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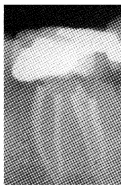

CASE REPORT

Generalized pulp stones: report of a case with 6-year follow-up

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Abstract

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Aim: To present a mildly mentally retarded patient with generalized pulp stone formation and the six-year follow-up and to discuss the differential diagnosis of the case.

Summary: Pulp stones were radiographically detected in the pulp chamber of all permanent teeth in a 25-year-old woman with mild mental retardation who presented for endodontic treatment on tooth no 11 (FDI). The patient's medical, dental and family history was noncontributory. The pulp stone in the pulp chamber of tooth no 11 was removed during canal filing, and root canal treatment completed uneventfully. Six years later, the patient was re-evaluated and the pulp stones were unchanged radiographically. The patient's family history, facial phenotype and karyotype as well as the radiographic, laboratory and physical examination were not consistent with any of the known genetic syndromes associated with generalized pulp stones. Molecular analysis for the DSPP gene proved negative. The aetiology of this case remains unknown.

Key points:

- Generalized pulp stones occur rarely;
- Such patients should be referred for genetic evaluation because pulp stones are mostly associated with genetic dentine defects;
- Pulp stones may hinder root canal treatment;
- Pulp stones may remain unchanged overtime.

Keywords: generalized pulp stones, mental retardation, short roots.

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Introduction

Pulp stones are calcified structures within the pulp of primary or permanent teeth and are even found in unerupted or impacted teeth (Moss-Salentijn & Klyvert 1988, Ranjitkar *et al.* 2002). They are detected radiographically, and their prevalence ranges from 10% to 46%

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(Tamse *et al.* 1982). Pulp stones occur in sizes from microscopic to large particles capable of obliterating the entire pulp chamber (Moss-Salentijn & Klyvert 1988, Ranjitkar *et al.* 2002). They are located more often in the coronal than in the radicular portion of the pulpal cavity and can be seen as free, attached or embedded (Goga *et al.* 2008).

Rarely pulp stones are generalized, appearing in all of an individual's teeth. Calcification in generalized pulp stone formation may be either diffuse, leading to complete pulp obliteration as found in tumoral calcinosis (Lyles *et al.* 1985) and dentine dysplasia type I (Ozer *et al.* 2004) or distinct as found in the present case. Distinct generalized pulp stone formation has been associated with multisystemic genetic syndromes (Goga *et al.* 2008) or with dentine genetic defects, such as dentine dysplasia (O'Carroll *et al.* 1991). In dentine dysplasia type II, the pulp stones are typically located within the pulp chamber, while in dentine dysplasia I within the root which is bulging (O'Carroll *et al.* 1991). In addition, there are reports of distinct generalized pulp stone formation of unknown aetiology (Tsatsas 1971, Siskos & Georgopoulou 1990, VanDenBerghe *et al.* 1999, Parekh *et al.* 2006).

Pulp stones have been rarely associated with pain (Moody *et al.* 1989). They may, however, complicate root canal therapy by blocking access to canal orifices and/or by altering the internal anatomy (Moss-Salentijn & Klyvert 1988, Lovdahl & Gutmann 1997, Ranjitkar *et al.* 2002). Attached stones may deflect the tip of exploring instruments preventing their passage through the canal. Thus, instrumentation is hindered, especially if the pulp stones are located at a curve (Pitt Ford *et al.* 2002). Ultrasonic instrumentation with the use of special tips makes their removal easier (Stamos *et al.* 1985).

Aim

The aim of this case report is: (i) to present a mildly mentally retarded patient with generalized pulp stone formation and the 6-year follow-up and (ii) to discuss the differential diagnosis of the case.

Report

A 25-year-old woman was referred by a general dentist to the Dental School, University of Athens, Greece, for the evaluation of multiple pulp stones and for root canal treatment of tooth no 11. She was the third child of healthy, nonconsanguineous, Albanian parents. Both her older siblings were clinically normal. Family history was unremarkable. She was born with a normal delivery after an uneventful, full-term pregnancy. Her mother reported that the patient had significant delay in speech acquisition.

Regarding her dental history, the mother reported a primary dentition of normal appearance, facial and dental trauma 4 years previously in a motor vehicle accident with avulsion of tooth no 21 and recent sharp persistent pain in tooth no 11. The patient had visited a dentist who alleviated the symptoms with pulp extirpation of tooth no 11 and then referred her to the university.

Facially, she had a slight right deviation of the nasal septum and eyebrows that were heavy medially and sparse laterally (Fig. 1a,b). This phenotype was highly reminiscent of her father.

Intraoral examination revealed bleeding gingival tissues and several periodontal pockets, up to 5 mm. Caries and multiple fillings were found, especially in the posterior teeth. She also had a high-arched palate and bilateral posterior cross-bite, 6 mm of overjet, and tooth no 21 was missing (Fig. 2a,b). Cranium, spine and hand radiographs as well as heart and abdominal ultrasound were unremarkable. Auditory brainstem response test and ophthalmologic evaluation were normal. Radiographic examination with a panoramic

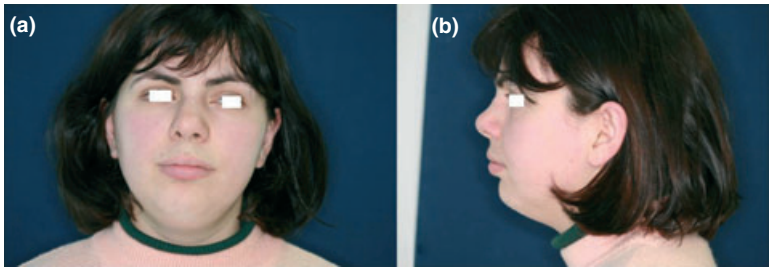

Figure 1 Patient's phenotype. (a) Frontal view. Medially heavy and laterally sparse eyebrows; right deviation of the nasal septum; obesity (b) Lateral view. Normal profile.

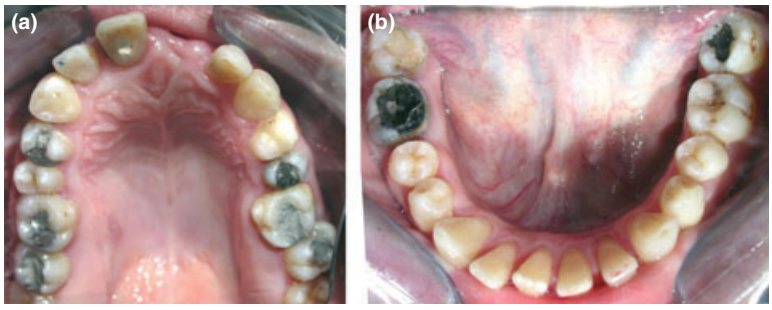

Figure 2 Patient's intraoral photographs. (a) Maxillary arch (b) mandibular arch.

radiograph revealed generalized multiple pulp stones, within the pulp chamber, in most of the teeth and short roots in the first premolars. Periapical radiographs, taken using receptor holding instruments XCP (Dentsply Rinn Co., Elgin, IL, USA), verified the presence of large pulp stones in all permanent teeth, located coronally within the pulp chamber. Representative radiographs are shown in Fig. 3a,b,c. More than one pulp stone was present in the canines and second molars. When the teeth were examined for pulpal status, all had a normal response to thermal and electrical stimuli except for the right maxillary central incisor, in which pulp extirpation had been performed. Both the family history and the panoramic radiographs of both parents were negative for pulp stones.

The patient was referred for genetic evaluation because it is known that generalized pulp stone formation is often being associated with multisystemic syndromes. Consultation with one of the authors (DS), a geneticist, revealed mild mental retardation. On

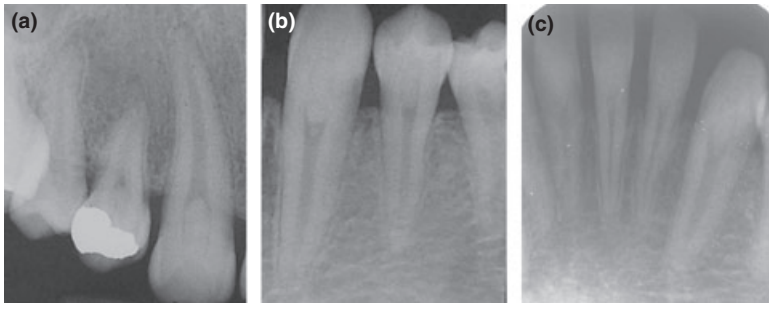

Figure 3 Periapical radiographs taken at the initial visit with apparent pulp stones located coronally in all of the teeth. (a) teeth 13, 14, 15 (b) teeth 33, 34, 35 (c) teeth 41-33.

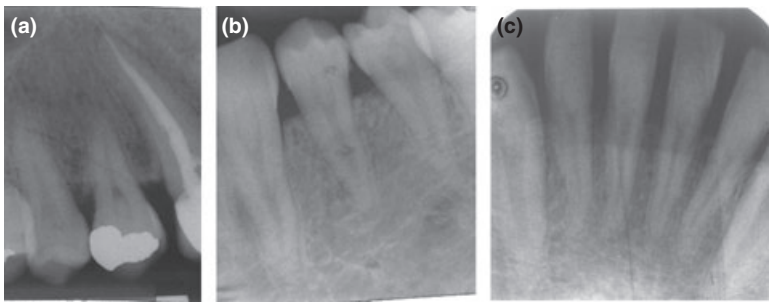

Figure 4 Periapical radiographs taken at the 6 years follow up. Pulp stones still present, seeming unchanged in all of the teeth. (a) teeth 13, 14, 15 (b) teeth 33, 34, 35 (c) teeth 43-33.

physical examination, the patient was obese and presented extremely dry skin that was exfoliating despite regular emollient application. She had mild bilateral hallux valgus. She had a normal female karyotype as revealed by the fibroblast chromosome analysis at 550 bands level. Molecular analysis of the DSPP gene proved to be negative.

The dental treatment plan included the following: completion of the root canal treatment of the right maxillary central incisor, periodontal therapy, restoration of carious teeth, orthodontic treatment and prosthodontic rehabilitation for the missing left maxillary central incisor. During the endodontic therapy of the right maxillary central incisor, removal of the pulp stone was attempted using a sharp endodontic excavator, to analyse its chemical composition. The pulp stone, however, was firmly attached to the dentinal walls and was gradually removed during the canal preparation. The endodontic treatment was completed uneventfully.

The patient did not keep her appointments but moved to her home country, where she sought dental treatment. However, she returned for re-evaluation 6 years later. New periapical radiographs were taken with the parallel technique, using receptor holding instruments to allow comparison with the initial radiographs. The radiographs revealed generalized pulp stones in all teeth. Representative radiographs are shown in Fig. 4a,b,c. These intraoral radiographs were compared with the initial ones by three experienced oral radiologists, according to a standardized protocol (Landin & Koch 1977). The consensus was that the pulp stones were unchanged in size and number in the six-year follow-up.

Discussion

This is a rare case report of generalized pulp stones within the pulp chamber, of unknown aetiology. There was no pain or other symptoms involved. Pulp stones remained unchanged during the 6-year follow-up. No previous reports exist on the long-term radiographic follow-up of pulp stones.

Generalized distinct pulp stones within the pulp chamber occur rarely, and the differential diagnosis includes the following genetic disorders: dentine dysplasia type II (O'Carroll *et al.* 1991), Saethre-Chotzen (Goho 1998), Elhers-Danlos (De Coster *et al.* 2005) and Otodental syndromes (Witkop *et al.* 1976).

The most probable diagnosis according to the pulp stones radiographic appearance, location and number is dentine dysplasia type II (MIM #125420), caused by heterozygote mutations of the dentine sialophosphoprotein DSPP gene. This was the reason for performing molecular analysis for this gene that proved, however, negative. In addition, according to her mother, the patient did not have primary teeth that were opalescent in colour, a characteristic of this condition. Second, in the differential diagnosis list, but not so likely, is Saethre-Chotzen syndrome (MIM #101400) caused by dominant mutations of

the TWIST1 gene. This condition is one of the most common craniosynostosis syndromes, characterized primarily by facial asymmetry, syndactyly and mild mental retardation, as presently found. Oral anomalies include the following: narrow or highly arched palate as presently found but also supernumerary teeth and enamel hypoplasia (Goho 1998), which were not present. This syndrome has only been once associated with pulp stones within the pulp chamber (Goho 1998). Although the mild mental retardation, nasal septum deviation, eyebrow pattern and bilateral hallux valgus have been reported in the Saethre-Chotzen syndrome, it is likely that they represent nonspecific, familial or casual features of the patient reported here. However, only the molecular analysis of *TWIST1* gene would exclude the hypothesis of this variably expressive syndrome.

Pulp stones may be also present in patients with Ehlers-Danlos syndrome (MIM #130000) (De Coster *et al.* 2005). However, in this syndrome, the pulp stones gradually increase in size, ultimately obliterating the entire pulp space, a feature not observed in the 6-year follow-up radiographs of the present case. Furthermore, the patient had none of the characteristics of this syndrome, i.e. hypermobility, skin hyperextensibility and/or increased tissue fragility with extensive scarring. Generalized pulp stones have been described in the rare Otodental dysplasia (MIM #166750). This syndrome is characterized by sensorineural hearing loss, globodontia or taurodontia in several teeth and bifurcated pulp chambers (Witkop *et al.* 1976). None of these features were present in this patient. Recently, a mentally retarded kindred with intracoronal and radicular pulp stones in most of the teeth was reported in a consanguineous family, suggested as a new syndrome (Martelli-Júnior *et al.* 2008). These patients, however, had also gingival fibromatosis and amelogenesis imperfecta, features not present in the oral phenotype of the present patient.

Similar cases of generalized pulp stone formation of unknown aetiology have been reported previously, in Greek teenagers, one in a 15 years old (Tsatsas 1971) and another in a 14 years old (Siskos & Georgopoulou 1990). Pulp stones were mostly coronally located within the pulp of all teeth, even in unerupted ones. No aetiology had been established in either of these cases but no molecular analysis had been performed. There has also been a report of adult twin sisters having generalized pulp stone formation of unknown aetiology in almost all of the teeth. In some of the teeth, the pulp stones obliterated the pulp chamber (VanDenBerghe *et al.* 1999). Another case of a 13-year-old boy has been reported in England. The location of pulp stones, however, differed from the present case because they were mostly located in single-rooted teeth and premolars and in the middle third of bulging roots (Parekh *et al.* 2006).

It has been suggested that chronic inflammation of the pulp caused by local factors such as periodontal disease, tooth abrasion, dental caries and abnormal occlusion may lead to pulp stone formation as an attempt by the pulp to repair (Sundell *et al.* 1968, Ranjitkar *et al.* 2002, Goga *et al.* 2008). However, if that was the aetiology for the present case, pulp stones would not be present in all of the teeth and those existing would have progressed in the 6-year follow-up period.

No definitive diagnosis could be made for the patient reported here. The phenotype of the patient could be a random association of mental retardation and generalized pulp stones, possibly a new phenotype.

Conclusion

Pulp stones appearing in almost all teeth of an individual are rarely found. Such patients should be referred for genetic evaluation because this condition is mostly associated with multisystemic syndromes or dentine genetic defects. Pulp stones rarely cause pain, but they may hinder canal instrumentation. The pulp stones observed in the present case remained unchanged over a 6-year observation period.

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