Case report

A unique case report of primordial odontogenic tumor

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Primordial odontogenic tumor (POT) has been included as a new entity in the latest WHO classification (2017). Only 17 cases have been documented worldwide, and here, we report the eighteenth case – the second in Europe and first in Poland. The study describes a case of a 16-year old boy, diagnosed with primordial odontogenic tumor. The patient was treated with enucleation and there was no recurrence in the follow-up period.
Due to the small number of cases, further reports are crucial to obtain pathomorphological and clinical characteristics of primordial odontogenic tumor.

Key words: primordial odontogenic tumor, odontogenic tumors, jaw tumors, mandible.

Introduction

The classification of head and neck odontogenic tumors is subject to constant change. More and more disease entities, distinguished on the basis of unique morphological features and processes, are added, while subtypes and variants of odontogenic tumors that are difficult to distinguish disappear. The latest version of the WHO classification from 2017 introduces two previously undescribed changes: sclerosing odontogenic carcinoma (SOC) and primordial odontogenic tumor (POT) [1, 2, 3]. Primordial odontogenic tumor (POT) belongs to the group of benign mixed epithelial and mesenchymal odontogenic tumors, due to its loose fibrous connective tissue and the cuboidal or columnar epithelium that surrounds it. Thus, the histological structure of the tumor resembles the internal epithelium of the enamel-forming organ. Randomly placed fibroblasts produce a small amount of collagen [1, 4]. There are numerous blood vessels in the abundant cellular stroma, providing a nourishing environment.

Analyzing the immunohistochemical profile of the tumor, Molina *et al*. suggest that the beginnings of its development are associated with the early stages of tooth germ formation. The presence of CK14, CK19 and amelogenin seems to be crucial in this theory [5]. The research of Mosqueda-Taylor *et al*., published long before the inclusion of POT in the WHO classification, describes 6 cases of primary odontogenic tumors. Unable to classify the changes into any of the categories at the time, the authors created the first description of the new disease entity [6]. To date, English-language literature describes 17 cases of POT. The present case report is the second in Europe and the first in Poland.

Case report

Based on a panoramic X-ray a 16-year-old patient was admitted to the Oral Surgery Department of the Medical University of Lublin due to an incidental finding in the right posterior mandible. The resorption of mandible cortical bone and extensive, multi-chamber osteolytic defect between teeth 45 and 46 measuring 20 mm × 15 mm was described in the performed CBCT imaging (Figs. 1, 2). The germ of a supernumerary tooth was visible inside the lesion. Adjacent teeth did not show any signs of resorption. The course of the inferior alveolar

Table I. Features of all POT published cases

Case number	Location	Age	Gender	Manifestation	Radiograph dimension [mm]	Radiograph appearance	Treatment method	Recurrence	Follow-up time	Author
1	Mandible	18	M	Asymptomatic	45 × 40	Radiolucent, unilocular	Enucleation with involved teeth	NO	20 years	Mosqueda-Taylor *et al.*
2	Mandible	16	M	Asymptomatic	55 × 50	Radiolucent, unilocular	Enucleation with involved teeth	N/A	Lost to follow-up	Mosqueda-Taylor *et al.*
3	Mandible	16	M	Asymptomatic	65 × 50	Radiolucent, unilocular	Enucleation with involved teeth	NO	10 years	Mosqueda-Taylor *et al.*
4	Mandible	3	F	Asymptomatic	90 × 70	Radiolucent, biloculated	Enucleation with involved teeth	NO	9 years	Mosqueda-Taylor *et al.*
5	Mandible	13	F	Asymptomatic	80 × 50	Radiolucent, biloculated	Enucleation with involved teeth	NO	3 years	Mosqueda-Taylor *et al.*
6	Maxilla	3	F	Asymptomatic	35 × 30	Radiolucent, unilocular	Enucleation with involved teeth	NO	6 months	Mosqueda-Taylor *et al.*
7	Mandible	19	M	Asymptomatic	N/A	Radiolucent, unilocular	Enucleation with involved teeth	NO	7 months	Slater *et al.*
8	Mandible	15	F	Asymptomatic	35 × 20	Radiolucent, multilocular	Enucleation with involved teeth	NO	3 months	Almazyad *et al.*
9	Mandible	18	M	Asymptomatic	17 × 12	Radiolucent, unilocular	Curettage and tooth extraction	NO	20 months	Almazyad *et al.*
10	Maxilla	8	F	Asymptomatic	N/A	Radiolucent, unilocular	Enucleation	NO	16 months	Ando *et al.*
11	Mandible	5	M	Asymptomatic	N/A	Radiolucent, unilocular	Enucleation with involved teeth	NO	7 months	Mikami *et al.*
12	Mandible	17	M	Asymptomatic	N/A	Radiolucent, multilocular	Enucleation with involved teeth	NO	6 months	Pardhe and Bajpai
13	Mandible	4	M	Asymptomatic	30 × 20	Radiolucent, unilocular	Enucleation with involved teeth	N/A	Lost to follow-up	Bomfim *et al.*
14	Mandible	2	M	Pain	30 × 40	Radiolucent, multilocular	Enucleation with involved teeth	NO	2 years	Amer *et al.*
15	Mandible	10	M	Asymptomatic	5 × 5	Radiolucent, unilocular	Enucleation	NO	12 months	Sun *et al.*
16	Mandible	13	F	Asymptomatic	N/A	Radiolucent, unilocular	Enucleation with involved teeth	N/A	N/A	Teixeira *et al.*
17	Mandible	N/A	M	Asymptomatic	22 × 20	Radiolucent, unilocular	Enucleation	N/A	N/A	Berdugo and Bilodeau
18	Mandible	16	M	Asymptomatic	20 × 15	Radiolucent, multilocular	Enucleation with involved teeth	NO	12 months	Present case

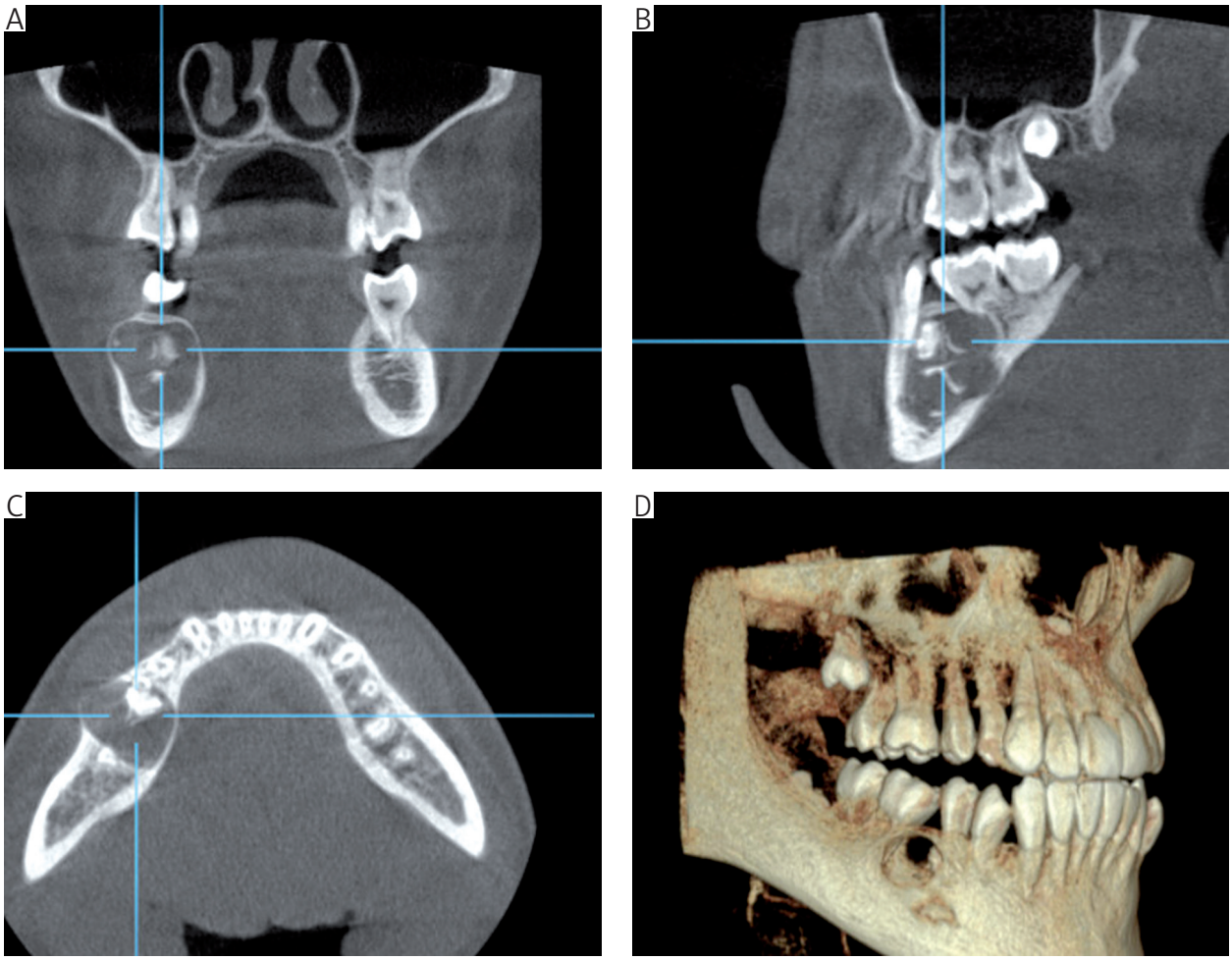

Fig. 1. Preoperative cone beam tomography pseudo 3D reconstruction revealed osteolytic radiolucency at right posterior mandible

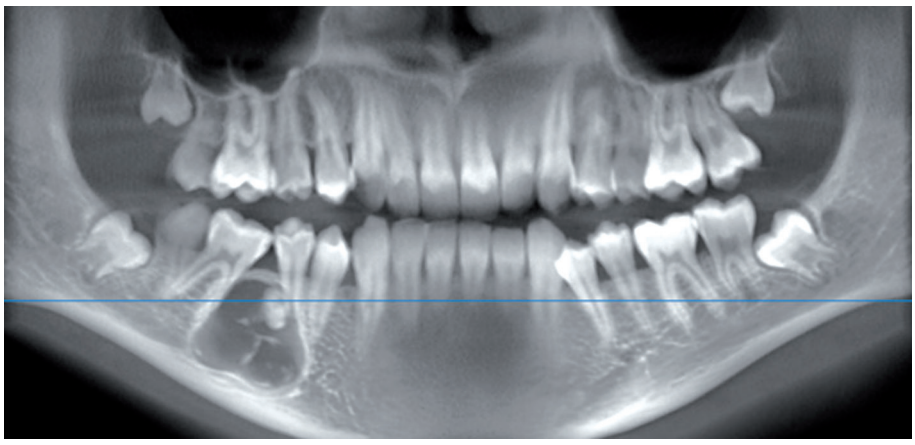

Fig. 2. Preoperative panoramic radiograph

nerve canal was distalized with preserved bone border. The patient did not report any symptoms related to the lesion. In the extraoral examination, no abnormalities were found. Intraorally, in the vestibule of the oral cavity, there was a slight bone dilation around 46, covered with unchanged mucosa. Submandibular lymph nodes were not palpable. The patient denied any systemic diseases and denied taking any medications. Due to the lack of cooperation, he was qualified for enucleation of the tumor under general anesthesia. Initial diagnosis was made: cystis follicularis. Under general endotracheal anesthesia,

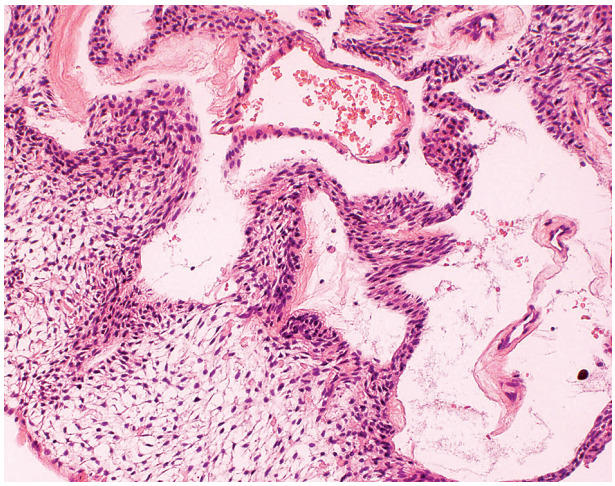

Fig. 3. Cystic multi-chamber nature of the lesion (HE, magnification 40×)

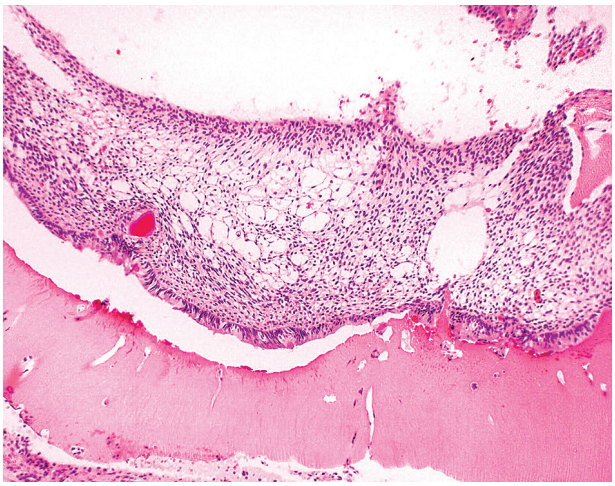

Fig. 6. General view of the lesion with the bone fragment (HE, magnification 20×)

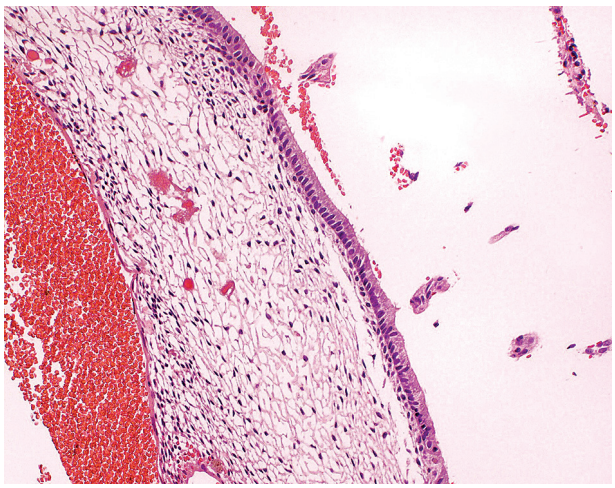

Fig. 4. Lining glandular epithelium, regular, cell nuclei perpendicular to the basement membrane and loose connective tissue stroma (HE, magnification 40×)

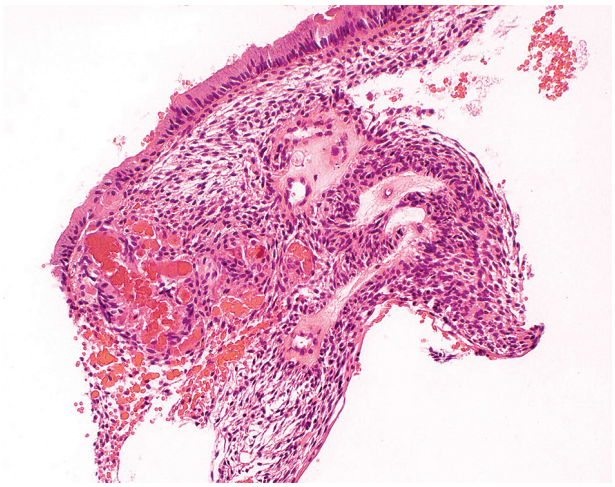

Fig. 5. Cell abundant stroma with numerous blood vessels, covered with regular glandular epithelium (HE, magnification 20×)

aspiration puncture was performed. A turbid, white content, which did not resemble typical odontogenic cyst fluid, was obtained. After incision and elevation of the triangular mucoperiosteal flap, the alveolar processes and mandibular body of region 44-46 were visualized. Cortical bone with a diameter of about 7 mm was found to be damaged. A solid, multi-chamber change was enucleated. Supernumerary tooth 45′ was removed. The cavity was rinsed with antiseptic. The flap was deposited and sutured with Monosyn 4.0 resorbable sutures. The obtained tissue material was sent for histopathological examination in 10% formalin solution. The result of the examination – primordial odontogenic tumor – fulfilled the clinical criteria of this lesion. During the follow-up appointment, normal healing of the wounds was observed. Tooth 46 and 45 vitality was checked using ethyl chloride, obtaining a positive reaction. Gradual filling of the cavity and correctly reproduced bone tissue were observed on both 6- and 12-month post-op panoramic radiographs. There were no signs of recurrence.

Discussion

Primordial odontogenic tumor affects patients at a young age (min. 2, max. 19 years) [7, 8, 9]. It does not have a sex predilection, and most often occurs in the mandible (88.89%), with only two case reports showing an association with the maxillary bone [4, 10]. It is usually associated with the impacted third inferior molar. Our case is the first to involve an impacted supernumerary tooth. Due to the asymptomatic course (pain symptoms were described only in the case presented by Amer *et al*. and were combined with earlier marsupialization and the young age of the patient), it is incidentally found on X-rays [8]. Cone beam computed tomography (CBCT) is crucial

in the correct diagnosis and treatment planning. The radiological image is described as an unilocular, less often multi-chamber, radiolucent lesion with associated impacted tooth. The classification proposed by Sun *et al*. organizes the relationship between the tumor and the associated tooth: type A – the POT has a pericoronal location in a dentigerous relationship; type B – the tumor appears to completely envelop an embedded tooth; and type C – the POT is in close proximity to the root of the tooth. Our case best meets the criteria of type B according to Sun *et al.* both in terms of tumor diameter and its location in relation to the germ of supernumerary tooth 45′ [11]. Due to the benign nature of the POT, simple enucleation of the tumor with the associated tooth and possible filling of the bone defect with bone substitute material seems to be an appropriate therapeutic approach. The authors strongly reject extending the procedure to include peripheral radicalization such as segmental resection of the mandible or the use of Carnoy's solution as a method of surgical treatment, describing it as too mutilating to the patient. In all presented cases, no recurrence was observed during follow-up visits (max. 20 years). Differential diagnostics must include disease entities such as odontogenic cyst, odontogenic myxoma (OM), odontoma complex (OC) (especially in early stage of development), ameloblastic fibroma (AF), odontogenic fibroma (OF), and calcifying odontogenic cyst (COC) [1, 12, 13]. Finding the differences between primordial odontogenic tumor and odontogenic myxoma is important because of the locally malignant nature of the latter, which implies more aggressive methods of treatment. OM is associated with an impacted tooth in only about 5% of cases. Additionally, it does not have a capsule, which allows cell infiltration of the surrounding bone. Since the recurrence rate of odontogenic myxoma is quite high, this creates difficulties during the procedure of simple enucleation of the tumor, therefore requiring segmental bone resection instead [1, 14].

The development of POT includes mesenchyme proliferation similar to that of dental papilla. Immunohistochemical studies indicate the activity of epithelial and mesenchymal components during tumor histogenesis, which justifies its classification among benign mixed epithelial-mesenchymal odontogenic tumors according to the WHO classification [1, 5]. Cytokeratins CK14 and CK19 and glucose transporter (Glut-1), characteristic for human epithelial cells, were present in epithelium in all preparations. The mesenchymal component showed a significant concentration of vimentin and syndecan-1 [5].

Although Fumio *et al*. question the existence of POT and treat it as a histological variant of ameloblastic fibroma (AF) or odontogenic myxoma (OM), it seems that the collected material clearly documents the correctness of isolating the new disease entity [15]. However, due to the small number of described cases, further research is needed in order to articulate the characteristic clinical and histological features of the tumor to allow proper diagnosis and treatment. The authors' own observations and analysis of published clinical cases confirm the benign nature of primordial odontogenic tumor, as well as simple enucleation of the tumor as the main method of surgical treatment of POT.

The authors declare no conflict of interests.

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