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CASE REPORT

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Noncalcifying type of calcifying epithelial odontogenic tumor: A rare case report and literature review

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Key Clinical Message

There has been a recent spike in reporting of noncalcifying variants of CEOT with the WHO 2022 classification of head and neck tumors. The present case describes a rare histopathological variant of CEOT of which a handful of cases have been reported.

Abstract

This work aimed to report a rare case of noncalcifying type of Pindborg Tumor involving the right mandible. The absence of calcifications in the calcifying epithelial odontogenic tumor presents a diagnostic challenge and prognostic implications. A literature review of the 16 reported clinical cases of this scarce variant was performed.

KEYWORDS

amyloid, calcifying epithelial odontogenic tumor, mandible, odontogenic tumors

1 | INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT) is a benign lesion that Thoma and Goldman first described, and then in 1958, it was described in detail by Jens Pindborg.[1] It is classified by the World Health Organization (WHO) among the benign odontogenic epithelial tumors.[2] Since this description, many cases have been reported in the literature. The mean age of occurrence is the fourth decade with no gender predilection, and it has a mandibular preference.[1,3] The 5th edition of the WHO Classification of Head and Neck Tumors, published in March 2022, contained significant changes regarding CEOT. Three subtypes have been described: clear cell CEOT, cystic/microcystic CEOT, and noncalcifying/Langerhans cell-rich CEOT.[2]

In this work, a rare clinical case was reported, and a literature review of the different reported cases of the noncalcifying type of calcifying epithelial odontogenic tumor was conducted.

2 | CASE REPORT

A 40-year-old female patient was referred to the oral surgery department for the management of a radiolucent lesion associated with the right mandibular impacted wisdom tooth

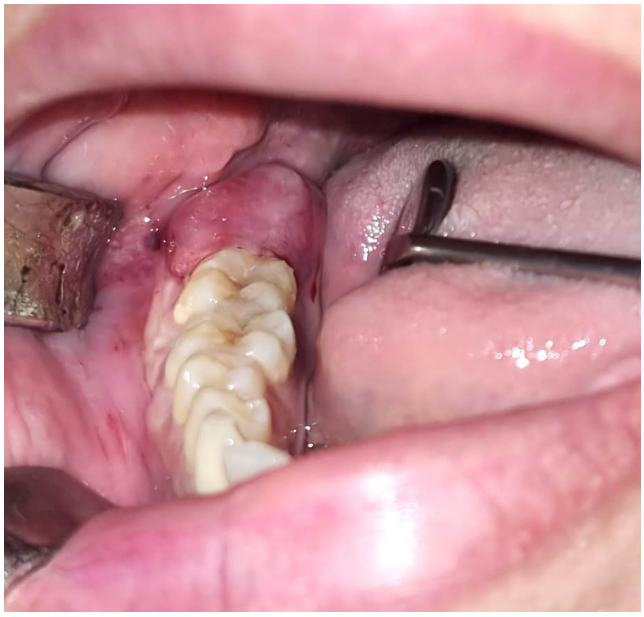

FIGURE 1 Intraoral aspect showing a lower well-circumscribed gingival exophytic mass extending distal to the right second mandibular molar.

of incidental discovery. Her familial and past medical history was noncontributory. She was asymptomatic.

The extraoral examination was normal. The intraoral examination showed a well-circumscribed mandibular sessile nodule distal to tooth 47, extending on both vestibular and lingual gingiva and measuring approximately 1 cm in diameter. It was covered by normal mucosa and firm on palpation. Tooth 48 was clinically absent (Figure 1). Radiographic examination using panoramic radiograph showed a unilocular expansile radiolucent homogenous image on the right mandibular angle, associated with an impacted wisdom tooth 48. A cone beam computed tomography was required to precisely determine lesion expansion and its limits. CBCT showed a well-defined, radiolucent lesion measuring 2 cm × 1.5 cm × 1 cm with the cortical expansion of the right posterior mandible and displacement of the inferior mandibular canal (Figure 2).

Differential diagnoses included odontogenic cysts, odontogenic keratocysts, dentigerous cysts, ameloblastoma, and osteolytic lesions related to the brown tumor. Parathormone, vitamin D, phosphor, and calcium serum

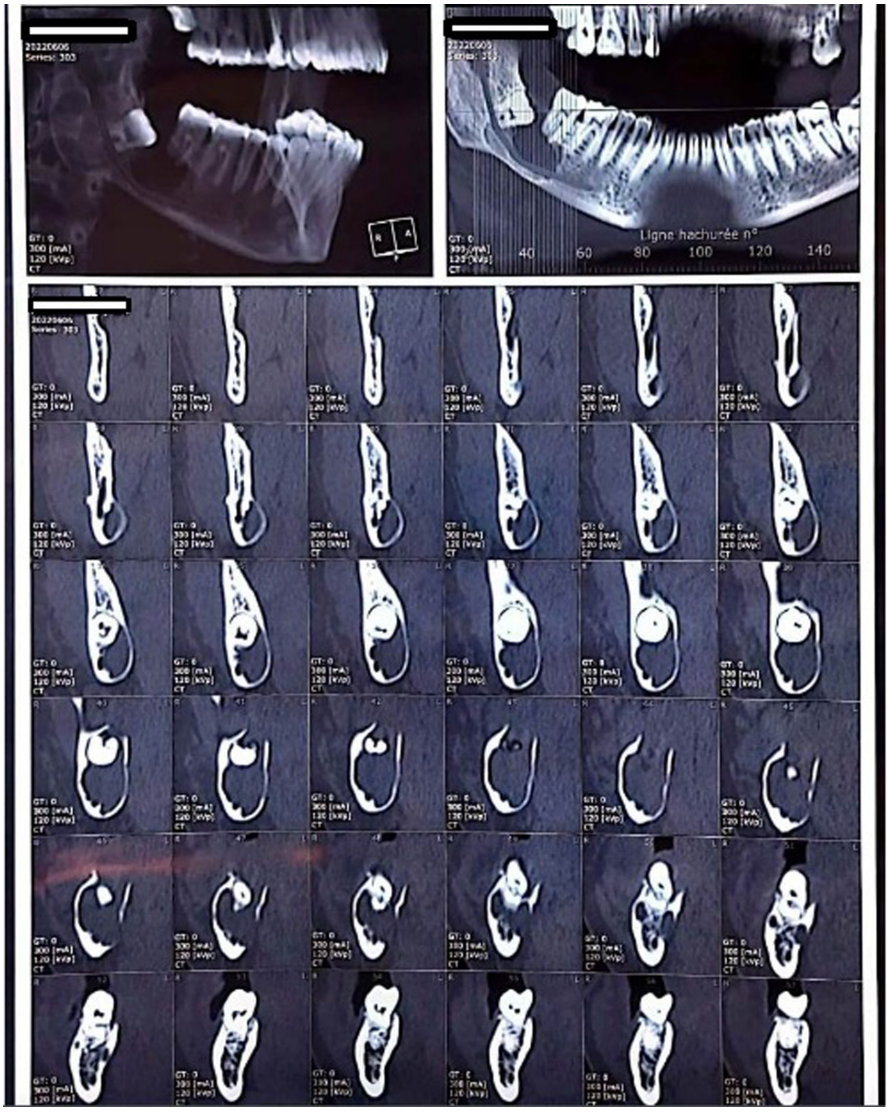

FIGURE 2 Cone-beam computed tomography shows a well-defined hypodense image on the right mandibular angle + impacted third molar.

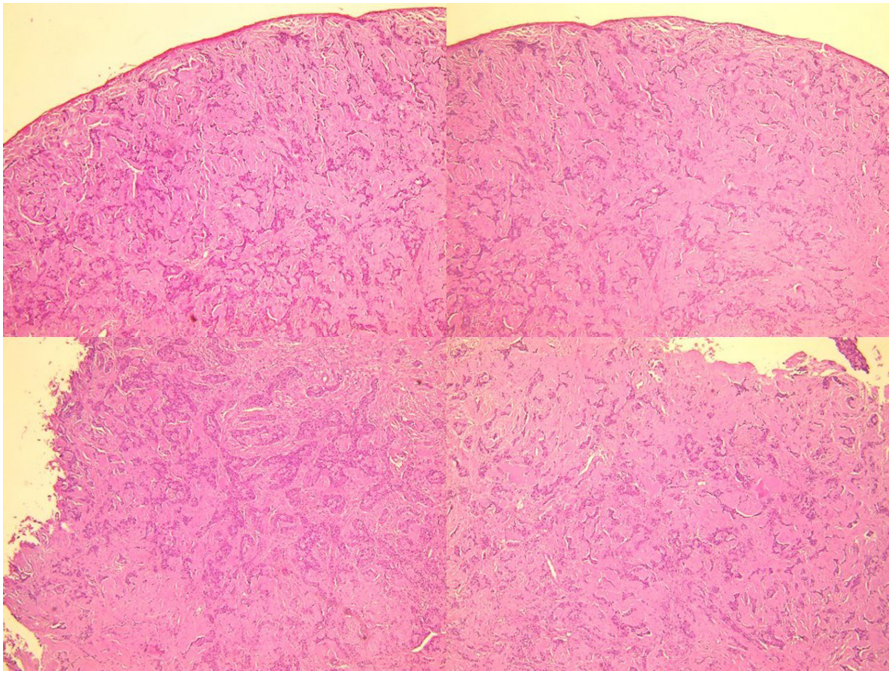

FIGURE 3 Histological sections stained with hematoxylin–eosin: Sheets and nests of polygonal epithelial cells with eosinophilic cytoplasm and large areas of amyloid-like material. (Hex40).

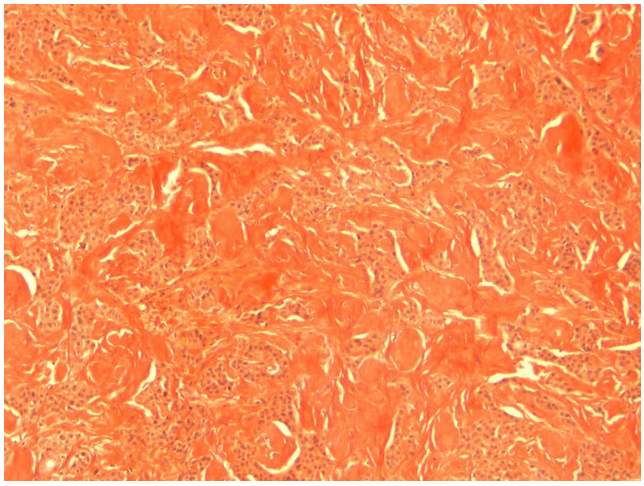

FIGURE 4 Amyloid-like material in tissue exhibits a deep red color with Congo red stain.

levels were requested. There were within the normal limits except for the decreased vitamin D level.

Total enucleation of the lesion was performed under local anesthesia, with the impacted wisdom tooth extraction.

Histopathological examination revealed a nodule formed by trabeculae and clusters of polyhedral epithelial cells, focally united by intercellular bridges. They had an eosinophilic cytoplasm and regular finely nucleated nuclei. These epithelial cells were separated by interspersed hyaline fibrous tissue, little cellular by places, globular, taking the aspect of amyloid-like material (Figure 3). This substance showed brick red color on Congo red staining (Figure 4). The diagnosis of a noncalcifying type of calcifying epithelial odontogenic tumor was consequently confirmed.

The postoperative period was uneventful. The patient continues to be on regular follow-ups. No sign of recurrence was detected after 6 months of follow-up (Figure 5).

3 | DISCUSSION

The calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor has been well documented in the literature since its first description. It is a rare lesion, the reported cases did not exceed 400,[1] representing less than 1% of all odontogenic tumors.[3] The noncalcifying type is rare; it is the least reported one.[1] The radiological and histological features of the noncalcifying type of calcifying epithelial odontogenic tumor are distinguished from those of the usual calcified variant. Consequently, there was a diagnostic challenge in the reported case, and the diagnosis of a Pindborg tumor was not suspected.

On radiographic examination, Pindborg tumor appears most commonly as a mixed radiopaque/radiolucent image. Both multilocular and unilocular presentations were described. The association with an impacted tooth was frequently found. The present case was exempt from radiopacities. This radiolucent aspect is insufficient to diagnose a noncalcifying variant since it was described in early diagnosed immature lesions, in which calcifications were found on histopathological examination. This radiolucent aspect was also described in some called cystic variants.[4] This radiolucent aspect may lead to the misdiagnosis of this variant as an odontogenic cyst, like in the reported clinical case.

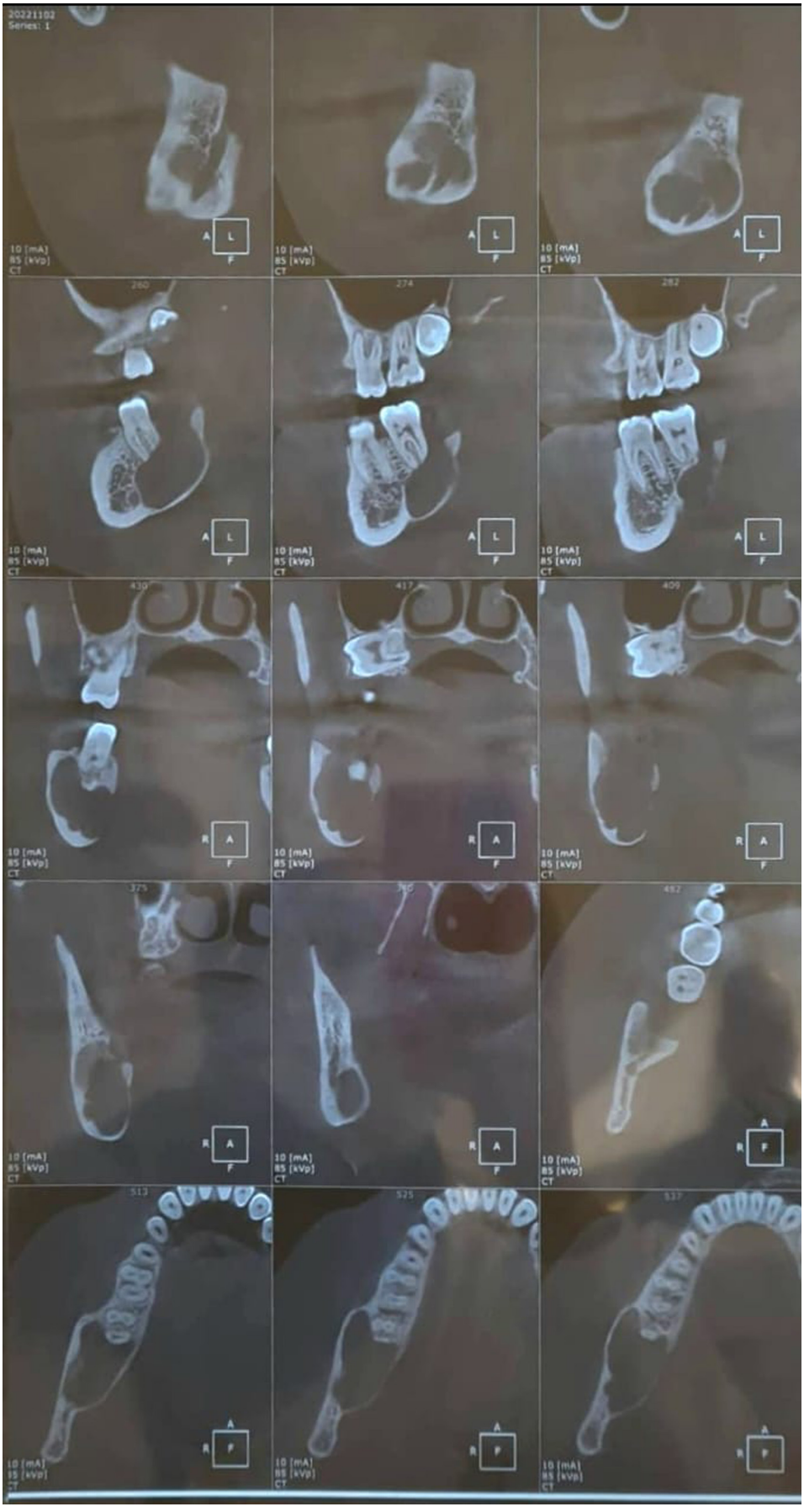

FIGURE 5 Control cone beam computed tomography 5 months postoperative.

TABLE 1 Review of the reported cases of noncalcifying type of CEOT.

Author	Year	Age	Sex	Location	Histopathology	Recurrence (follow-up period)
Aufdermaur et al.[6]	1981	68	M	Mandible	No calcification	No (1 year)
Asano et al.[7]	1990	44	F	Maxilla	No calcification Langerhans cells Clear cells	Nonspecified
Takata et al.[8]	1993	58	M	Maxilla (posterior)	No calcification Langerhans cells	Nonspecified
Hafian et al.[9]	2004	61	M	Maxilla (anterior)	No calcification	Nonspecified
Wang et al.[10]	2007	52	F	Maxilla (anterior)	No calcification Langerhans cells	Nonspecified
Wang et al.[11]	2007	38	M	Mandible	No calcification Clear cells	Nonspecified
Wang et al.[11]	2007	39	F	Maxilla	No calcification Clear cells	Nonspecified
Kaushal et al.[3]	2012	57	M	Mandible	No calcification	No (1 year)
Afroz et al.[12]	2013	20	F	Maxilla (anterior)	No calcification Langerhans cells Clear cells	Nonspecified
Chen et al.[13]	2014	40	F	Maxilla (anterior)	No calcification	No (5 years)
Chen et al.[13]	2014	58	M	Maxilla	No calcification Langerhans cells	No (10 years)
Tseng et al.[14]	2015	24	M	Maxilla	No calcification Langerhans cells	Nonspecified
Kurihara et al.[15]	2017	60	M	Mandible (anterior)	No calcification Clear cells	No (3 years)
Taneeru et al.[16]	2017	27	F	Mandible (posterior)	No calcification	Nonspecified
Santosh et al.[17]	2019	43	F	Maxilla (anterior)	No calcification Langerhans cells	No (18 months)
Patankar et al.[18]	2021	43	M	Mandible (posterior)	No calcification Clear cells	Nonspecified

The most distinctive microscopic feature of classical CEOT is the presence of sheets of polyhedral cells, amyloid globules, and Liesegang ring calcifications in the tumor tissue.[5] According to the literature, noncalcifying cases described are primarily associated with the presence of Clear Cells and Langerhans Cells. This led authors to the definition of variants of this tumor: the Clear Cells variant and the Langerhans Cells-rich variant.[1]

The absence of calcification not only poses a diagnostic problem but also has been suggested to be an indicator of poor differentiation of the tumor. Consequently, the noncalcifying epithelial odontogenic tumor would have more recurrence risk and require radical treatment and a long-term follow-up.[3]

The international literature was reviewed; only 16 cases of non-calcifying type of calcifying epithelial odontogenic tumor were found from 1981 to 2021 and are resumed in Table 1. No sex predilection was noted (seven females/nine males). Age ranged from 20 to 68 years old. The present reported case adds a female patient aged 40 years old. Ten cases were developed in the maxillary bone over seven cases in the mandible, including the present reported case. Histologically, four of the reported noncalcifying variants contained clear cells, five contained Langerhans cells, two contained both clear and Langerhans cells, and five showed neither clear cells nor Langerhans cells like the reported clinical case. Treatment modalities varied between enucleation and partial resection with 1 cm margins to prevent recurrence. Enucleation was preferred as a more conservative approach in the present case since the diagnosis of Pindborg tumor was unlikely suspected before histopathology, especially with the absence of calcification and the sporadic similar reported cases. Also, there were no clinical or radiological signs of aggressivity of the lesion.

Considering the prognostic implications that present the absence of calcifications, close follow-up appointments are mandatory to assess any possible recurrence early. In the literature, no cases of recurrence were reported over follow-up periods ranging from 6 months to 10 years.

4 | CONCLUSION

The absence of calcifications in CEOT was reported in very few cases of Pindborg tumor. A careful diagnostic approach should be conducted to diagnose this rare variant. Some authors suggested that it is a poorly differentiated form with more unpredictable behavior. This possible aggressiveness still represents a controversial subject; however, no recurrence has been reported to date. More case reports of noncalcifying type of calcifying epithelial odontogenic tumor are needed to investigate this entity better.

AUTHOR CONTRIBUTIONS

Hela Zouaghi: Writing – original draft. **Maroua Garma:** Writing – original draft. **Afef Slim:** Writing – review and editing. **Abdellatif Chokri:** Writing – review and editing. **Manel Njima:** Writing – review and editing. **Jamil Selmi:** Writing – review and editing.

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All authors of this manuscript have no conflict of interest.

DATA AVAILABILITY STATEMENT

All supporting data are available.

CONSENT STATEMENT

Written informed consent had been obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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