Ehlers-Danlos Syndrome (Type VIII)

Aysegul Apaydin

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Abstract

Ehlers-Danlos syndrome (EDS), a group of rare, autosomal dominantly inherited connective tissue dysplasias, characterized mainly by abnormal collagen synthesis, has been shown to exhibit extensive heterogeneity with at least 11 clinical entities differentiated by their clinical, biochemical, and genetic features. Of these, Type VIII EDS is of special interest from a dental viewpoint, due mainly to the resulting fragility of the oral mucosa and blood vessels, and an aggressive type of periodontitis causing severe loss of alveolar bone; all of these clinical manifestations of the entity make it necessary to exercise care during the dental treatment procedures. Here we present a case of EDS Type VIII with typical clinical findings, together with details of the dental treatment modalities, placing emphasis on surgical methods.

Introduction

Ehlers-Danlos syndrome (EDS) is a collective group of rare, autosomal dominantly inherited connective tissue dysplasias, characterized by abnormal collagen synthesis and associated with clinical manifestations such as tendency for excessive bruising, hyperextensibility and abnormal fragility of the skin, defective and delayed healing of skin wounds (instead of contraction, scars show a spreading tendency), and hypermobility of the joints[1-14]. Ocular, cardiovascular, and gastrointestinal abnormalities are also often associated with the syndrome[3]. Dermatofragility, dermatorrhexis, cutis hyperelastica, and Meekein-Ehlers-Danlos are some of the synonyms cited in the literature[15,16].

The condition known today as Ehlers-Danlos Syndrome was first described by Job a Meek’ren in 1682 as an extraordinary elasticity of the skin, and later named after a Danish dermatologist, Ehlers, and a French physician, Danlos, who first pointed out the syndrome as a clinical entity[15].

The severity of the disorder varies from relatively mild to life-threatening, the latter being characterized by spontaneous rupture or aneurysm formation in major vessels and by spontaneous visceral rupture[17-19].

Joint hypermobility, usually a primary feature of the syndrome depending on the type of EDS, affects both large and small joints and may be severe enough to cause recurrent dislocations, including subluxation of the temporomandibular joints[12,17].

On the basis of clinical, biochemical, and genetic features, 11 distinct types of EDS have been described[20]. Because of the ultrastructural inconsistency of the connective tissue abnormalities and the lack of pathognomic laboratory tests, EDS is diagnosed using clinical features[6,21].

From a dental viewpoint, Type VIII EDS is of special interest because of various types of oral involvement[11,12,15,17,22] such as aggressive-type periodontitis with gingival recession and early tooth loss. The present paper describes a case of EDS Type VIII with dental findings.

Case Report

A 26-year-old male patient was referred to the Department of Oral and Maxillofacial Diseases and Surgery because of pain in the right ear and swelling in the posterior part of the right mandible of 10 days duration.
The patient’s medical history revealed a previous referral to the Department of Dermatology with complaints of chronic Al skin lesions on the nose and forehead, while a pathological report on a dermatological biopsy suggested Ehlers-Danlos syndrome. The patient’s dental history included loss of the majority of the mandibular teeth at the age of 3-4 y and use of a partial denture since the age of 7 y.

Hypermobility of the joints at the shoulders and the elbows (Fig. 1), and easy accessibility of the tongue to the tip of the nose were some of the interesting clinical features observed. No unusual pathological findings were evident upon pulmonary and ophthalmological examination. Similar clinical findings have not been found in any other member of the patient’s family.

Extra oral examination revealed characteristic scar formations on the nose and forehead skin (Fig. 2) and herpetical lesions at the angles of the mouth. Widely set eyes and a broad bridge to the nose gave a significant appearance to the face.

Examination of the dentition revealed 7 6 5 4 3 2 1−b1 3 4 5 6 7 8, 7 5−b1 2 3 7.

Radiographical analysis revealed that tooth 20 was in a horizontally impacted position, with extensive resorption of the mandibular bone around the sites where teeth were absent, suggesting severe periodontal involvement of the mandible (Fig. 3). In the light of the clinical and radiographical findings, an acute apical abscess was considered to be present around tooth 31.

After administration of a broad-spectrum antibiotic, tooth 31 were extracted. Although great care was taken to perform atraumatic extraction, a delay in wound healing and some cracks in the underlying alveolar bone inevitably occurred.

Since then, the patient has appeared for recall regularly at monthly intervals for 20 months.

Discussion

The diagnosis of EDS, in the general sense, requires clinical, biochemical, and genetic data. On the other hand, Type VIII, which is considered to be more important from a dental viewpoint, is diagnosed mainly on clinical grounds based on the characteristic clinical findings. This is due to inconsistency of the connective tissue abnormalities and the lack of specific pathognomic laboratory tests[6,20,21].

Our patient was considered to have EDS on the basis of pathological examination of dermatological biopsy material. This preliminary assumption was confirmed by the clinical features, and the EDS was diagnosed as Type VIII mainly on clinical grounds. The hypermobility of the joints in the upper extremities, characteristic scarring on the nose and forehead skin, a broad bridge to the nose, and widely set eyes provided extraoral evidence supporting the clinical diagnosis. However, hypermobility and subluxation of the temporomandibular joint, which is a frequently reported feature of the syndrome, was not evident.

The diagnosis of EDS is of importance for subsequent dental treatment[12]. Fragility of the oral mucosa results in a tendency for tearing on suturing, even when only slight tension is applied[12]. Inferior and posterior superior dental block anaesthesia should be avoided in order to eliminate the possibility of injury to the fragile blood vessels, which would result in large dissecting hematomas[15,22]. Intraligamentary analgesia, as an alternative to the alveolar block anaesthesia, is advisable[12].

In addition to these general considerations, Type VIII EDS, which is distinguished from other forms because of its strong association with periodontal disease and loss of alveolar bone[21], should be managed from a different viewpoint. Scaling, periodontal surgery, and raising of mucoperiosteal flaps should be done with the greatest care to minimize gingival damage and prevent any lacerations[12]. Preoperative acrylic plate construction is suggested, so that suturing can be avoided in flap surgery[12]. Because of the fragility of the mucosa, suturing should again be performed with utmost care, especially when silk suture is used. Sutures, placed only with minimum tension, should be left intact for a longer period than usual[22].

Our patient also showed severe periodontal breakdown similar to cases reported in the literature. On the other hand, there was no ongoing active periodontal disease when the patient was referred to our department, due probably to previous extraction of the periodontally involved teeth. The only surgical treatment performed was extraction of tooth 20, which was unaccompanied by laceration of the gingiva or bleeding complications. However, although the extraction was intended to be simple and atraumatic, post operative
radiographic examination indicated minor cracks in the alveolar bone and moreover, healing of the wound was delayed. Fortunately, however, neither of these resulted in long-term complications. The patient is now under regular control.

It is quite clear that protection of the EDS patient by taking precautions against unnecessary trauma and stress is the most important consideration for dental treatment. A conservative approach should be sought, as in the case of most other disorders. Recognition of the dentofacial implications and the provision of the patient with appropriate information about daily home care and the need for regular follow-up should be considered by the dental professional as an indispensable part of the maintenance phase of the therapy.

**Conclusion**

Being a rare, autosomal dominantly inherited connective tissue disorder, Ehlers-Danlos syndrome (EDS) presents a number of important features for the clinician. The dental findings in a case of Type VIII EDS, which is of special interest to the dental practitioner, are reported and clinical management is discussed, giving special emphasis to a conservative approach.

**References**


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ecchymosis leading to diagnosis of Ehlers-Danlos syndrome: report of surgical management and 


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**Fig 1** Hypermobility of the joints at the shoulders

**Fig 2** Characteristic scar formation on the nose and forehead skin

**Figure 3** Panoramic radiograph showing horizontal impaction of tooth 5 and extensive resorption of the mandible