Calcifying Epithelial Odontogenic Tumor of The Anterior Maxilla: An Unusual Location

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ABSTRACT

Calcifying epithelial odontogenic tumor (CEOT) is a benign tumor of odontogenic epithelium. It is a locally invasive neoplasm with a characteristic feature of having amyloid. The mandible is twice more commonly affected than the maxilla and in the maxilla, posterior location is the preferred site of involvement. Here we present a case in a 50 years old male patient who presented with a non-tender hard swelling of the anterior maxilla measuring 2.5 x 1.5 cm, just right to the midline and at the base of the lateral incisor and canine. Imaging revealed a radiolucent lesion in the maxilla with small foci of calcification without any evidence of impacted tooth. Thus a provisional diagnosis of an Ameloblastoma was made. A local excision of the swelling was done along with extraction of the lateral incisor and canine. On histopathological examination, a diagnosis of Calcifying epithelial odontogenic tumor was rendered.

Keywords: Key Words: Benign Tumor Jaw, CEOT, Maxilla, Pindborg

Introduction

Calcifying epithelial odontogenic tumor (CEOT) is a benign tumor of odontogenic epithelium, first described by the Danish pathologist, Dr. Jens J. Pindborg in 1955.¹ CEOT is listed as a benign tumor of odontogenic epithelium with mature fibrous stroma without odontogenic mesenchyme by World Health Organization.² It is a "locally invasive neoplasm with a characteristic feature of having amyloid which tends to calcify".² The origin of CEOT is said to arise from the enamel organ's stratum intermedium in the tooth development stage, a few also believe that they may arise from the remnants of the dental lamina of initial stages of odontogenesis.³

Case Report:

A 50 year old male presented with swelling in the right maxilla of 3 months duration. There was no associated pain or nasal stuffiness. Extra-oral examination showed a mild swelling along the right naso-labial fold. The overlying skin was normal. An intra-oral examination revealed a firm to hard swelling of the anterior maxilla, just right to the midline and at the base of the lateral incisor and canine. The swelling measured about 2.5x1.5 cm and was non-tender. The overlying mucosa and the adjacent teeth were unremarkable. The patient did not give any previous or recent history of tooth extraction or any other dental complaints or procedures.

Imaging revealed a radiolucent lesion in the maxilla with small foci of calcification. There was no evidence of any impacted tooth. A provisional diagnosis of an ameloblastoma was considered. A local excision of the swelling was done

along with extraction of the lateral incisor and canine, and the specimen sent for histopathological examination.

Histopathology, revealed an unencapsulated tumor composed of sheets and islands of polyhedral epithelial cells with moderate to abundant eosinophilic cytoplasm and distinct cell borders. The cells have pleomorphic nuclei, but no mitosis. The surrounding stroma is fibrous to hyalinized. There are numerous amorphous pale eosinophilic rounded masses of amyloid like material, with few showing calcification, and surrounding acute on chronic inflammatory infiltrate (Fig-1). Thus the tumor was diagnosed as CEOT of right anterior maxilla.

Fig.1: Microphotograph showing sheets and islands of polyhedral epithelial cells in a fibrous to hyalinized stroma with presence of numerous amorphous pale eosinophilic rounded masses of amyloid like material, with few showing calcification. (H&E x100)

Discussion

The mean age of presentation is around 40 years of age, with range of 20 to 60 years without any sex predilection, our patient was 50yrs old.⁴ Younger patients have also been reported, recently CEOT has been described in two female children of the ages of 12 & 13 years respectively.⁵ Majority of the cases (95%) are intraosseous, as in the present case,⁶ however a small section of them happen to be extraosseous (gingival). The mandible is known to be twice more commonly affected than the maxilla, with a predilection for the ramus.⁷ In cases affecting the maxilla, the posterior location is more common with the premolar to molar ratio being 1:3.⁸ The present tumor was located in the anterior maxilla at the base of the lateral incisor and canine, which is an uncommon presentation. About 50% of the tumors are associated with an un-erupted tooth, most commonly the third molar.²

Clinical presentation of CEOT varies, from being asymptomatic, accidentally discovered on radiology, to being symptomatic enough for the patient to seek medical help. The patient may have nasal stuffiness and pain, besides the swelling which causes cosmetic problems. Also due to tooth resorption in advanced cases, there may be mobility of involved tooth or tooth tipping.⁹

Histopathologically, CEOT shows a population of polyhedral epithelial cells with distinct borders and intercellular bridges. There is anisonucleosis with pleomorphic nuclei. The surrounding stroma is fibrous to hyalinized with amyloid deposits and concentric lamellated calcifications, called Liesegang's rings. These rings though not exclusive, are quite characteristic of CEOTs. Tumors without calcification have also been reported, and are said to be poorly differentiated with a high rate of recurrence.¹⁰ A histological subclassification has also been proposed, dividing tumors into four patterns based on cell arrangement, calcifications and the amount of amyloid present. However the authors do not claim any prognostic significance of these subdivisions.¹¹

Many variants of CEOT have been reported, namely; clear cell, non calcifying CEOT with Langerhans cells, CEOT with cementum and bone, mixed tumor with ameloblastoma and combined adenomatoid odontogenic tumor and CEOT.¹² Multifocal presentation of CEOT has also been reported, involving both mandible and maxilla in a 50 year male.¹³ Recurrence rates of upto 14% have been seen, with the clear cell variant being the most notorious.¹⁴

Malignant transformations in CEOTs have been reported in literature, with three cases showing evidence of

metastasis. Malignant transformation is determined by the presence of increased and abnormal mitosis, focal necrosis and an increased proliferative index. Aggressive treatment of maxillary CEOTs is recommended, as they are unencapsulated and grow rapidly.⁴ Primary intraosseous squamous cell carcinoma is a close histopathological differential for CEOT.

Conclusion

CEOT though rare, are exclusive to the maxillofacial skeleton. The reported case involves the unusual location of the anterior maxilla, as opposed to the more common posterior mandible. A close follow up is advised in all cases, in view of the invasive nature of the tumor.

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