



Research Paper

Primordial Odontogenic Tumor in a Pediatric Patient; a unique case report, 2nd from India and 19th reported world over.

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Abstract: Primordial odontogenic tumor (POT) was recently recognized in the 2017 World Health Organization classification as a distinct mixed odontogenic tumor, with only 18 cases reported to date. This is the 19th reported case of POT in the world and 2nd from India. Diagnosed in an 8-year-old boy, this case shows unique radiographic features of the tumor encircling the developing 1st premolar of the second quadrant, extending into the maxillary sinus, having notable opacification, inducing buccal and lingual cortical expansion and root resorption of deciduous teeth of the region. Macroscopically, the tumor was an encapsulated, slippery, solid mass and microscopically composed of dental papilla like myxoid fibrous connective tissue, surrounded by a single layer of tall columnar epithelial cells with reverse nuclear polarization. Provisionally diagnosed as an ameloblastic fibroma, the tumor was enucleated as a whole.

Key-words: Primordial odontogenic tumour, Opacification, Developing 1st Premolar

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I. Introduction:

Primordial odontogenic tumor (POT), first reported by Mosqueda-Taylor et al. in 2014^[1] is a new entity, classified as a benign mixed epithelial odontogenic tumor in the WHO classification of Head and Neck Tumors, 2017.^[2] Owing to its rarity, evidence regarding its etiopathogenesis and clinical behaviour remains largely unknown. It is typically seen in the posterior mandible region, with only 3 reported cases of the maxilla.^[3] This report is about a case of POT in an 8-year-old boy, involving the developing 1st premolar in the left maxilla and also having notable radiographic opacification, making it an even rarer entity.

II. Case History:

An 8-year-old boy was referred to the Department of Pediatric and Preventive Dentistry of Government Dental College, Thiruvananthapuram, with the chief complaint of a swelling of the left upper jaw region, noticed about 3 months prior. Even though largely asymptomatic, a gradual increase in size of the swelling had occurred from the time since it was first noticed. There was no relevant medical/dental history or that of any traumatic dental injuries. Extra oral palpation revealed a bony hard swelling over the left malar region, extending from the ala of the nose to the zygoma (figure-1). Intra orally, a non-tender bony hard swelling extending from the infraorbital region superiorly and obliterating the buccal vestibule from 62 to 26 with pathological mobility of 63, 64 and 65 were noted (figure-2). Panoramic imaging (figure-3) and cone beam computed tomography (figure-4) revealed a well-defined, large, expansile, unilocular lesion of size 30×30 mm in the left maxilla, extending antero-posteriorly from the distal aspect of 62 to the mesial aspect of developing tooth bud of 27 and apical to 26. The lesion was involving the anterior and mesial walls and roof of the left maxillary sinus, extending superiorly to the orbit and inferiorly to the radicular region of 63, 64 and 65. Opacification was noted, except for the superior most region of the left maxillary sinus. The buds of 23, 24 & 25 appeared displaced, with 23 being pushed mesially towards the nasal floor, 24 superiorly towards floor of the orbit and 25 distally towards the periapical region of 26. Pathologic root resorption of 63, 64, 65 and buccal cortical expansion of the segment were also seen (figure-5). Diagnostic hypothesis from radiology was that of a dentigerous cyst with respect to developing 24, because of its location and radiologic features. Aspiration yielded a clear cystic fluid mixed with blood (figure-6).

Even though opacification was noticed, marsupialization of the lesion was planned, along with extractions of mobile teeth, because the aspirated fluid from the lesion showed very few inflammatory cells and

also the CBCT report, both being suggestive of a dentigerous cyst developing from 24. But, after extraction of 63, 64 and 65 the surgical site revealed a slippery movable mass that appeared whitish yellow (figure-7). An incisional biopsy of the mass, with 3 bites were sent for histopathological examination (figure-8), initial diagnosis made from the specimen, was that of a dentigerous cyst undergoing ameloblastomatous transformation or an ameloblastic fibroma, because of its microscopically shared common histological features. An excision of the lesion was planned and done under General Anaesthesia (figure-9), along with the extraction of buds of 23, 25 (displaced towards periphery to the lesion) and 62 with pathologic root resorption. The bud of 24 was embedded within the excised lesion (figure-10). After thorough curettage, iodoform+glycerine paste was impregnated along with gauze and buccal and labial flaps were sutured with 4-0 vicryl suture. The pack was changed every 3 days for 2 weeks and the patient was under observation for 3 months. Wound healing was good (figure-11).

The excised lesion (figure-10), which was a large encapsulated mass, white in colour, intermingled with brown areas, with a smooth surface, firm in consistency and of a similar size as seen on imaging, was sent for histopathological examination. Histopathology, confirmed the lesion as a primordial odontogenic tumor, involving the developing 24.

III. Discussion:

Based on the information in the English-language literature ^{[1],[4],[5],[6],[7]} and the latest WHO classification, ^[2] the clinical, radiographic and microscopic features of this case fulfil the diagnostic criteria of a primordial odontogenic tumor (POT). The age of this patient and that of previously reported cases having a mean age of 11.5±6.15 years, suggests that this tumor occurs exclusively in young age. ^[8] Male to female ratio was 10:7, suggestive of a slight male predilection. ^[3] The mean largest diameter of POT was 42.54mm ± 24.42mm, mainly occurring in the posterior mandible, ^[7] with only 3 cases of the maxilla reported till date. ^[3] This is the 19th reported case of POT world over, 4th of the maxilla and 2nd from India. Previously reported cases were associated with molars and had well-defined unilocular radiolucencies, resembling a dentigerous cyst. ^[7] The presenting case was associated with an unerupted 1st premolar and had notable opacification. Importantly, this is only a second case of POT demonstrating a radiopaque focus. ^[5] Buccal cortical plate expansion and the root resorption of deciduous teeth were remarkable, like previous reports.

Histopathological examination was consistent with the previous reports of POT and described a highly cellular, myxomatous, delicately fibrillar connective tissue stroma with loosely arranged cells resembling dental papilla, ^[1] lined by a single layer of cells which was tall columnar with hyperchromatic nuclei showing reversal of polarity and subnuclear vacuolation. ^{[1],[4],[9]} In a few areas, polygonal to squamous cells were seen superficial to the tall columnar cells, without proliferation in the connective tissue. ^[4] A fibrous connective tissue capsule was seen surrounding the tumour. ^{[1],[10]}

POT is a benign neoplasm and can be treated by simple enucleation. Although POTs may show a variation in radiographic features, it should be included in the differential diagnoses of lesions with unilocular radiolucencies, irrespective of any opacification, particularly when it is associated with developing teeth buds. As a POT is a less reported and new entity, further understanding of its nature will depend on future reports and studies. Importantly, a Pediatric dentist must be aware of this new and rare tumour in order to avoid a misdiagnosis and inappropriate treatment.

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Legends

Figure-1; extra oral swelling involving the left malar region

Figure-2; intra oral bony hard swelling extending from 62 to mesial aspect of 26

Figure-3; panoramic radiograph showing a well-defined unilocular radiolucency with displaced buds of 23,24,25 and resorbed roots of 63,64,65

Figure-4; CBCT OF Maxilla, showing involvement of the left maxillary sinus and opacification

Figure-5; CBCT axial view of maxilla showing buccal cortical expansion

Figure-6; clear fluid mixed with blood, aspirated from the lesion

Figure-7; extraction site of 64,65 exposing the lesion

Figure-8; bites of incisional biopsy

Figure-9; lesion exposed after buccal flap reflection and cortical plate removal

Figure-10; excised lesion along with embedded 24 and extracted 23,25

Figure-11; uneventful postoperative healing and erupting 21