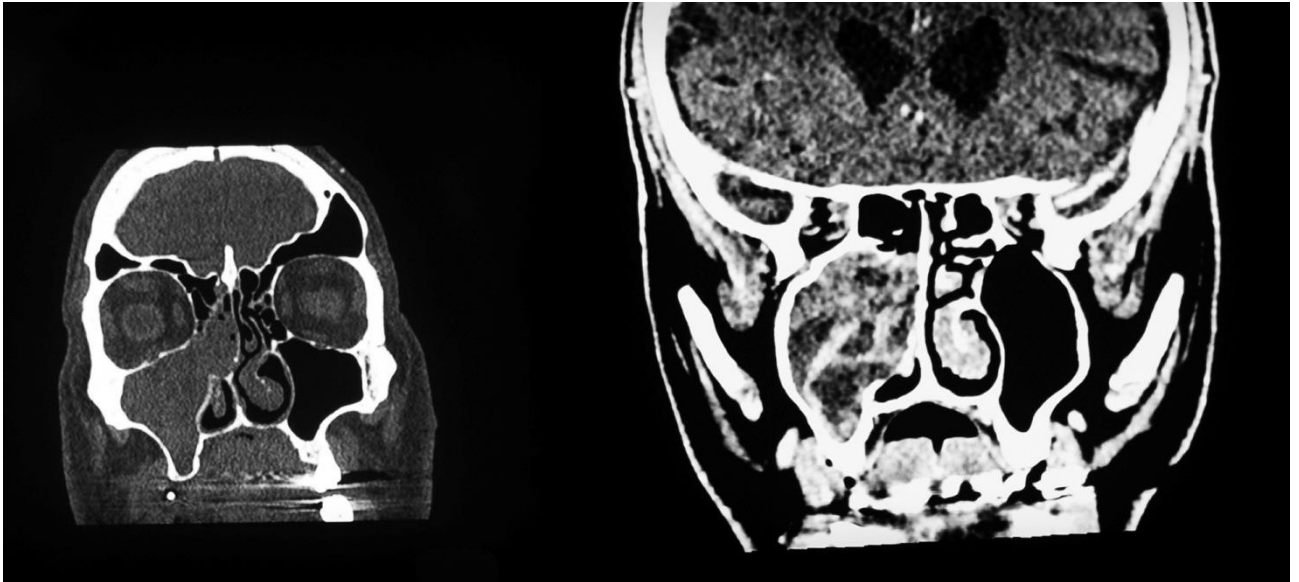
Most ameloblastomas in the sinonasal region appear secondary to an extension of a tumour of gnathic origin into this area [3]. However, primary sinonasal ameloblastomas without evident gnathic involvement, have also been described [4,5]. Unlike their gnathic counterparts which appear usually between 35 and 45 years of age with no distinct gender predilection, sinonasal ameloblastomas mostly affect male patients in their 6th and 7th decades of life [6]. The proximity of the odontogenic apparatus and sinonasal cavity during embryogenesis could potentially result in misplacement of odontogenic cells in the sinonasal epithelium or the abnormal differentiation of the pluripotent basal cells of the sinonasal mucosa. The possibility of origin from the bony structures of the nasal turbinates has also been proposed [5,6]. It is unclear whether the chronic inflammation of the sinonasal mucosa may be the triggering event in the pathogenesis of ameloblastoma or if it is secondary due to tumour presence in the sinonasal tract [7]. Histologically, ameloblastomas are benign neoplasms with locally

aggressive behaviour and marked tendency for late recurrence. Cases of malignant alteration and distant metastases, although exceptionally rare, have also been reported [8,9]. They manifest with nonspecific nasal symptomatology, including progressive nasal obstruction, recurrent epistaxis, facial swelling or sinusitis. On examination, a soft tissue mass in the nasal cavity is usually noted. Clinically and radiologically, sinonasal ameloblastomas are indistinguishable from more common nasal pathology such as polyps, chronic sinusitis or inverted papilloma. Unilateral nasal involvement and CT signs of bone affection should raise suspicion of a neoplastic process, and definitive diagnosis of ameloblastoma is only possible with a biopsy followed by histopathological analysis. A wide surgical excision is the treatment of choice [10]. The choice of operation is usually dictated by the extent of the disease. A variety of transfacial approaches, such as lateral rhinotomy, sublabial or Weber-Ferguson incisions provide good visual control and enable wide excision. Simple curettage of maxillary sinus is usually associated with recurrence. Wider excisions by means of partial or radical maxillectomy have better outcomes. Recently, endoscopic management of ameloblastomas has been described, with reportedly less perioperative morbidity and better disease control [11]. A combined transfacial and endoscopic approach was also reported [12]. In the case described we have chosen medial maxillectomy through modified Weber Ferguson incision, based on involvement both of nasal cavity and of maxillary sinus, as well as CT evidence of bone affection of its medial wall. Currently there is no consensus regarding the choice of surgical technique, as a result of the small number of reported cases. In cases of infiltrative tumours where complete removal proves difficult or impossible due to its adherence to the surrounding structures, postoperative radiotherapy can be used as an adjuvant treatment. [13] In our case, the tumour was well-defined and was completely removed through the selected approach without difficulties. Radiotherapy could be used as a single therapy in the case of locally advanced disease uncontrollable by surgery alone. [14].

Given the benign histologic nature of sinonasal ameloblastoma and its high potential for local recurrence, we would favour the wide surgical excision which we achieved through transfacial approach. The definitive choice of surgical treatment for sinonasal ameloblastoma is still controversial and requires a larger series of patients. Due to the tumour potential for late recurrence, regular periodic checkups are essential and the exact treatment outcome should only be assessed after long-term follow-up. In our case, wide communication of the nasal cavity with the remnant of the maxillary sinus was created, thus providing relatively simple follow-up using rigid Hopkins telescopes in outpatient service.

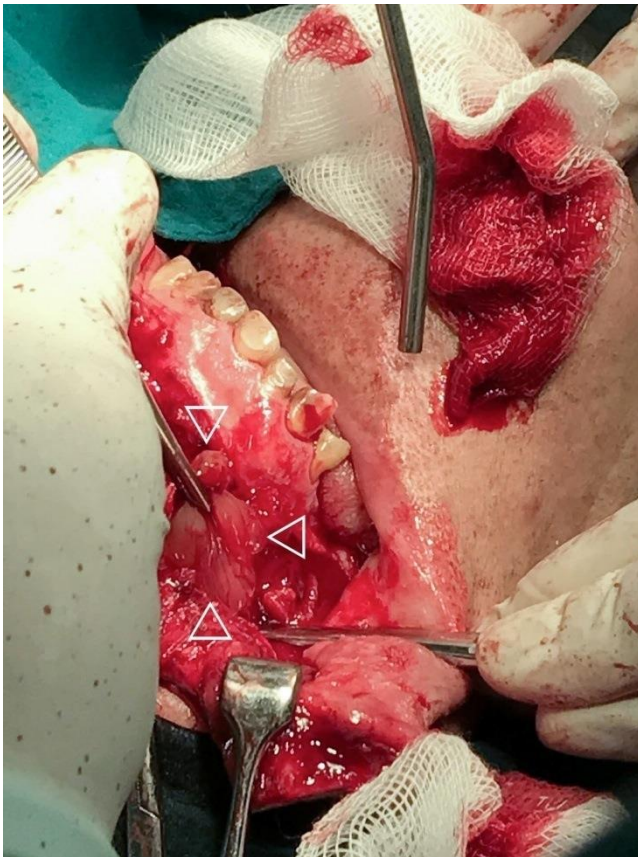
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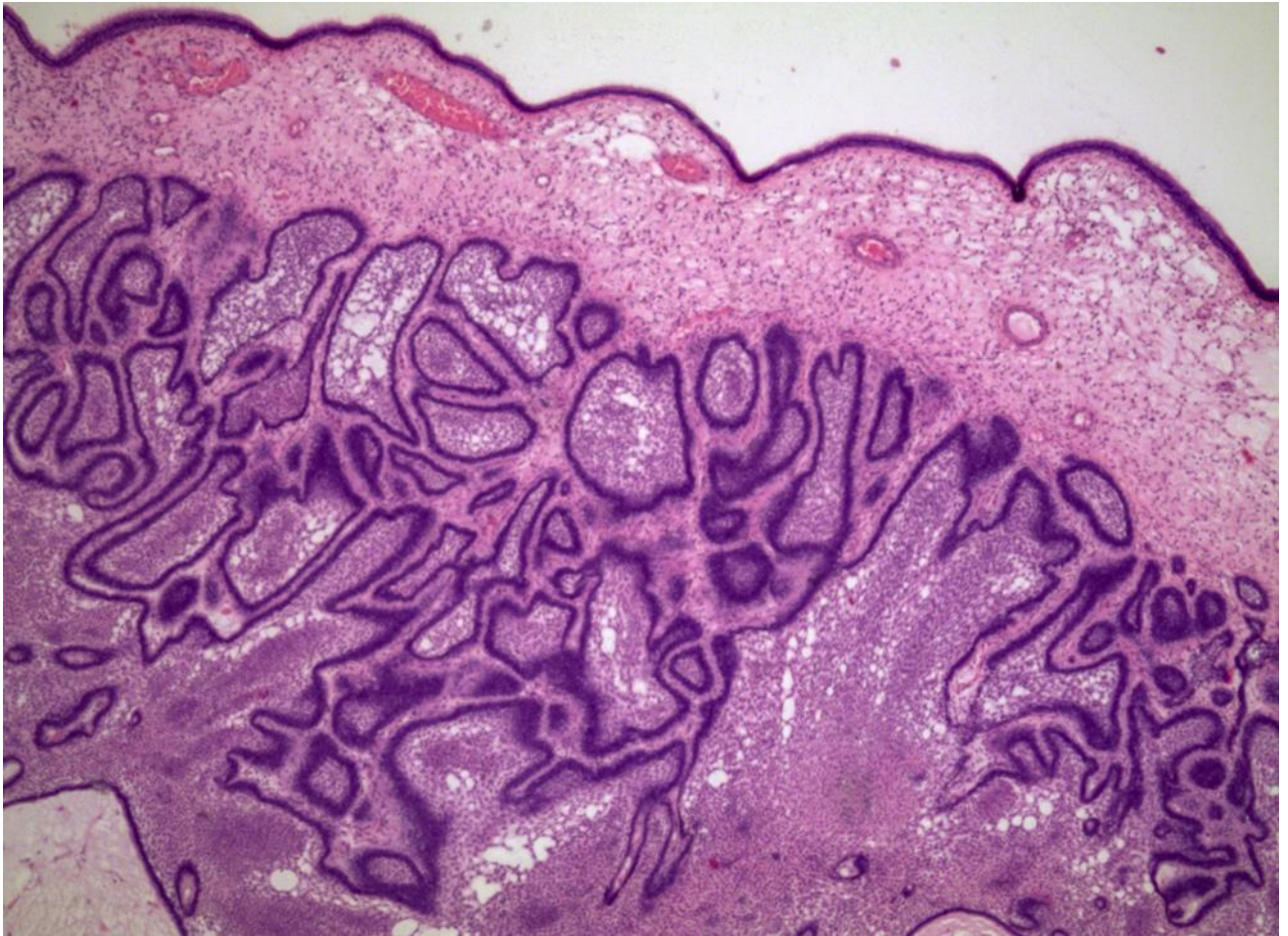


**Figure 1.** Coronal CT of the nose and paranasal sinuses (bone and soft tissue windows) demonstrating a mixed density mass in the right maxillary sinus and with an extension to the right nasal cavity

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**Figure 2.** The tumour (triangles) exposed through the modified Weber-Ferguson incision



**Figure 3.** The tumour consisting of loosely arranged stellate cells resembling the stellate reticulum of the tooth germ, surrounded by peripheral rim of palisading cells (H&E,  $\times 25$ )

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