Library Progress International
Vol.44 No. 3, Jul-Dec 2024: P.11136-11142
Print version ISSN 0970 1052
Online version ISSN 2320 317X
Case Report
Available online at www.bpasjournals.com

Familial Presentation of Dentin Dysplasia Type-I in siblings: A Case report

Pranali B. Bahadure[1]*, Bhakti Puranik[2], Mahesh Chavan[3] , Saurabh Chandra Pawar[4] Dhanashree Kamble[5] , Priya Shinde[6]

1 Lecturer, Department of Oral Medicine and Radiology, D. Y. Patil Dental School, Lohegaon, Pune, Maharashtra, India. bahapranali25406@gmail.com Orcid ID:0000-0002-1518-299X.

2 Post Graduate student, Department of Oral Medicine and Radiology, Sinhgad Dental College and Hospital, Pune, Maharashtra, India. bhaktigp300@gmail.com

3 Professor and Head of Department, Department of Oral Medicine and Radiology, Sinhgad Dental College and Hospital, Pune, Maharashtra, India. drmaheshschavan@gmail.com

4 Private Practitioner, Nashik, Maharashtra, India. pawarsaurabhachandra@gmail.com

5 Post Graduate student, Department of Oral Medicine and Radiology, Sinhgad Dental College and Hospital, Pune, Maharashtra, India. dhanashreekamble.dk@gmail.com

6 Post Graduate student, Department of Oral Medicine and Radiology, Sinhgad Dental College and Hospital, Pune, Maharashtra, India. piyaadangel@gmail.com

How to cite this article: Pranali B. Bahadure, Bhakti Puranik , Mahesh Chavan , Saurabh Chandra Pawar, Dhanashree Kamble , Priya Shinde (2024) Familial Presentation of Dentin Dysplasia Type-I in siblings: A Case report.Library Progress International, 44(3), 11136-11142

Abstract

Dentin dysplasia is a rare genetic condition with autosomal inheritance. In this case study, we report a male and female patient who are siblings, diagnosed with dentin dysplasia type I. They were the first affected generations in their family. The diagnosis of dentin dysplasia type I was confirmed based on the clinical and radiographic features. Dentin dysplasia I has distinctive radiographic features that differentiate it from other dentin developmental disorders. Since periodontitis and root morphology can cause early exfoliation of teeth, the goal is to provide preventive treatment for optimal oral function

Keywords: Dentin dysplasia, dentin, rootless teeth, pulpal obliteration.

INTRODUCTION

Dentin dysplasia (DD) is the rare dental anomaly of dentin formation characterized by normal enamel but atypical dentin formation with abnormal pupal morphology. It impacts either the primary dentition or both the primary and secondary dentitions, occurring in about one individual per 100,000 [1]. Based on clinical and radiographic presentation, Shields et al. [2] have proposed two primary classes of DD: type I, and type II. Later, Witkop [3] used the terms "radicular dentin dysplasia" for type I and "coronal dentin dysplasia" for type II. The clinical characteristics of DD have been extensively documented since the condition was first identified in 1920 as "rootless teeth" and later as "dentin dysplasia" by Rushton et al [4] in 1933. However, the pathophysiology of dentin abnormalities and their clinical and pathological ramifications remain largely unknown to present.

This case report aims to elucidate the intricate nature of dentin dysplasia, emphasizing its clinical and radiographic implications and therapeutic considerations. By delving into this intriguing case, we can glean valuable insights

into the complexities of this condition and foster a deeper understanding of its impact on oral health and patient management.

CASE PRESENTATION:

An 18-year-old female patient and a 19-year-old male patient reported to the Department of Oral Medicine and Radiology with the chief complaint of missing teeth in the upper and lower front region of the jaw since 6 years, discomfort during mastication, and aesthetic problems. Family history reveals they both were siblings and their parents had a consanguineous marriage. There was no contributory medical history. Premature loss of primary teeth was reported in both siblings. On clinical examination, both siblings had multiple missing permanent teeth due to early exfoliation of teeth due to loosening, mobile teeth, dental caries, and the generalized bulbous shape of the crown of upper and lower molars with an extra cusp tip (Figure 1,2).

Patient A

The dental findings of the female patient revealed missing teeth with 11 12 13 21 22 31 32 36 41 42, Advanced occlusal caries with grossly destructed crown with 38 47 48, Moderate occlusal caries with 18 27 37 Early Occlusal caries with 17 35 45 46. Grade I Mobility 23 33 34 Grade III Mobility 35 43 (Figure 1)

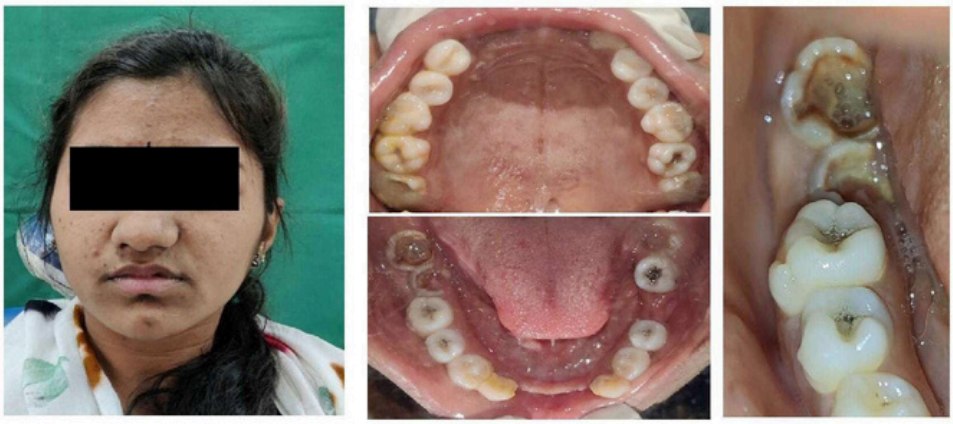


Figure 1: Patient A Extra oral and Intra Oral Photographs

Patient B

The dental findings of the male patient revealed missing teeth with 11 12 21 22 31 32 41 42, Early Occlusal caries with 45 47 48 38 Grade I Mobility 23 33 43. The color was the same as a normal tooth. (Figure 2) Based on family history and clinical findings, a provisional diagnosis of dentin dysplasia was made. However, considering the detailed history and findings, a differential diagnosis of dentinogenesis imperfecta was considered. Further investigations, including complete blood tests (within normal limits) and radiographic examination (RVG and OPG), were conducted to confirm the diagnosis.

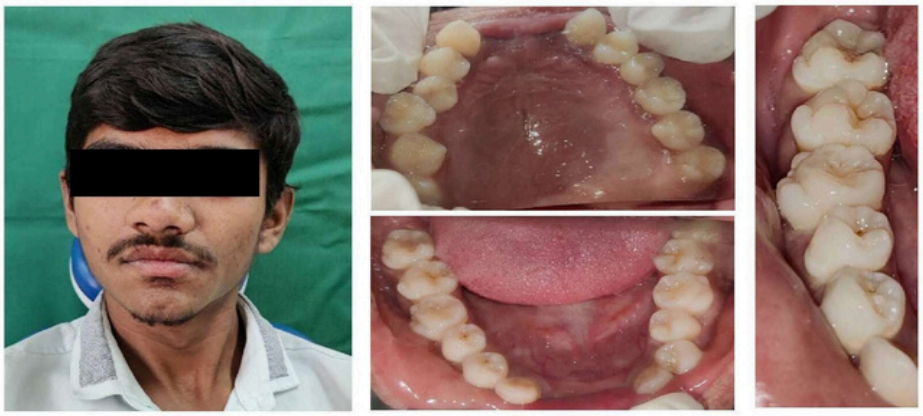
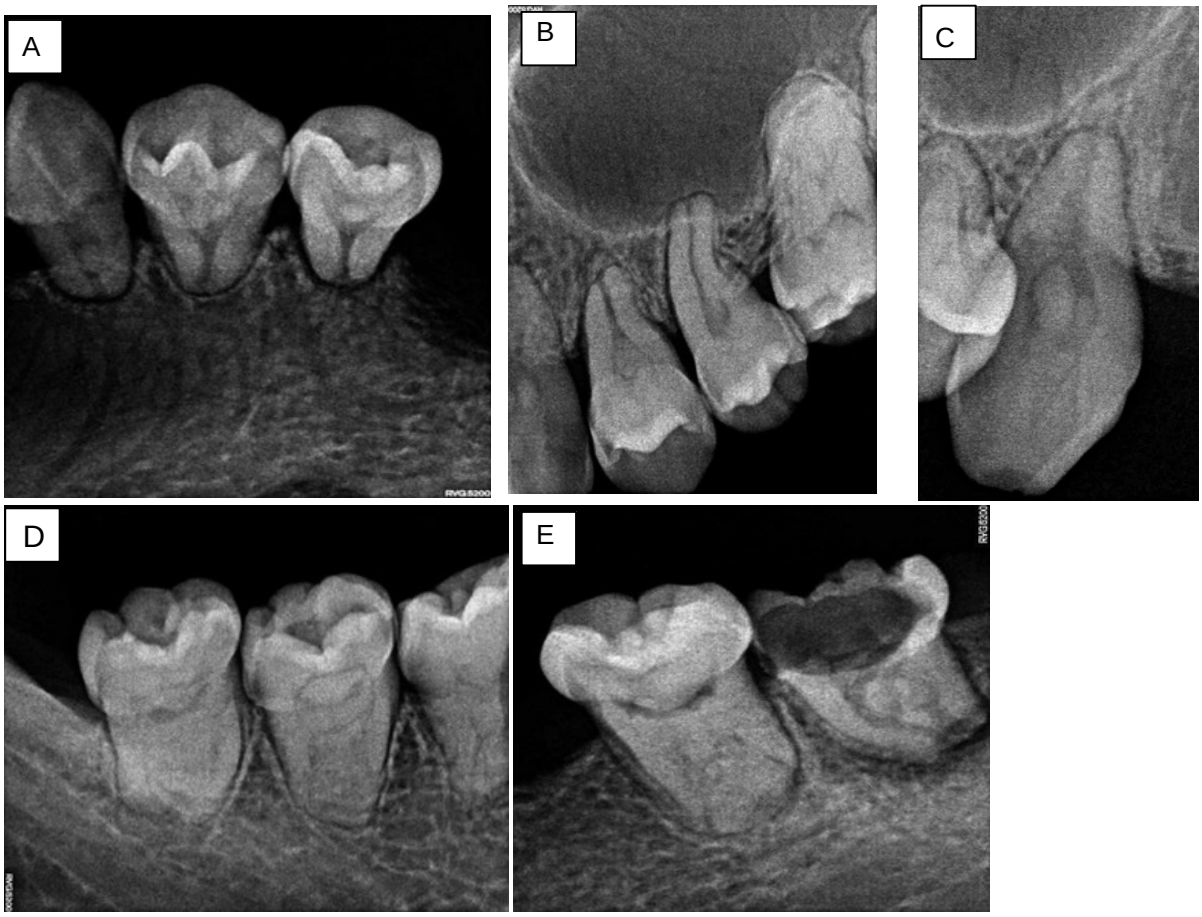
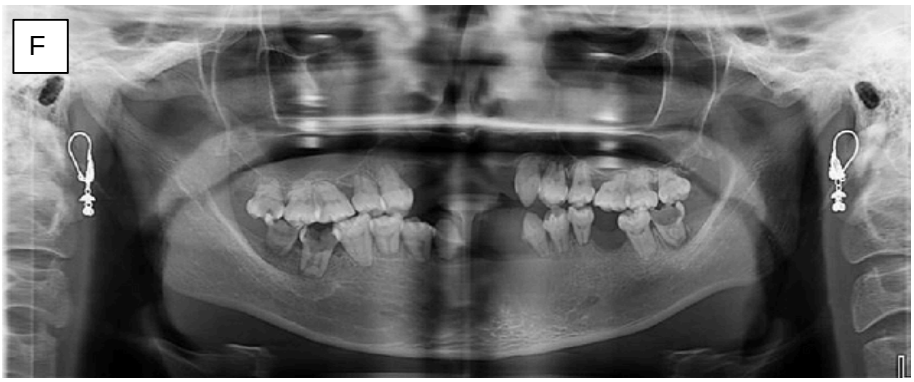


Figure 2: Patient B Extra oral and Intra Oral Photographs

RADIOGRAPHIC EXAMINATION

On radiographs, bulbous crown structure was seen. Roots were short, blunt, conical, and malformed. Pulp chambers and root canals were partially obliterated in the female patient (Patient A) and completely obliterated in the male patient (Patient B). Multiple pulp stones were seen in the pulp chamber. Crescent-shaped pulpal remnants were seen in the pulp chamber. Generalized radiolucency was evident in the periapical area. Generalized horizontal alveolar bone loss was seen. The final diagnosis of Dentin Dysplasia type I was given after considering all the investigations. (Figure 3)





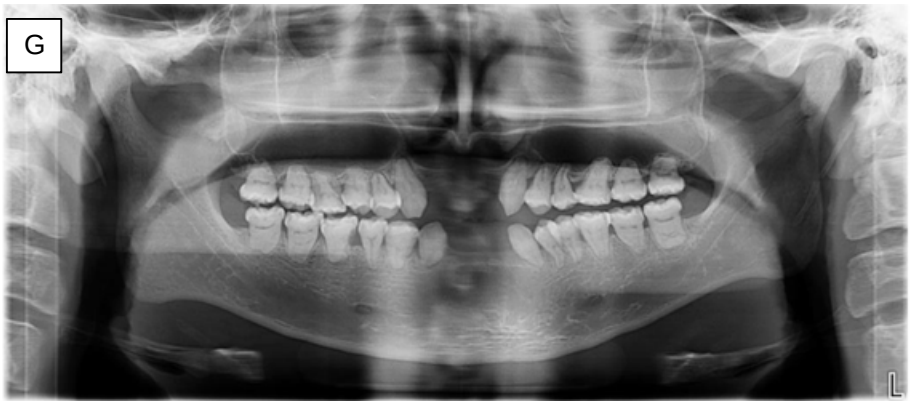


Figure 3: Radiographic examination (A, B, C) Bulbous crown appearance and multiple pulp stones in the pulp chamber, (D) obliterated pulp chamber, (E) crescent-shaped remnants in the pulp chamber (F, G) OPG of Patient A and Patient B.

DIFFERENTIAL DIAGNOSIS

Other dentin developmental disorders such as DD II and Dentinogenesis imperfecta, were ruled out (Table 1). [5] In our patients, the radiographic characteristics like bulbous crown structure, short and blunt roots, obliterated pulp chamber and root canals, crescent-shaped remnants and multiple pulp stones in pulp chamber proved characteristic for the diagnosis of dentin dysplasia type I.

	Dentin Dysplasia type I	Dentin Dysplasia type II	Dentinogenesis Imperfecta
Mode of inheritance	Autosomal dominant	Autosomal dominant	Autosomal dominant
Dentition affected	Primary and permanent teeth-normal	Primary teeth-discolored Permanent teeth-normal	Primary and permanent teeth discolored
Mobility	Present	Absent	Absent
Taurodontism	Present	Absent	Absent
Radiographic appearance	Small or no roots and obliteration of the pulp chambers-rootless teeth	Thistle tube shaped or flame-shaped pulp chambers	Bulbous crowns, cervical constriction, thin roots and early obliteration of pulp chamber
Histopathologic features	Central portion of the root forms whorls of tubular dentin giving the appearance of "stream flowing around boulders."	Coronal dentin- Primary teeth– irregular tubules Permanent teeth-normal tubules True denticles, excessive dentin remodelling	Coronal dentin exhibiting short misshapen tubules within atypical granular dentin matrix

Table 1 : Differential diagnosis for Dentin Dysplasia

Additional diagnosis were dental caries, partial edentulism, and chronic generalized periodontitis. The treatment plan included oral prophylaxis, restoration of carious teeth, and prosthetic rehabilitation for edentulous areas. The female patient also required extraction of severely decayed teeth. Preventive measures, including brushing technique demonstrations and oral hygiene instructions, were provided to both.

DISCUSSION

We report a case of dentin dysplasia type I in siblings born to parents with a consanguineous marriage, increasing the risk of autosomal conditions through the expression of recessive deleterious alleles. Approximately 2% (1 to 50) of couples are at risk of having a child with a serious or lethal medical condition [6] Putrino et al. [7] reported two cases of dentin dysplasia type I, in two siblings and their father. According to the theory put forth by Logan et al. [8], the anomalies in root growth are caused by the dental papilla. They proposed that several degenerative foci inside the papilla calcify, inhibiting development and ultimately obliterating the pulp space. Sauk et al. [9], suggested Dentin dysplasia is a defect where the root sheath invades too early, causing a stunted root with a whorl-like dentin pattern, obliterating the pulp chamber. Melnick et al. [10] suggested that the abnormal root morphology is caused by abnormal differentiation and/or function of the odontoblasts. The exact etiology has yet to be explored. The initial symptom of the disease is tooth mobility, leading to primary and permanent premature tooth loss due to minimal trauma or spontaneous exfoliation. Diagnosis can made using a combination of patient history, distinctive clinical and radiographic features, histopathological examination, and genetic analysis revealing an SMOC2 gene deficiency.[11] Management of patients with Dentin dysplasia type I is a great challenge to dentists because of the changes in tooth morphology. Endodontic treatment will be challenging because of obliteration of root canals and pulp chambers. Extraction remains the only treatment alternative for such patients. Conservative management, periapical surgery, retrograde filling, prosthetic rehabilitation can be done. In cases of resorption, bone grafting, and sinus lift procedures may be necessary for implant placement [12] .

CONCLUSION

This case report highlights the unique radiographic features of dentin dysplasia I, and explores its clinical implications, diagnosis, and treatment options, distinguishing it from other dentin disorders.

ACKNOWLEDGEMENTS

The authors would like to extend their sincere appreciation to the staff and colleagues at the Department of Oral Medicine and Radiology, D.Y. Patil Dental School, and Sinhgad Dental College and Hospital for their support in the diagnosis and management of the patients presented in this case report. Special thanks to the patients and their family for their cooperation throughout the diagnostic and treatment process.

CONFLICT OF INTEREST

The authors declare no conflict of interest related to the publication of this case report.

FUNDING

No external funding was received for the research, authorship, and publication of this case report.

REFERENCES

1. Shankly PE, Mackie IC, Sloan P. Dentinal dysplasia type I: report of a case. Int. J. Paediatr. Dent. 1999; 9:37-42.
2. Shields ED, Bixler D, El-Kafrawy AM. A proposed classification for heritable human dentine defects with a description of a new entity. Arch. Oral Biol. 1973 1; 18:543-IN7.
3. Witkop Jr CJ. Hereditary defects of dentin. Dent. Clin. N. Am. 1975 1; 19:25-45.
4. Rushton MA. A case of dentinal dysplasia. Guys Hosp Rep. 1939; 89:369-73.
5. Neville B, Damm D, Allen C, Bouquot J, Bouquot J. Abnormalities of teeth. 2nd ed. Philadelphia: Saunders Elsevier; 2005. Oral and Maxillofacial Pathol; 94–101.
6. Lakshmayya Naidu D, Srinivasa Raju M, Goel S. Effects of consanguineous marriages on oral and craniofacial structures: a study on dental patients in north India. Ann Essences Dent. 2010 1; 2:199-203.
7. Putrino A, Caputo M, Galeotti A, Marinelli E, Zaami S. Type I Dentin Dysplasia: The Literature Review and Case Report of a Family Affected by Misrecognition and Late Diagnosis. Medicina. 2023 17; 59:1477.
8. Logan J, Becks H, Silverman Jr S, Pindborg JJ. Dentinal dysplasia. Oral Surg Oral Med Oral Pathol. 1962 1; 15:317-33.
9. Sauk JJ, Jr, Lyon HW, Trowbridge HO, Witkop CJ., Jr An electron optic analysis and explanation for the etiology of dentinal dysplasia. Oral Surg Oral Med Oral Pathol. 1972; 33:763–71.
10. Melnick M, Levin LS, Brady J. Dentin dysplasia type I: A scanning electron microscopic analysis of the primary dentition. Oral Surg Oral Med Oral Pathol. 1980; 50:335–40.
11. Chen D, Li X, Lu F, Wang Y, Xiong F, Li Q. Dentin dysplasia type I-A dental disease with genetic heterogeneity. Oral Dis. 2019: 25:439-446.
12. Toomarian L, Mashhadiabbas F, Mirkarimi M, Mehrdad L. Dentin dysplasia type I: a case report and review of the literature. J. Med. Case Rep. 2010 4:1-6.