

## Primordial Odontogenic Tumor: Report of a Case with 10 Years Follow-Up

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### Abstract:

The primordial odontogenic tumor (POT) has recently been classified as a new entity in the WHO Classification of Head and Neck Tumours (2017). POT has been added to the section on benign mixed epithelial and mesenchymal odontogenic tumors. At the time of publication of the WHO classification 7 cases of POT had been published. Due to the small number of cases a new case of POT in a 6-year old male patient is presented. The patient presented with an osteolytic lesion in the anterior maxilla associated with odontoma-like structures that had caused retention of the central incisors. The lesion was surgically removed. Histopathology showed a mesenchymal tissue proliferation surrounded by columnar epithelium. The epithelium showed one or two layers of flat cells. In some areas reverse nuclear polarity in the cores was present. Both clinical and histopathological findings are in accordance with the diagnosis of POT. Because of the young age of the patient he was followed-up for 10 years. At present he is under orthodontic treatment for stimulation of zone growth followed by rehabilitation using dental implants.

**Keywords:** primordial Odontogenic tumour; Odontogenic tumours, dental papilla, inner enamel epithelium.

### Introduction

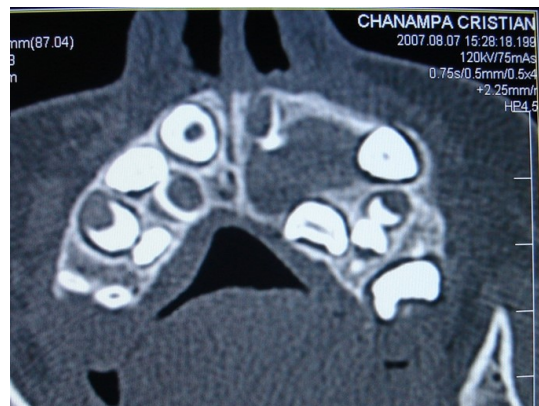
Generally, tooth formation starts at the sixth week of intrauterine life with formation of the dental lamina, bud stage, cap stage, early bell and and late bell stage. During the last phase first lines of mineralized tissue are produced. POT forms similar tissues compared to the dental germ, therefore it has been given its name: primordial odontogenic tumor. POT does not develop mineralized tissues such as dentin or enamel and odontoblastic proliferation in tumor stroma simulating the early bell stage. POT occurs at a mean age of 12.5 years with a range of 3-19 years with no gender predilection (1). POT occurs intraosseously. Most cases are located in the posterior mandible (1, 2). Radiologically, POT appears as a uni- or multilocular osteolytic lesion often associated with a retained tooth (2, 3, 5, 7). Enucleation and curettage is the treatment of choice. To date no recurrences have been reported with follow-up periods of 6 months to 20 years (1).

### Case Report

A 6-year old patient presented with non-erupting maxillary central incisors. Panoramic radiography revealed a small odontoma-like structure associated with the right maxillary incisor interfering with its eruption. A radiolucent lesion was associated with the left maxillary incisor (Fig.1). Computer tomography confirmed the presence of a radiodense structure palatally to the right central incisor. A small dentigerous cyst-like lesion associated with the left central incisor had eroded the vestibular bone plate. The capsular structure showed a higher radiodensity and a peripheral osteogenic reaction was also noted (Fig. 2). At the site a small odontoma-like structure was also present.



**Figure 1:** Panoramic radiography showing retention of both maxillary central incisors associated with odontoma-like structures and an osteolytic lesion adjacent to the left maxillary central incisor.



**Figure 2:** Computer tomography (CT) which shows a dentigerous cyst-like lesion associated with the left maxillary central incisor with slight erosion of the vestibular bone plate (see arrows).

Under general anesthesia enucleation of the right central incisor and the palatal odontoma as well as removal of the left central incisor including the cystic lesion and the calcified structure of small size was performed. The surgical defect was treated by guided bone regeneration and was filled with granulated tricalcium phosphate. Healing was uneventful. Postoperatively, orthodontic was indicated.

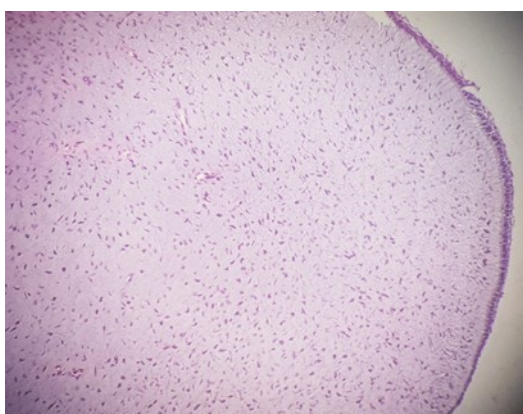
Histopathology revealed a compound odontoma of the right side. Macroscopically (Figs. 3 and 4), the cyst-like lesion was of cartilagenous consistency. Nodular proliferation of a tissue similar to the dental papilla with a myxomatous tissue with spindle cells surrounded by a columnar epithelium similar to the internal dental epithelium with reverse nuclear polarity was found. The periphery of the tumor was covered by columnar or cuboidal epithelial cells, a characteristic finding for POT (Figs. 4 - 6). The capsule was formed by connective tissue.



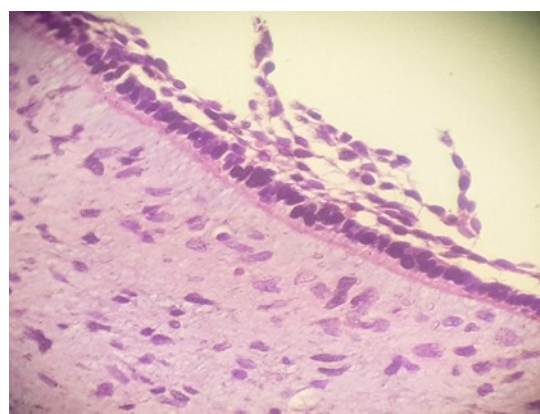
**Figure 3:** Macroscopic view of the odontoma-like lesion with a diameter of 25 mm



**Figure 4:** Macroscopically, the cyst-like lesion was of cartilagenous consistency no evidence of cystic changes.

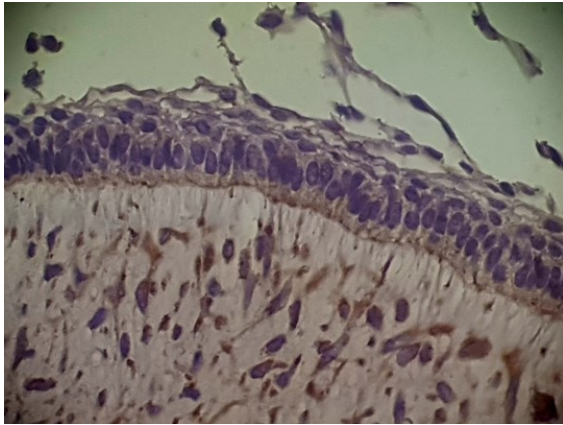


**Figure 5:** Tumor lesion with proliferation of loose fibrous tissue surrounded by columnar epithelium with reverse nuclear polarization. H&E x 40.

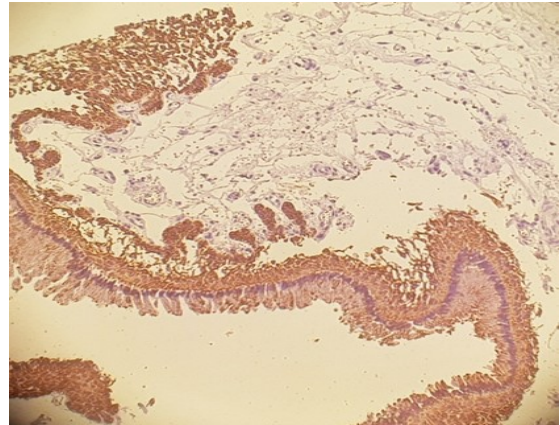


**Figure 6:** Surface epithelium which shows reverse nuclear polarization. H&E x 1000

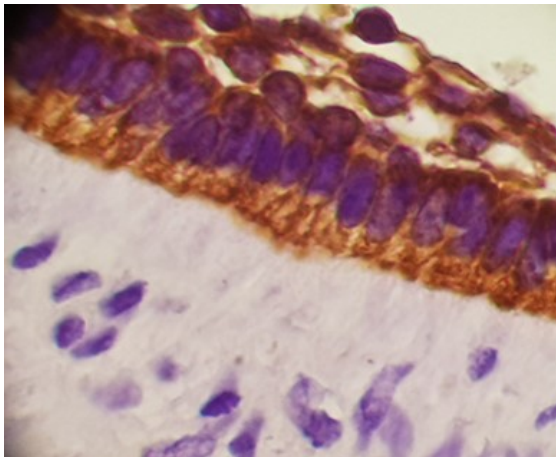
Immunohistochemistry showed positive reactions of epithelial cells for cytokeratins 5 and 18 (7 and 14 were also positive but are not shown in figures) (Figs. 7 - 9). Mesenchymal tissues were positive for vimentin (Fig. 10 and 11). Ki-67 was found in isolated cores at the epithelial level (Fig. 12).



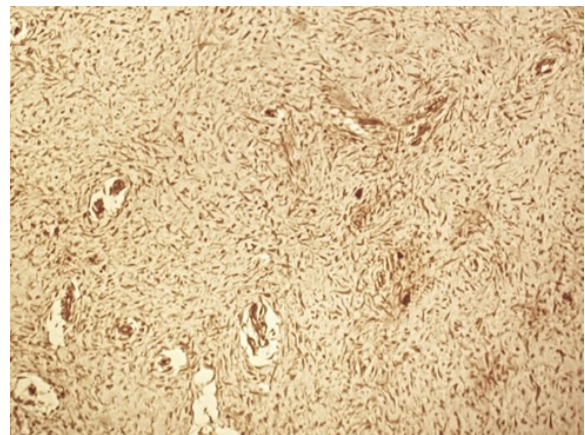
**Figure 7:** Overexpression of CK5 in surface epithelium of POT; x 1000.



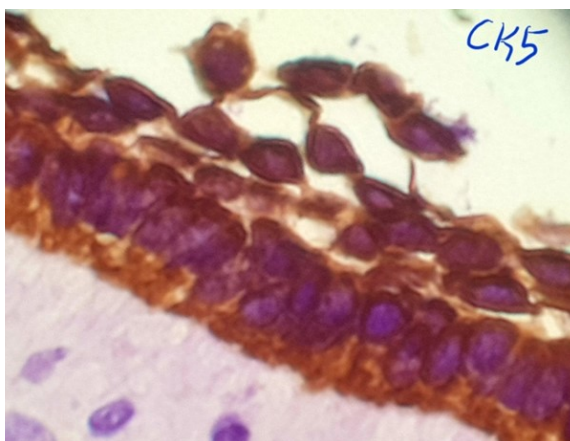
**Figure 8:** Cytokeratin 18 expression in surface epithelium of POT; x 400.



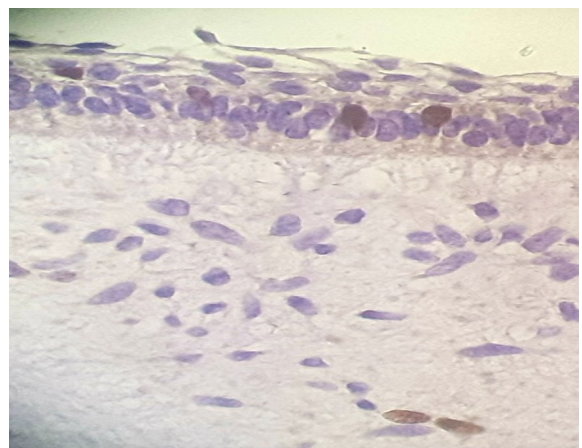
**Figure 9:** Cytokeratin 18 expression at epithelial level; x 1000



**Figure 10:** The mesenchymal component of the tumor was positive for Vimentin



**Figure 11:** Staining for vimentin is positive in mesenchymal stroma at basement membrane; x 1000.



**Figure 12:** Ki-67 indicates low activity in the epithelium of POT; x 1000.

## Discussion

The POT has been added to the WHO Classification of Head and Neck Tumours in 2017 as a new entity of benign mixed epithelial and mesenchymal odontogenic tumours (1). POT has been defined as „a tumour composed of variably cellular loose fibrous tissue with areas similar to the dental papilla, entirely surrounded by cuboidal to columnar epithelium resembling the internal epithelium of the enamel organ“(1). At present the number of POT cases reported in the literature is still very small (1). POT formation corresponds to early tooth formation in which the dental germ in its early stages develops from bud stage to cap stage and early bell stage. The latter stage is characterized by presence of a loose mesenchymal (ectomesenchymal) tissue with spindle cells and blood vessels. The entire mesenchyme (ectomesenchyme) is surrounded by an enamel organ characterized by presence of a layer of columnar cells with reverse nuclear polarization (internal dental epithelium) followed by a layer of spindle cells in the intermedium stratum followed by the stellate reticulum (named by its star-shaped cell appearance). A superficial layer of cuboidal cells – the outer dental epithelium – envelops the enamel organ. At this stage no dentin or enamel has been formed as yet.

Histopathology of POT is characterized by loose fibrous tissue with fusiform or stellate fibroblasts. POTs are covered by columnar or cuboidal epithelium. In some areas superficial layers of fusiform cells may be found with a thin fibrous capsule. Epithelial strands or islands may be detected within POTs as a result of tangential sectioning (1). Immunohistochemical studies have shown positive staining for pancytokeratins (AE1/AE3, CK5 and CK14. CK 19 is expressed by columnar epithelium (1, 4, 8, 9). Mesenchyme is positive for vimentin. The Ki-67 index is very low (>2%) (1).

The histopathologic differential diagnosis may include ameloblastic fibroma, central odontogenic fibroma, hyperplastic dental follicle and odontogenic myxoma. Also it should be considered that differential diagnosis of POT might include immature soft tissue odontoma and soft tissue odontoma with marked papillomegaly. In ameloblastic fibroma strands and islands of odontogenic epithelium within the cellular stroma. While strands and islands of epithelium may also be observed in POTs due to tangential sections POTs are covered by epithelium on their surface. The central odontogenic fibroma reveals nests of primitive odontogenic epithelium within a fibrous stroma but does not have any surface epithelium. The odontogenic myxoma does not have epithelial structures within its loose myxomatous structure. The hyperplastic dental follicle is characterized by fibroblastic proliferations and islands of inactive odontogenic epithelium (10).

As in the present case impaction of teeth with follicular cyst formation is a common finding in POTs. Usually, however, the mandibular third molar area is mostly affected compared to the present case in which the maxillary central incisors were involved. The age of 6 years of our patient fits the age range of 3 to 19 years (1). Odontoma-like structures in the present case have not been reported so far in POTs, however, the maxillary incisor area is well known for the common formation of odontomas compared to the posterior mandible (10).

POTs representing a benign pathology seem to have no tendency for recurrence because of a rather low index of cellular proliferation with Ki-67 being less than 2% (1). In contrast ameloblastomas have a high tendency for recurrence, although their Ki-67 index is also low. Therefore, some authors have suggested that Ki-67 and PCNA index proliferation are not viable for POTs and its odontogenic benign tumor evolution (4-10).

The discussion whether or not POT is a novel and neoplastic lesion or not is still not decided at this point. From the point of view of the present authors additional research on POT should be published before final conclusions concerning the biological and behavioral profile of POT can be drawn.

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