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Case Report

Primordial odontogenic tumor- rare and novel entity: A case report

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ABSTRACT

The primordial odontogenic tumor (POT) is a recently described benign entity with histopathological and immunohistochemical features suggesting its origin during early odontogenesis that has been included in the World Health Organization classification 2017. Only 27 cases that conform to the clinico-pathologic criteria for diagnosis have been reported worldwide. We report an case of POT in a 9-year-old child patient who presented with diffuse swelling in pre-auricular region. Large expansile osteolytic lesion involving inverted 47 causing erosion of superior margin with persistent expansion of buccal and lingual cortical plate was observed radiographically. Along with the present case, we also present here a summary of the cases reported till date and the review of literature.

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1. Introduction

Primordial Odontogenic Tumor (POT) is a rare and novel entity first described by Mosqueda- Taylor et al.¹ which has been incorporated in the latest WHO classification² under benign mixed epithelial and mesenchymal odontogenic tumor. The name Primordial Odontogenic Tumor (POT) was suggested because of its location and process of odontogenesis imply that this lesion may originate from the early stage of tooth development.³ Majority of the cases predominantly occur in young patients in the first and second decades of life with mandible being the most common site.⁴ POT appears as a well-defined unilocular or multilocular radiolucent lesion adjacent to the crown of an unerupted tooth with asymptomatic bony swelling causing root resorption and buccal or lingual cortical expansion.⁵

Histopathologically, it is characterized by a variably cellular immature connective tissue stroma with dental

papilla like areas covered by cuboidal or columnar epithelium resembling the inner enamel epithelium of the enamel organ.^{6,7} So, far 27 cases worldwide and 6 cases in India have been reported in published literature (Table 1). Here we present a new case of developing POT at right ramus of mandible with emphasis on its biological analysis.

2. Case Presentation

A 9-year old male patient was reported to the Out Patient Department (OPD) of a tertiary care centre with primary complaints of swelling in the right pre-auricular region persisting for five months with no associated pus discharge. Patient reported a history of swelling 5 months back with associated mild, non-radiating pain and fever which resolved after taking non-steroidal anti-inflammatory medications. The patient underwent Ultrasonography (USG) Neck which revealed large lytic lesion suggestive of osteomyelitis in right ramus of mandible. Further Magnetic Resonance Imaging (MRI) revealed large expansile lesion in

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right ramus of mandible with cortical erosion and associated cervical nodes. Patient was advised ATT for 3 weeks after that there was no change in reduction in size. The patient was fully oriented to time, place and person without any underlying systemic disease.

Extra-oral examination revealed a diffuse swelling in the right preauricular area with palpable level II and III lymph nodes. On exploration -multiple osteolytic bone lesion was present in right ramus over lingual aspect of 47,48. CBCT was advised which depicted osteolytic lesion involving inverted 47 causing erosion of superior margin wrt Inferior alveolar nerve canal with persistent expansion of buccal and lingual cortical plate in 46,47 regions (Figure 1).

Based on radiographic examination, a provisional diagnosis of odontogenic tumor was made. Case was planned for curettage and biopsy under General Anesthesia. Via Intra-oral approach, incision was given buccal to ascending border of ramus. Curettage and biopsy was done from osteolytic lesion. (Figure 2A, B). After achieving haemostasis, primary closure done using a gelatin sponge. Medications were advised for 5 days and the patient was recalled for follow up.



Figure 1: Pre-Op CBCT showing lytic defect

The gross specimen shows multiple grey white soft tissue and hard tissue bits together measuring 12.5x1.8x0.4 cm. Histologically, sections showed tumor composed of loose fibrous to myxoid tissue with numerous fusiform and stellate fibroblast- resembling odontogenic ectomesenchyme. The surface was lined by discontinuous cuboidal to columnar epithelium resembling inner enamel epithelium, the features were suggestive of a benign mixed epithelial and mesenchymal odontogenic tumor- possibility

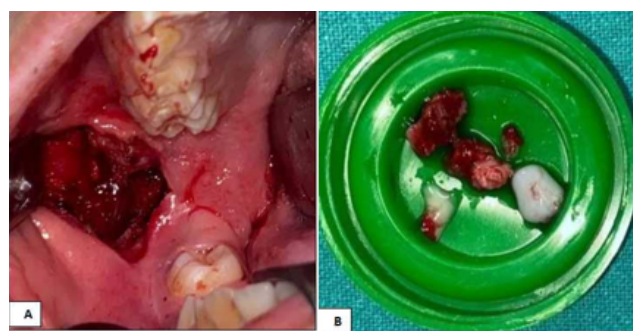


Figure 2: A: Intra-op procedure; B: Gross of the enucleated specimen

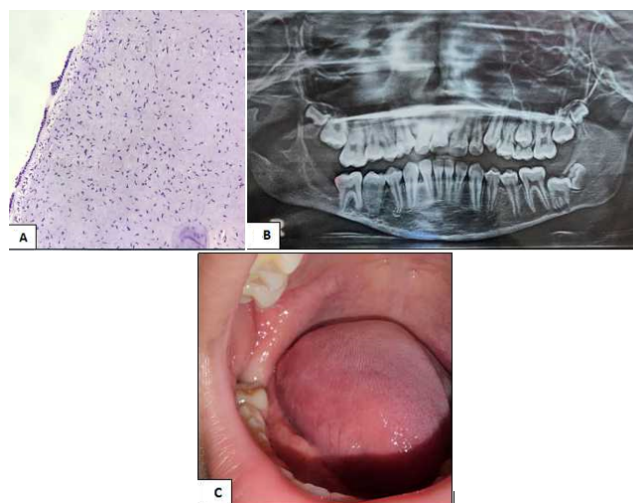


Figure 3: A: Photomicrograph showing thin lining; B: Post op follow up imaging epithelium and the underlying stroma (HE 100x); C: Post op follow up intra-oral picture

of Primordial Odontogenic Tumor (POT) with associate dense acute on chronic inflammatory granulation tissue (Figure 3A). Final diagnosis of POT was made and the patient has been followed up without any any signs and symptoms (Figure 3B,C)

3. Discussion

POT is a new neoplastic entity, classified as benign mixed odontogenic neoplasms in the fourth edition of WHO Head and Neck book in 2017.² First described by Mosqueda-Taylor et al. in their series of six cases, these authors reported a new odontogenic lesion that did not fit or fulfill the diagnostic criteria in any category of odontogenic tumors described before ¹. According to the existing literature, POT may be defined and described as a benign odontogenic tumour that develops and is diagnosed during the first two decades of life.^{1-7,9,11} It is worth noting that the geographic regions of the POT cases were mainly located in North and South America (68.8%).⁵ Till date

Table 1: Summary of previously reported cases of primordialodontogenic tumour

Case Number	Reference	Age (Year)	Gender	Location	Involved Tooth	Treatment	Follow up	Recurrence
1	Mosqueda-Taylor et al., ¹ 2014	18	M	Man	Unerupted third molar	Enucleation & tooth extraction	20 years	No
2	Mosqueda-Taylor et al., ¹ 2014	16	M	Man	Unerupted third molar	Enucleation & tooth extraction	13 years	No
3	Mosqueda-Taylor et al., ¹ 2014	16	M	Man	Unerupted third molar	Enucleation & tooth extraction	10 years	No
4	Mosqueda-Taylor et al., ¹ 2014	3	F	Man	Unerupted second deciduous molar	Enucleation & tooth extraction	9 years	No
5	Mosqueda-Taylor et al., ¹ 2014	13	F	Man	Unerupted third molar	Enucleation & tooth extraction	3 years	No
6	Mosqueda-Taylor et al., ¹ 2014	3	F	Max	Unerupted second deciduous molar	Enucleation & tooth extraction	6 months	No
7	Slater et al., ⁶ 2016	19	M	Man	Unerupted third molar	Excision & tooth extraction	7 months	No
8	Mikami et al., ⁸ 2017	5	M	Man	Unerupted second deciduous molar	Excision & tooth extraction	7 months	No
9	Amer et al., ⁹ 2018	2	M	Man	Unerupted tooth	Excision & tooth extraction	2 years	No
10	Pardhe et al., ¹⁰ 2018	17	M	Man	Unerupted third molar	Enucleation & tooth extraction	6 months	No
11	Almazyad et al., ¹¹ 2018	15	F	Man	Unerupted third molar	Excision & tooth extraction	3 months	No
12	Almazyad et al., ¹¹ 2018	18	M	Man	Unerupted third molar	Eurettage & tooth extraction	20 months	No
13	Bomfim et al., ⁴ 2019	4	M	Man	Unerupted second deciduous molar	Excision & tooth extraction	—	No
14	Almazyad et al., ¹¹ 2018	19	F	Man	Unerupted third molar	Excision & tooth extraction and hemimandibulectomy after recurrence	4 years	Yes
15	Kayamori et al., ¹² 2021	10	M	Max	Unerupted tooth	Excision & tooth extraction	30 months	No
16	Naina et al., ¹³ 2021	14	M	Max	Unerupted canine	Excision & tooth extraction	3 years	No
17	Ando et al., ¹⁴ 2017	8	F	Max	Unerupted first deciduous molar	Enucleation	16 months	No
18	Suresh et al., ⁷ 2023	03	F	Man	Unerupted first deciduous molar	Incisional biopsy	—	No

only six cases have been reported in India according to the published literature, ours will be the seventh case. The majority of the cases were diagnosed in young patients with age range 2-19 years old. All cases occurred in the posterior region of the jaw, with the mandible (87.5%) being the main anatomic site of occurrence.^{10,14} All cases reported were asymptomatic and presented an expansion of cortical bone. The present case showed a rare location with a small tumor, whereas clinical, radiological, and pathologic findings are similar to those of the previous reported cases. The location of the tumor was near right ramus of mandible with cortical erosion Radiographically, POT appears as a well-defined radiolucent unilocular lesion always associated with unerupted teeth, such as deciduous or third molars, with a mean size of 4.1 cm, showing tooth displacement and

frequent root resorption.^{9–11,13,14}

Microscopically, POT is a solid, multilobulated whitish and glossy mass with no evidence of cystic changes, and this tumor is well demarcated from the surrounding structures. Considering these aspects, it is convenient to rule out the possibility of ameloblastic fibroma in excision specimens and of odontogenic myxofibroma in incisional biopsies.¹² Histologically, POT is characterized by variably cellular to loose fibrous tissue with areas mimicking dental papilla, being circumscribed by epithelial cells showing morphologies ranging from cuboidal to columnar. In our case, these microscopical findings were also observed as demonstrated in the images present in the manuscript.⁸

The prognosis of all cases of POT was excellent after surgery, except for two cases which were lost

to followup, recurrences of all reported cases have not been reported to date (median follow-up years 4.53 ± 6.09 , ranging from 3months to 20years).^{5–7,10–13}. In majority of cases, enucleation followed by tooth extraction was done. However, in our case, the patient underwent curettage and biopsy and until now after curettage there was no recurrence either. It seems that curettage and extraction of involved tooth were effective treatments because the peripheral columnar epithelium or fibrous pseudocapsule of the tumor clearly delimited the boundaries of the tumor from adjacent tissues.

4. Conclusion

This case report highlights the characteristics and management of Primordial Odontogenic Tumor (POT), a rare benign jaw lesion predominantly affecting young individuals. With distinct radiological and histopathological features, early recognition is essential for appropriate treatment. Surgical interventions, such as enucleation or curettage, have shown promising outcomes in preventing recurrence. Continued documentation and analysis of cases are crucial for refining diagnostic and therapeutic strategies, ultimately improving patient care for this intriguing odontogenic tumor.

5. Conflict of Interest

None.

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None.

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