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Case Report

Acute gingival bleeding in Ehlers–Danlos syndrome

Neha Bansal[a,*], Narinder D. Gupta[a,1], Himanshu Trivedi[a,2], Sandhya Maheshwari[b,3]

[a] Department of Periodontics & Community Dentistry, Dr Ziauddin Ahmad Dental College (AMU), Aligarh 202002, U.P., India
[b] Department of Orthodontics & Dental Anatomy, Dr Ziauddin Ahmad Dental College (AMU), Aligarh 202002, U.P., India

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ABSTRACT

Ehlers–Danlos syndrome (EDS) is an inherited syndrome in which there is a defect in collagen synthesis. Dental component of many such cases frequently goes unnoticed, or presents as secondary features. Dental health professionals can play an important role in diagnosis of such rare genetic syndromes by correlating oral manifestations with systemic signs and symptoms.

1. Introduction

Ehlers–Danlos syndrome (EDS) is a heterogeneous group of inherited connective tissue disorder in which there is a defect in collagen synthesis. Other terms such as “elastic man” or “Indian rubber man” are also being used synonymously for this syndrome.

The occurrence of joint hypermobility with spontaneous hemorrhage is a common feature in Ehlers–Danlos syndrome. But there is no report of this rare entity with the chief complaint of intra-oral bleeding. The reasons of acute gingival bleeding may be numerous but when it occurs in conjunction with other clinical features as in this particular case, the need of a thorough examination and investigations cannot be disregarded. The cases of EDS with chief complaint of intra-oral bleeding are not reported yet and this makes this case worth reporting.

2. Case report

An 8-year-old girl referred to the Department of Periodontics of Dr Ziauddin Dental College with chief complaint of gingival bleeding since last 2 days. Initially, bleeding was intermittent in nature which was aggravated by oral prophylaxis by a local dentist. She gave a history of recurrent fall while walking and easy bruising all over the body since as long as she remembered. There was no history of any other previous dental treatment or bleeding from any other site of body. There was no significant medical and drug history. Occurrence of similar manifestations was found in two out of three sisters of the patient.

On extra-oral examination of the patient, typical long face with prominent eyes, staring look due to loss of infraorbital adipose tissue and pinched nose was noted (Fig. 1a). On intra-oral examination a hematoma (8 by 20 mm) was observed in relation to buccal gingiva of upper right primary second molar and permanent 1st molar (Fig. 1b). Periodontal status appeared to be normal. Patient’s palate was deep and dome-shaped (Fig. 1c). IOPA X-rays and panoramic radiograph revealed no significant abnormal finding.

On general examination, articular hypermobility was observed (Fig. 1c). However, no episodes of joint dislocation have occurred in the past. Muscle bulk, tone and power were noticeably reduced. On Beighton scale, a Score of 7/9 was given to patient for joint hypermobility (Fig. 2). Trendelenburg sign and mild hyperelasticity of the skin could be elicited (Fig. 3a and b). Subdermal ecchymosis on shin of her right leg and bruises were observed on forearms (Fig. 3b and c).

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

* Corresponding author. Tel.: +91 98 975 17350.
E-mail addresses: nehabansal2k5@gmail.com (N. Bansal), ndguptaligarh@gmail.com (N.D. Gupta), trivedi.heman@gmail.com (H. Trivedi), sandhyaligarh21@yahoo.com (S. Maheshwari).
[1] Tel.: +91 0571 2721184; mobile: +91 94 128 17009.
[2] Tel.: +91 99 972 37723.
[3] Tel.: +91 0571 2721184; mobile: +91 94 122 73293.

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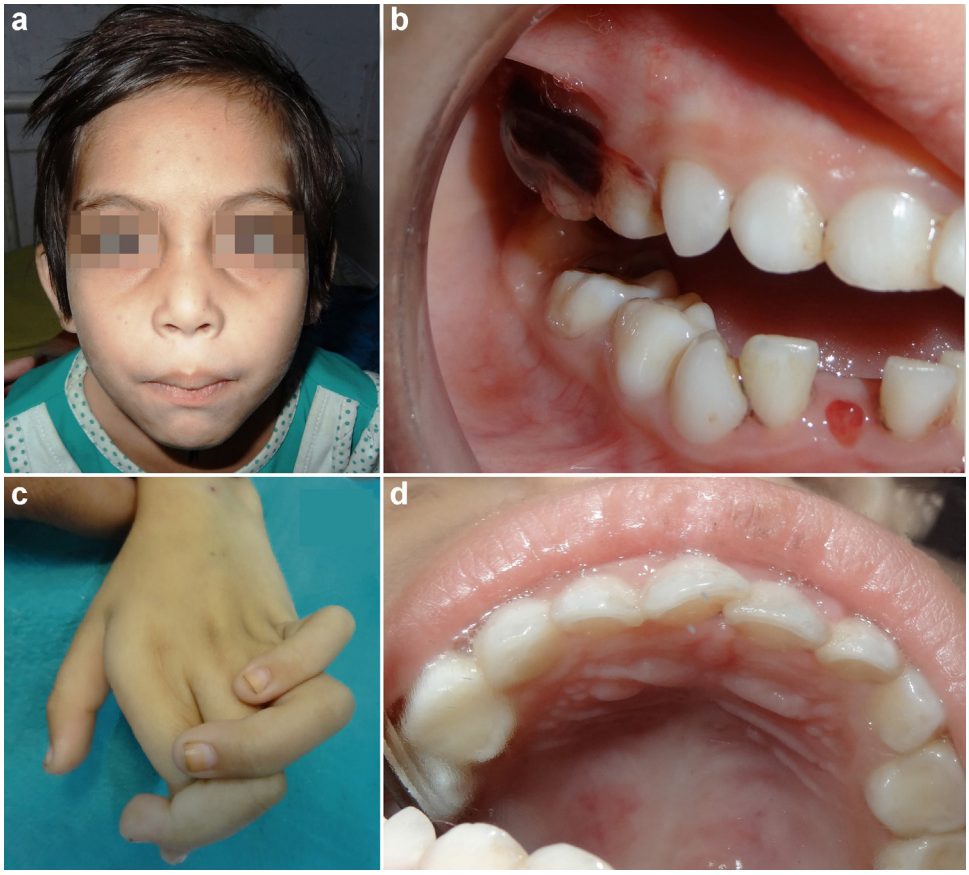

Fig. 1. (a) Typical long face with prominent eyes, staring look due to loss of infraorbital adipose tissue and pinched nose. (b) Intra-oral hematoma in relation to buccal gingiva of maxillary primary second molar and permanent 1st molar. (c) Showing articular hypermobility. (d) Deep and dome-shaped palate.

The patient was referred to the Department of Pediatrics for evaluation of cardiovascular, respiratory and gastrointestinal and renal system. Other routine investigations such as complete blood picture, serum electrolyte analysis, BT, CT, INR ratio, RFT and LFT were advised. Blood biochemical revealed increased INR (1.75), microcytic normochromic anemia with low hemoglobin (6.5).

Treatment provided was palliative only. The patient and her parents were educated and motivated about syndrome. The patient was advised ice pack application at the site of hematoma. A gum paint containing 1 percent iodine and 0.5 percent tannic acid (w/v) was advised to the patient for local application on the site of hematoma thrice daily. 1.5 g/day of vitamin C in chewable tablet form was prescribed to the patient for 3 months. 5 mg vitamin K was administered to the patient by intravenous route for 3 days. Significant improvement was noted at the site in the patient with complete resolution of hematoma.

3. Discussion

Ehlers–Danlos syndrome is a rare genetic syndrome with 1:5000 prevalence rates for all types of EDS, with EDS-hypermobility type accounting for about half of all registered cases [1]. Job Van Meerkeran was the one who reported index case in 1882 which was later elaborated by Ehlers in 1901 and in 1908 by Danlos.

EDS comprised of characteristic but variable features and is primarily diagnosed by clinical findings and family history. The syndrome has both oral and extra-oral manifestations but degree of involvement varies depending on the type of EDS.

Characteristic principle clinical features of EDS are joint hypermobility, poor muscle tone, skin hyperextensibility, bone pain and fractures, fragile and hyperelastic skin, easy bruising, ecchymosis and hematomas, delayed wound healing and atrophic scarring [2]. Bleeding time may or may not be prolonged, but no consistent coagulation factor abnormalities have been reported [3].

Intra-oral manifestations are included in minor diagnostic criteria and include fragile mucosa and gingival bleeding leading to acute hemorrhage, early-onset generalized periodontitis, hypoplasia, microdontia, pulp stones, deformed roots, vault shaped palate, and very supple tongue (Gorlin's sign) [4]. Bleeding from the gums usually follows brushing of the teeth, or excessive bleeding after minor trauma following treatments such as prophylaxis, periodontal surgery or extraction.

Recently Ferré et al. reported some new specific oral signs of disease whose combination may be of greater value in diagnosing the condition. These include temporomandibular disorders including joint pain, excessive dentin formation leading to reduction in pulp volume, molar root fusion and increased root length and marked gingival fragility [5].

Usually, the complications of the syndrome develop early in life mostly in 1st and 2nd decade of life and may predispose patient to severe physical impairment. Therefore, prompt early diagnosis is very necessary for improving the patient's prognosis and quality of life. Many complications of syndrome such as joint luxations and life threatening bleeding episodes can be prevented by early diagnosis. Also being an inherited disorder, early diagnosis helps in educating parents for prenatal and postnatal diagnosis of syndrome in other siblings.

EDS is considered an invisible disability. There is generally poor knowledge of the syndrome among practitioners; hence, many patients remain undiagnosed until severe physical impairment