

Short Case Report

Odontogenic fibroma-like lesions associated with amelogenesis imperfecta: short case

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Abstract – Observation: We present a case of a 21-year-old Pakistani female with hypoplastic amelogenesis imperfecta (AI), microdontia, and multiple gingival swellings overlying impacted teeth. The gingival swellings were diagnosed as AI-associated odontogenic fibroma (OF)-like lesions on histology. **Commentary:** AI is an inherited enamel defect that may present in association with microdontia, root abnormalities, taurodontism, pulp stones, gingival hyperplasia, and rarely, gingival tumors. These gingival tumors share histologic features with odontogenic fibroma (OF), a benign odontogenic neoplasm, and are referred to as AI-associated OF-like lesions in the literature. **Conclusion:** AI-associated OF-like lesions are rare, this case will inform oral surgeons and other dental practitioners about this process and its management.

Observation

A 21-year-old Pakistani female was seen at the out-patient department at Riphah International University, Islamabad, with a complaint of unerupted teeth and swellings on the mandibular gingiva. There were also esthetic concerns owing to the small size of her teeth. According to the history primary dentition had been similarly affected. There were some people on the girl's father side with similar symptoms, however none the siblings were "obviously" affected. There was no significant medical history and the patient was not using any medications.

Clinical examination revealed microdontia. The teeth were yellow in color and showed signs of chipping (Fig. 1). Physiological pigmentation in the maxillary anterior region was seen. Gingival swellings were noted above impacted mandibular right canine and third molars (Fig. 1). Orthopantomogram (OPG) showed small teeth with well-developed roots. The enamel layer was thin. Follicular enlargement was prominent in mandibular third molars. Pulp spaces were intact with homogenous bone pattern and normal periodontal ligament space (Fig. 2).

Management

Scaling and polishing was performed. The patient underwent excisional biopsy for the gingival swelling above the mandibular canine. The excised tissue was submitted for histopathologic evaluation.

Histopathology

Histopathology showed a proliferation of fibrous connective tissue that supported nests of odontogenic epithelium and numerous psammomatoid calcified bodies. Scattered plasma cells were also seen (Fig. 3). These findings resembled odontogenic fibroma, therefore a conclusion of odontogenic hamartoma was provided.

Diagnosis

The dental condition was diagnosed as Amelogenesis imperfecta (AI). A literature search was carried out to identify AI-associated tumors. A few similar entities were identified. Based on the literature, the case was finally diagnosed as AI associated OF-like lesions.

Treatment

The patient underwent excision of the remaining two tumors at the site of her third molars. The histology was consistent with the lesion removed from the canine region. She was recalled after 2 weeks and provided with composite buildups of maxillary anterior teeth. Two years on we have no report of recurrence in this patient.

Commentary

Amelogenesis imperfecta (AI) is an inherited developmental enamel defect that has a prevalence ranging between 1 in

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Fig. 1. (A) Clinical photograph showing microdontia, yellow color teeth and chipping. (B) Clinical photograph showing gingival swelling above impacted mandibular right canine.



Fig. 2. Orthopantomogram showing thin enamel especially at the cemento-enamel junction and follicular enlargement being prominent in mandibular third molars.

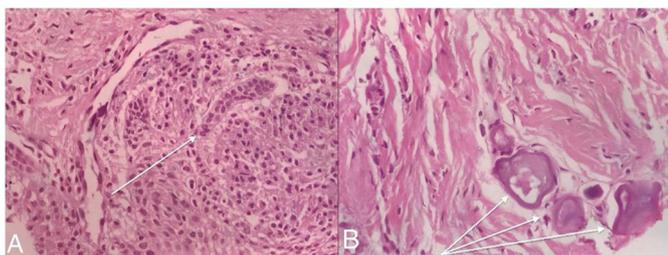


Fig. 3. Histologic sections showing: (A) The odontogenic island, pointed with arrow, (B) Pssammomatoid bodies.

700, to 1 in 1400. It is broadly classified into hypoplastic, hypomaturation, and hypocalcified types. Hypoplastic type presents as thin, soft enamel that may exhibit pitting. Other types can show yellow or brown discoloration, deformation or loss of tooth structure. These features can cause pain, eating difficulties, and esthetic concerns [1].

Radiographically the enamel appears less dense and may show variation in thickness. Other than defective enamel, delayed eruption, microdontia, dilacerated root, short roots, taurodontism, pulp stones, gingival hyperplasia, crowding, gingivitis and periodontitis, and gingival tumors are infrequently seen [1]. The gingival tumors are usually located pericoronally and impede the eruption of underlying teeth. They are either called odontogenic fibroma (OF)-like lesion or odontogenic hamartomas, we will be favoring the OF-like lesion in this text [2].

OF is a benign mesenchymal odontogenic neoplasm that is defined by the World Health Organization (WHO) as “a rare neoplasm of mature fibrous connective tissue, with variable amounts of inactive-looking odontogenic epithelium with or without evidence of calcification” [2]. Central tumors can occur in either jaw and show a female predilection. Peripheral present as variably sized gingival swellings. While there is no established correlation between AI and OF-like lesions, there are several case reports that describe an association. Presence of psammomatoid deposits is the only described feature used to distinguish true OF from OF-like lesions [3].

The first two cases of rough hypoplastic AI and OF-like lesions overlying impacted teeth were described in South Africans by Van Heerden. The patients also had follicular enlargements, root malformations, hypercementosis and missing teeth [3]. The third case was also reported in a South African. This patient’s presentation was similar to the previous cases, however pulpal calcifications were also noted [3]. All three cases showed OF-like histology and presence of psammomatoid deposits. Our case also had rough hypoplastic AI, hypodontia, microdontia, follicular enlargement, nodular fibrotic gingiva. Although our case does not share every clinical feature with these patients, pericoronal gingival growths impeding the eruption of underlying teeth and histology was consistent.

Larger OF-like lesions have also been reported. Raubenheimer documented a case from a 19-year-old with AI who developed COF-like lesion developed in the site of multiple teeth. Calcified psammomatous laminar deposits were seen in histology [3] (Table I).

While most OF-lesion are pericoronal, there are two reports of central occurrences. This includes a case of rough hypoplastic type AI with multiple OF-like lesions causing displacement of impacted teeth, anterior open bite, pulpal calcifications, hypercementosis and generalized gingival overgrowths. While hypodontia and clinical bone expansion or perforation was not evident, the OF-like lesions were located within the jaw rather than the gingiva and were larger than 10 mms in size [3]. The other case was reported in one of the two siblings with AI. The patient was a 17-year-old male with OF-like lesions located centrally around impacted teeth [4].

While OF-like lesion appears to be more popular term, some authors have favored the term dental follicular hamartomas for similar presentations. This includes a case of a 10-year-old black, male patient with enamel dysplasia, delayed eruption, malformed roots, pulpal calcifications, hypercementosis, generalized gingival hyperplasia with dystrophic calcifications and pericoronal follicular hamartomas. No sign of hypodontia was reported in this case [3]. O’Connell S also used it for his report of a 7-year-old female with AI, dental follicular hamartomas, generalized gingival overgrowth, anterior open bite, multiple impactions and dilacerated roots [5].

Table I. Literature review.

Authors	Patients	Dental findings	Odontogenic fibroma
Van Heerden WF <i>et al.</i>	14 year old female	Rough hypoplastic Amelogenesis imperfecta, delayed eruptions, malformed molar roots, pulpal calcifications, hypercementosis,	Pericoronal WHO type COF like lesion
	26 year old female	Rough hypoplastic amelogenesis imperfecta, impacted molars, malformed roots, hypercementosis	Pericoronal WHO type COF like lesion
Peters E <i>et al.</i>	26 year old male	Rough hypoplastic amelogenesis imperfecta, malformed moalr roots, pulpal calcifications, nodular fibrotic gingival hyperplasia, hypodontia	Pericoronal WHO type COF like lesion at maxillary and mandibular molar region
Raubenheimer EJ <i>et al.</i>	19 year old male	Enamel dysplasia, anterior open bite, hypodontia,	COF like lesions at multiple sites in mandible
Feller L <i>et al.</i>	12 year old male	Enamel dysplasia, anterior open bite, delayed eruptions, pulpal calcifications, hypercementosis, generalized gingival hyperplasia	Pericoronal WHO type COF like lesion throughout the dentition
Roquebert D <i>et al.</i>	10 year old male	Hypoplastic amelogenesis imperfecta, delayed eruption, malformed molar roots, pulpal calcifications, hypercementosis, generalized gingival hyperplasia	Pericoronal dental follicular hamartomas at maxillary and mandibular posterior teeth
O'Connell S <i>et al.</i>	7 year old female	Hypoplastic amelogenesis imperfecta, generalized gingival overgrowth, anterior open bite, multiple impactions, dilacerated roots, pulpal calcifications	Pericoronal Dental follicular hamartomas in maxillary first and second molars
Sai Sankar AJ <i>et al.</i>	17 year old male	Hypoplastic amelogenesis imperfecta, retained deciduous canines and molars, missing permanent teeth, generalized gingival hyperplasia.	WHO type COF like lesions in multiple sites in maxilla and mandible

As in our case, majority of these cases were treated with excisional biopsies and no recurrences were reported. However, the impacted teeth that these lesions were overlying, failed to erupt [5].

Conclusion

Here we present a case of AI in association with OF-like lesions in a 21-year-old Pakistani female. While rare, these tumors have been associated with the condition and create complications by impacting the underlying teeth. The aim of this report was to describe this rare presentation so oral surgeons and other dental health practitioners are made aware of its presence and association with AI.

Conflicts of interest: The authors declare that they have no conflicts of interest in relation to this article.

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