Pindborg Tumor Involving Maxillary Sinus and Ethmoid Plate in Pediatric Patient: A Case Report

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Abstract: Calcifying epithelial odontogenic tumor (CEOT), also known as Pindborg tumor, is a rare benign tumor deriving from odontogenic epithelium. The mean age at diagnosis is 40.3 years for the central variant and 31.8 years for the peripheral form. We report a case of a 12-years-old female patient with maxillary and ethmoid sinus involvement treated with conservative surgical approach and Leukocyte and Platelet Rich Fibrin (LPRF) regenerative procedures.

Keywords: Odontogenic tumor, CEOT, Pindborg Tumor, Pediatric.

1. INTRODUCTION

Calcifying epithelial odontogenic tumor (CEOT), also known as Pindborg tumor, is a rare, benign but locally aggressive odontogenic tumor, described for the first time by Pindborg in 1955 [1]. CEOT represents less than 1% of all odontogenic neoplasms [2], and approximately 200 cases have been reported until date.

The vast majority of cases occurs at intraosseous level, while peripheral CEOT accounts for less than 5% of cases [3]. Pediatric cases of odontogenic tumors are extremely rare; therefore, we want to describe an interesting case referred at Special and Surgical Stomatology Department, “Ospedali Riuniti” Hospital of Ancona: a 12-year-old girl with sinus localization of CEOT.

2. CASE REPORT

A 12-year-old female patient was reported to Complex Operating Unit of Special and Surgical Stomatology Department (“Ospedali Riuniti” Hospital of Ancona, Italy) for the evaluation of a worsening pain and swelling from upper left posterior region of maxilla for 3 months. No discharge and numbness were associated. Past medical history was insignificant.

Extraoral examination showed a diffuse swelling on the left side of the face with associated facial asymmetry. Palpation confirmed the findings, revealing a non-tender, non-fluctuant and hard swelling. The neck was soft with no evidence of lymphadenopathy or tenderness.

Intraoral examination showed swelling area extending from 2.6 to 2.3, associated with 2.5 element agenesis and persistent 6.5 deciduous tooth. Vitality tests on near teeth revealed positive results and no alterations in color, shape and mobility were reported (Figure 1a).

Radiographic examination was conducted using Cone Beam Computed Tomography (CBCT), revealing a hypodense formation in the left jaw, rounded, with regular contours, of about 3.7 cm in diameter, with 2.5 tooth embedded. Margins of the lesion erode the root of the 6.5 element, the lower wall of the maxillary sinus and the lateral plate of the left ethmoid sinus (Figure 2).

Differential diagnosis included: residual cyst, dentigerous cyst, keratocystic odontogenic tumor, calcifying epithelial odontogenic tumor, ameloblastoma ( multicystic type), and odontogenic myxoma.

To reduce the risk of recurrence, radical surgical resection of the mass with clean margins was conducted. Under general anesthesia, a mucoperiosteal flap was conducted in vestibular surface of left maxilla from 2.7 to 2.1, using piezoelectric bone surgery (Mectron Medical Technology, I-16042 Carasco). Osteotomy of vestibular plate in 2.5 position was performed and after exposition of the cleavage plane, the cystic lesion was enucleated (Figures 1b, 1c). A small fenestration was observed between residual cavity and maxillary sinus. The remaining cavity was filled with platelet-rich fibrin (PRF) (Figure 1d) and sutured using 3-0 silk suture (Figure 1e).

Gross pathology showed a white specimen, including a tooth, measuring 2.7 x 2 cm in size. The cut revealed calcifying areas. Histopathology showed a
neoplasm with solid nests growth pattern, with intracytoplasmic lumen and ductal-like structure. A fibrous capsule enclosing epithelioid-like cells with abundant and eosinophilic cytoplasm. Nuclei were pleomorphic in appearance with rare mitosis.

Large areas of amorphous, eosinophilic, hyalinized material (amyloid-like) were observed and Liesegang ring appearance calcification (Figure 1f). Immunohistochemical analysis revealed positivity for CK19, p63 and bcl2, while CK7 and CK20 were negative.
Proliferative activity, evaluated with Mib-1, was 2%. The histopathological features suggested diagnosis of calcifying epithelial odontogenic tumor.

Post operation period was favorable for the patient; a follow-up CBCT was conducted 1 and 2 years after, revealing the formation of a new alveolar bridge (Figures 3a, 3b).

3. DISCUSSION

We have reported an extremely rare case of pediatric CEOT. This tumor, as in this case, frequently occurs at intraosseous level, while less than 5% of cases can develop within gingival tissues as a nodular mass. There is no sex prevalence, and the onset age range is from first to tenth decade, more frequently in the 20-60
years age group and with a peak of incidence in the fifth decade [4]. Posterior mandibular area is the most frequently involved region by CEOT; moreover, this tumor can be associated with an impacted, displaced tooth or root resorption due to the ability to infiltrate surrounding tissues [5].

The young age, the small dimensions and the presence of well-defined borders justify more conservative approaches, such as enucleation or curettage, followed by judicious removal of a thin layer of bone adjacent to the lesion.

Recurrence free follow-up of the patient make think that conservative approach is an optimal way to treat this tumor [6]. Lesions greater than 4 cm without well-defined radiological margins need to be treated with segmental resections and reconstruction procedures [7], in order to reduce the risk of recurrence, which is about 10-15% [8].

In our experience, PRF has proved to be an optimal strategy to encouraging and speed up healing as we can observe in follow-up CBCT. Several investigations should be conducted to develop a clear surgical protocol for CEOT improving use of piezoelectric bone surgery, a safe and accurate way to perform this treatment. Several studies describe its use in dental therapeutics but no one in CEOT treatment.

In conclusion, even if CEOT typically arises in adult patients, this case demonstrates that also in very young patient can be detected. The piezosurgery conservative enucleation, associated to PRF can be considered the election treatment for limited size lesions.

4. STATEMENTS

This article does not contain any studies with human or animal subjects performed the authors.

Conflict of interest: none declared.

REFERENCES