

# Calcifying Epithelial Odontogenic Tumor: Report of a Recurrent Destructive Case with Review of Literature

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## Abstract

Calcifying epithelial odontogenic tumor (CEOT) is an uncommon odontogenic tumor with well-known histopathological features and a challenging treatment plan. Although some investigators advocate conservative approach as the treatment of choice, others believe in radical surgical excision to avoid recurrence or malignant transformation. The main objective of this case report is presenting an unusual destructive recurrence of CEOT after 8 years in a 34-year-old woman, and discussing an effective treatment plan for this tumor.

**Keywords:** Odontogenic tumor; Maxillary sinus; Neoplasm, Recurrence

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## INTRODUCTION

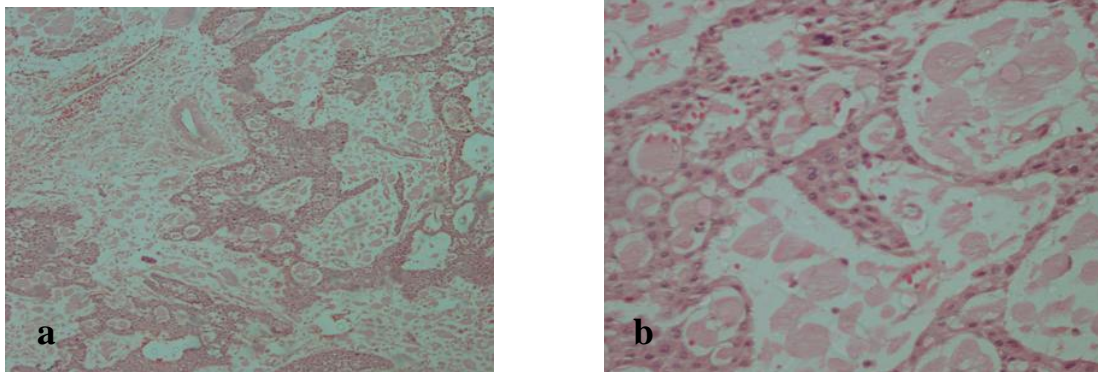
Calcifying epithelial odontogenic tumor, also known as Pindborg tumor [1], is a rare benign odontogenic tumor with an uncertain histogenesis and unique microscopic features, which can be easily distinguished from other odontogenic lesions.

The intraosseous variant of this neoplasm most commonly occurs in the posterior area of the mandible. Extraosseous forms of the tumor have been reported in less than 5% of the cases [2].

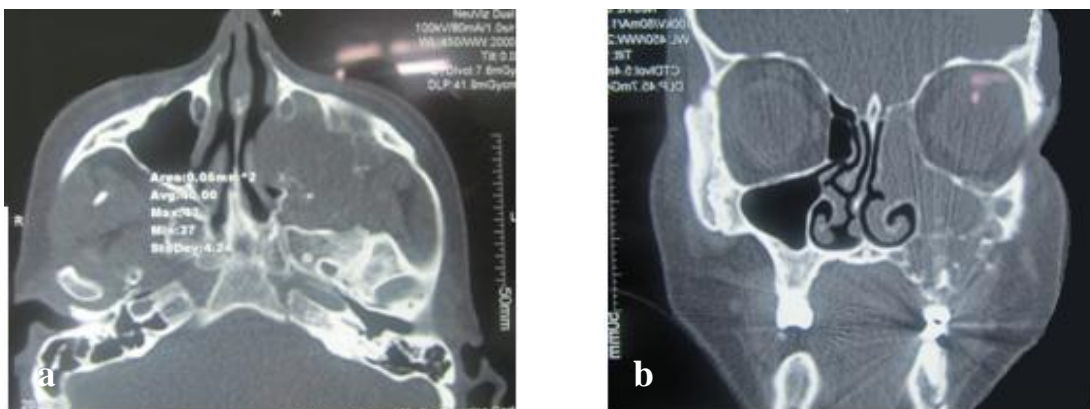
During the past ten years more than 200 cases of CEOT have been reported, most of which were found in young adults with a mean age of 40 years, with approximately similar incidence in men and women [3].

Although this neoplasm is considered to be less aggressive than ameloblastoma, the most common odontogenic tumor, several reports of destructive CEOT have been published over the past years [4, 5] and reports of recurrent CEOT has made the investigators focus on the necessity to revise the conventional surgical treatment [6, 7]. Therefore, it appears that conservative local resection, which is the current surgical treatment plan of choice, [8], needs to be modified based on reliable prognostic clinical and histopathological criteria.

Here, we report a destructive recurrence of an intraosseous CEOT after 8 years, in the upper jaw of a 34-year-old female, which emphasizes on the necessity of meticulous follow up for similar lesions.



**Fig 1.** Photomicrographs (of primary lesion) show islands and sheets of polygonal epithelial cell with prominent nuclei and eosinophilic cytoplasm. Also, extensive amyloid deposition is remarkable. (H&E staining; X100) (a); (X400) (b)”



**Fig 2.** Axial (a) and coronal (b) computed tomography scan sections showing extension of the tumor to the left maxillary sinus and destruction of adjacent tissues.

Also, we review the most common risk factors, which could be utilized to predict clinical behavior of this lesion. Finally, we propose an effective treatment plan.

### CASE REPORT

A 34-year-old female with mild facial swelling on the left side was referred to the Oral and Maxillofacial Surgery Department. Intra-oral examination revealed a painless, non-tender swelling at the upper left molar teeth area with intact overlying mucosa. The teeth were not mobile and examination of cervical lymph nodes was unremarkable.

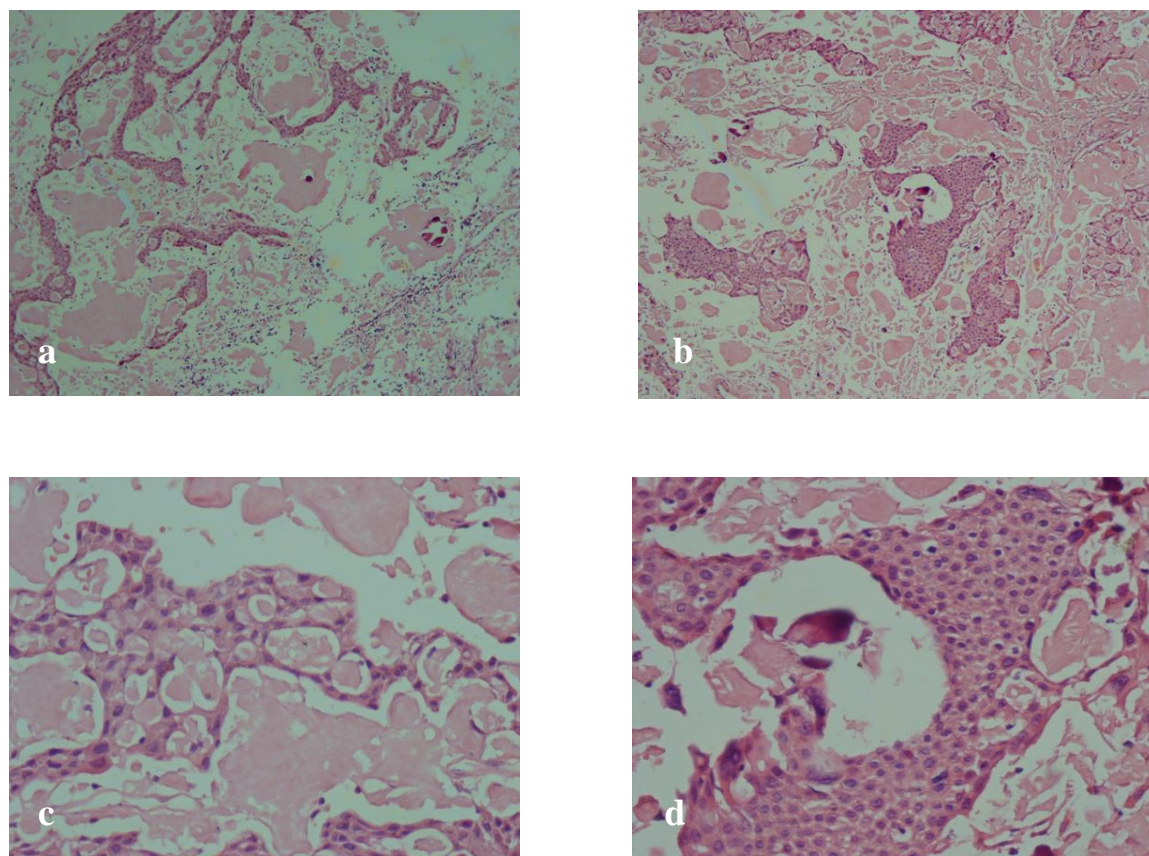
There were no focal neurological deficits on examination. About eight years ago, she was referred to the author for discomfort at the same area. At that time, her chief complaint was mild swelling of her face.

The radiographic examination had shown a small ill-defined radiolucency at the periapical region of premolar and molar teeth. No obvious destructive impact on adjacent structures was observed. Gross examination of the surgically excised lesion revealed ten small, irregular, brown tissue densities measuring 2.3 x 2 x 0.6 cm in total.

Histopathological examination of the excised lesion revealed a Pindborg tumor (Figure 1). The microscopic examination showed well-known sheets and strands of polyhedral, polymorphous epithelial cells with clear intercellular bridges.

Huge aggregations of amyloid material and very small foci of calcification were also observed.

Panoramic view taken from the present illness showed an ill-defined radiolucency in the same area.

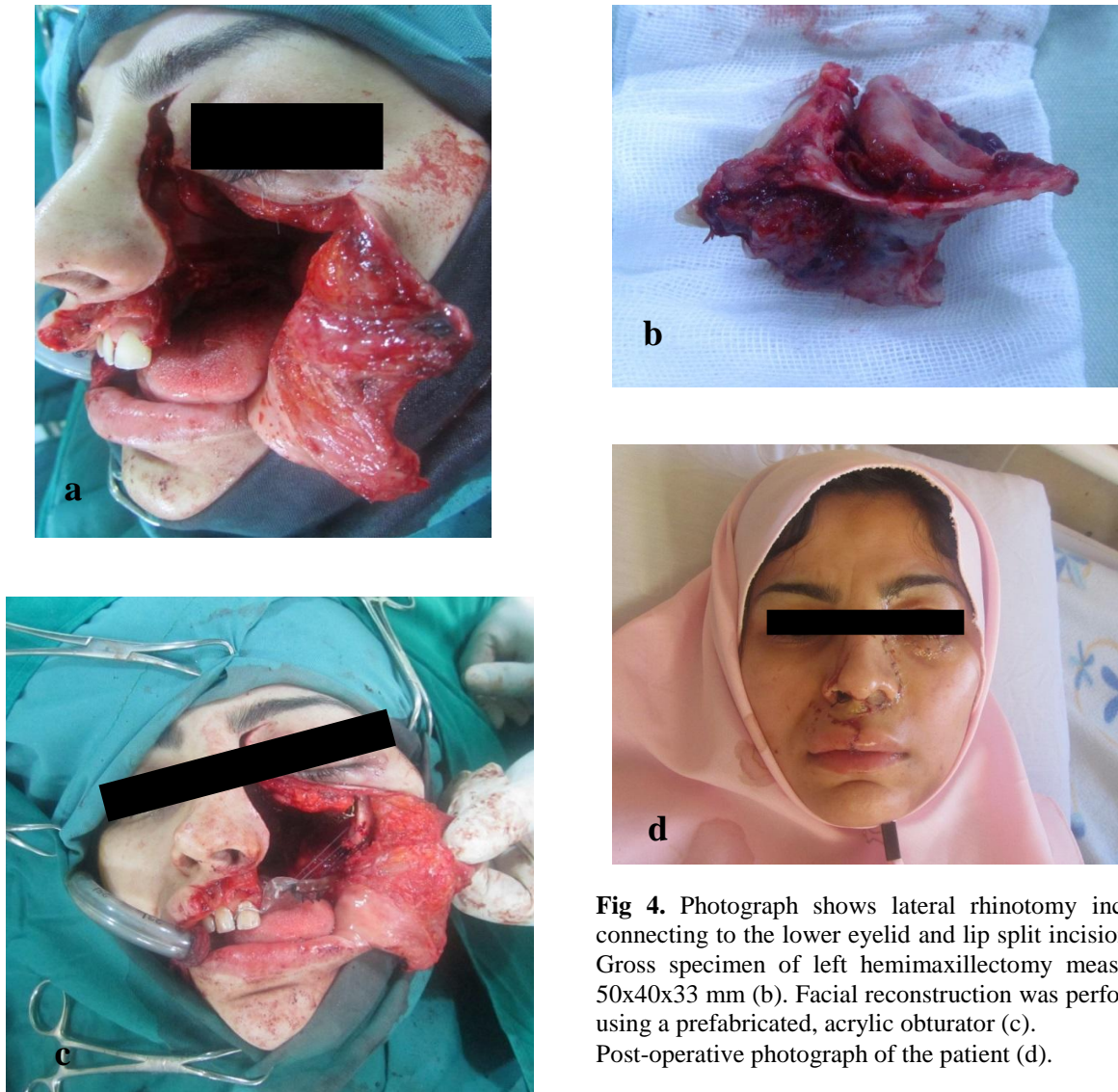


**Fig 3.** Microscopic evaluation shows nests and strands of large polyhedral epithelial cells with mild nuclear pleomorphism. Also, variable sized sheets of amyloid and small irregular foci of calcification are observed. (H&E staining; X100) (a, b); (X400) (c, d)

On computed tomography scan, a massive intraosseous lesion was disclosed in the left maxilla involving the orbit and ethmoidal air cells superiorly and the nasal airway medially (Figure 2). Incisional biopsy and histopathological examination verified the clinical diagnosis of CEOT (Pindborg tumor), which confirmed the recurrence of the previous tumor (Figure 3). The microscopic findings of the incised lesion were similar to the primary one, in which amyloid aggregation was remarkable. Conventional epithelial cell features of CEOT, including polyhedral cells with mild pleomorphism and prominent intercellular bridges were also evident. The only apparent distinction was the presence of more foci of calcification in the recurrent lesion.

Surgical access to the tumor was gained through a lateral rhinotomy incision connecting to lower eyelid and lip split incisions (Weber Fergusson's approach) (Figure 4a). Lateral rhinotomy incision was extended superiorly to provide access to the ethmoidal air cells. After complete excision of the entire tumor associated with partial ethmoidectomy (Figure 4b), a prefabricated acrylic obturator was inserted into the surgical defect for postoperative esthetics and to preserve function.

This was secured in place using three 12 mm titanium mini-screws (Figure 4c). Orbital floor reconstruction was carried out using a titanium mesh plate (Figure 4d). Recurrence was not detected until one year after the surgery (Figure 5).



**Fig 4.** Photograph shows lateral rhinotomy incision connecting to the lower eyelid and lip split incision (a). Gross specimen of left hemimaxillectomy measuring 50x40x33 mm (b). Facial reconstruction was performed using a prefabricated, acrylic obturator (c). Post-operative photograph of the patient (d).

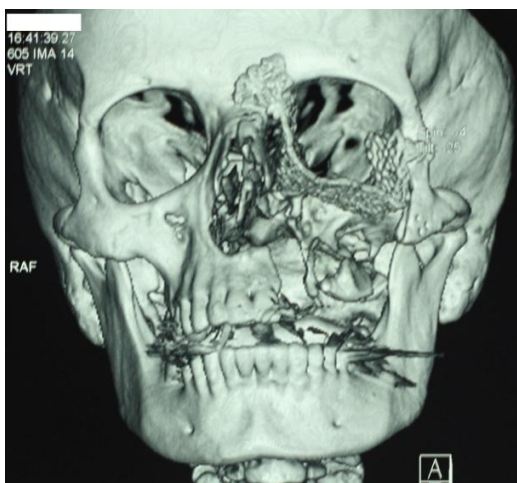
## DISCUSSION

CEOT is a benign neoplasm with unpredictable behavior, which usually presents as painless, slow growing expansion of the jaws, accompanied with destruction of the associated normal tooth structure [3].

The tumor is usually treated surgically, ranging from simple enucleation to extensive radical resections [9]. Being able to predict the risk of recurrence or locoregional destruction could allow surgeons to come up with an effective treatment plan especially in destructive cases of the upper jaw [7].

Several prognostic factors have been proposed to estimate recurrence risk of the tumor and consequently to choose the extension of the surgical margins. Microscopically, less amyloid aggregation and foci of calcification have been suggested in association with more aggressive tumor behavior [10].

In the present case, while the small and scattered foci of calcification confirm the above concept, the abundant amyloid aggregation does not support this belief. Furthermore, the uncommon histopathological variants of CEOT, especially clear cell type, have made predicting the probability of tumor recurrence and prognosis particularly challenging [11].



**Fig 5.** Three-dimensional CT scan revealed no recurrence in the follow up.

Although some investigators advocate more aggressive behavior of the clear cell variant of CEOT, the lack of reliable long term follow up information from the published case reports is apparent [12].

In addition, few cases of malignant transformation have been presented, in which cytological features were abundantly representative of the transformation [13].

Also, it seems that the clinical features of CEOT consisting of size, anatomic site, health status of the patient, and reconstruction methods have an obvious impact on the extent of the planned surgical resection [14, 15]. In our case, following the recurrence, because of the involvement of the maxillary bone and the remarkable extension into the adjacent struc-

tures, such as maxillary and ethmoidal sinuses, the tumor was resected with wide surgical margins.

The unusual destructive behavior of CEOT in primary or recurrent lesions has been published in several case reports [15], which confirm the necessity to revise the current treatment plan in similar clinical situations.

In a brief review of the literature, we found six cases of Pindborg tumor with an extension to the maxillary sinus, which are summarized in Table 1.

Although the current protocols suggest a follow up period of five years to detect any probable recurrence of Pindborg tumor [9], in the present case, we found a rare recurrence eight years after the primary diagnosis. While some authors suggest an individualized treatment based on the clinical and histologic findings of the lesion, others insist on a more aggressive surgical resection similar to what is performed for ameloblastoma [20].

## CONCLUSION

In conclusion, we are in favor of designing a meticulous treatment plan for CEOT in order to prevent the risk of recurrence and local invasion by performing an extensive primary surgical resection.

This compels us to evaluate future cases clinically and microscopically to come up with an appropriate treatment plan, which may range from a simple enucleation or curettage to hemimandibulectomy or hemimaxillectomy.

**Table 1.** Clinicopathological features of the published cases of Pindborg tumor involving the maxillary sinus.

Reference	Age	Gender	Microscopic type	Follow up
Gon [16]	35	Female	Conventional	NP
Stimson et al. [17]	35	Male	NF	NF
Lee et al. [18]	27	Female	Conventional	7 yrs.
Bridle et al. [13]	30	Female	Conventional	NP
Mohtasham et al. [7]	18	Male	Conventional	NP
da Rosa et al. [19]	33	Female	Conventional	NP
Sahni et al. [15]	52	Male	Conventional and clear cell	3 yrs.
Present case	34	Female	Conventional	8yrs.

NP: Not Performed/ NF: Not Found

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