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Case Report

A Rare Cause of Tooth Failure of Eruption: Morphea with Intraoral Involvement

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Abstract

Background: In this study, the clinical and radiographic manifestations of a patient with linear morphea as well as her clinical features are reported with more emphasis on the oral effects of this connective tissue disease. This patient was referred to the orthodontic department due to the misalignment of her teeth and a diagnosis of linear scleroderma made three years earlier by her dermatologist. The most important intervention in such patients is to not start treatment without knowledge, since it will cause more harm to the patient. Therefore, it is necessary to find the best treatment plan with the fewest complications as soon as possible.

Keywords: Dermatologists, Female,Humans, Scleroderma, Localized

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Case Presentation

A 16-year-old girl presented to Orthodontic Department of Shahid Beheshti University of Medical Sciences with an aesthetic complaint. There was no history of facial trauma. The general health of the patient was good and she had no complaints of pain. Moreover, she neither smoked nor she used any drugs. At the time of evaluation, she described the presence of a linear indurated plaque that started along her head and spread to her lip and gingiva since she was 10 years old when a diagnosis of linear morphea was made by a dermatologist. Morphea, also known as Localized Scleroderma (LS), is a chronic inflammatory connective tissue disorder with variable clinical presentations, that affects both adults and children and the incidence rates vary from 3.4 to 27 per 100,000, and women are more susceptible than men. The differential diagnosis for morphea may include the following: skin granulomas, interstitial and granulomatous dermatitis, erythema migrans (1) also known as localized scleroderma, is a chronic inflammatory connective tissue disorder with variable clinical presentations, that affects both adults and children. It is characterized by inflammation and fibrosis of the skin and underlying soft tissue, in certain cases even of the surrounding structures such as fascia, muscle, bone and central nervous system. While the etiology is still unknown, many factors may contribute to disease development, including genetic predisposition, vascular dysregulation, T(H. For morphea, physicians may prescribe an immunosuppressive medication, such as oral methotrexate (Trexall), corticosteroid pills or both. Her only regular medication was EBETREX Tablet 2.5 *mg* for 3 years.

Among her facial problems, the most important one with the greatest impact on her social communication was her malocclusion and smile. Her oral problems started about 2 years after the onset of the cutaneous lesions. In fact, in the further stages of the disease, deciduous teeth fell out and the permanent teeth erupted late and incompletely. This issue itself caused a cant in the maxillary occlusal plane of the patient. Her family history was negative for genetic or autoimmune diseases.

In the extra-oral examination, there was a continuous depression involving the forehead, eyebrows, orbital rim, zygoma area, upper and lower lips, and chin on the left side. Facial expression muscles as well as cranial nerves had their proper functions. The eyebrows were not hairless in the hollow area. On the other hand, the vermilion thickness of the upper and lower lips was thinner in the affected area, and the buccal corridor looked wider. In the neck area, there was no sign of indentation, and no stiffness was felt upon touching the area (Figure 1 A-B); therefore, she had facial asymmetry and a linear patch on the left side.

The intraoral view showed the cant of occlusal plane due to the incomplete eruption of the teeth in the linear morphea area. In terms of oral hygiene, gingivitis and dental plaque indicated poor hygiene;

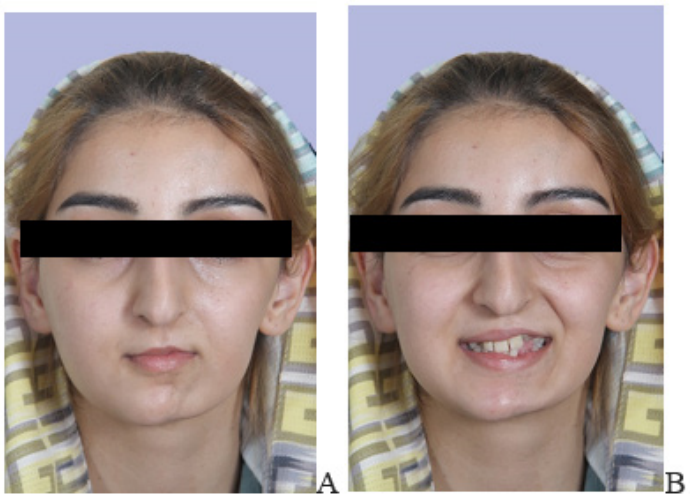

Figure 1. Clinical appearance. A: linear region on the left ala of the nose, lips and chin. B: asymmetric smile

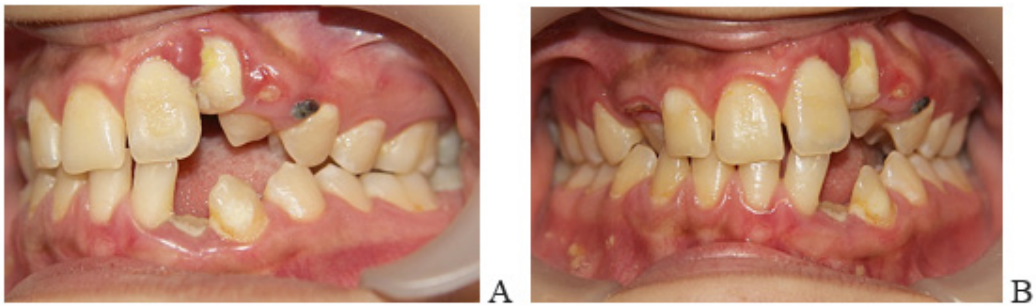

Figure 2. Intraoral manifestations. A: Periodontal inflammation, plaque and gingival enlargement B: the infraocclusal teeth are seen in the left side.

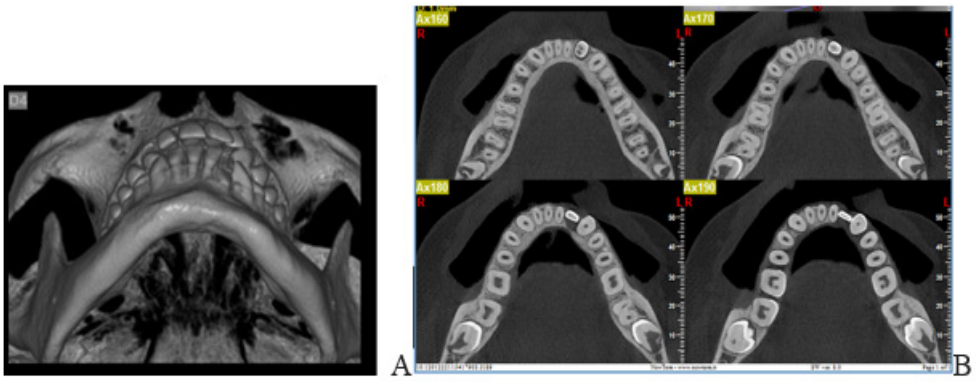

Figure 3. A: Submental view: asymmetric mandibular border. B: Axial section of mandibular cone beam computed tomography image.

furthermore, the residual roots of deciduous teeth were obvious. The orientation of the dental axis had changed in the area of linear morphea, the upper left canine and lateral incisor were in crossbite due to the lingual positioning of these teeth, and the central teeth were tipped toward the distal side and the midline was inclined. The position of these teeth was more palatal and gingival, which led to a one-sided open bite (Figure 2 A-B).

Cone Beam Computed Tomography (CBCT) radiography was used to evaluate the patient's bone status. In the initial overview of the RPR re-constructed from the CBCT, the difference in the size of the two sinuses was quite clear and the left sinus appeared larger than the right sinus. Submental view showed the asymmetric mandibular border (Figure 3A).

The axial section of the mandible revealed a radiolucent area around the lower left lateral, which could indicate bone resorption or lack of bone formation in that area. The cross-section of the roots, pulp chamber, and root canal can be seen in this view, which seemed normal with no noticeable difference between the two sides (Figure 3B).

Bone level deficiency could be seen in the left quadrants, and the occlusal level of the teeth in this area was lower than other teeth, thus the reverse curve of Spee could be visualized (Figure 4A). The length of the roots of the central, lateral, canine, and premolar teeth on the left side was short, while the crowns of the teeth did not show this shortness (Figure 4 B-D).

In the frontal view of the maxilla, the apexes of the left central and canine were blunt and there was periodontal ligament widening around the central apex. The maxilla had a reverse curve of Spee on the left side, both canines (R and L) were in crossbite, and the midline was deviated to the left. The upper canine and first premolar teeth and the lower lateral and canine teeth on the left side have not fully erupted. The oblique view on the left side of the 3D reconstructed image of the patient's CBCT showed low bone coverage of the roots and depression of the bone tissue. The ramus and posterior mandibular bone were normal (Figure 5 A-D).

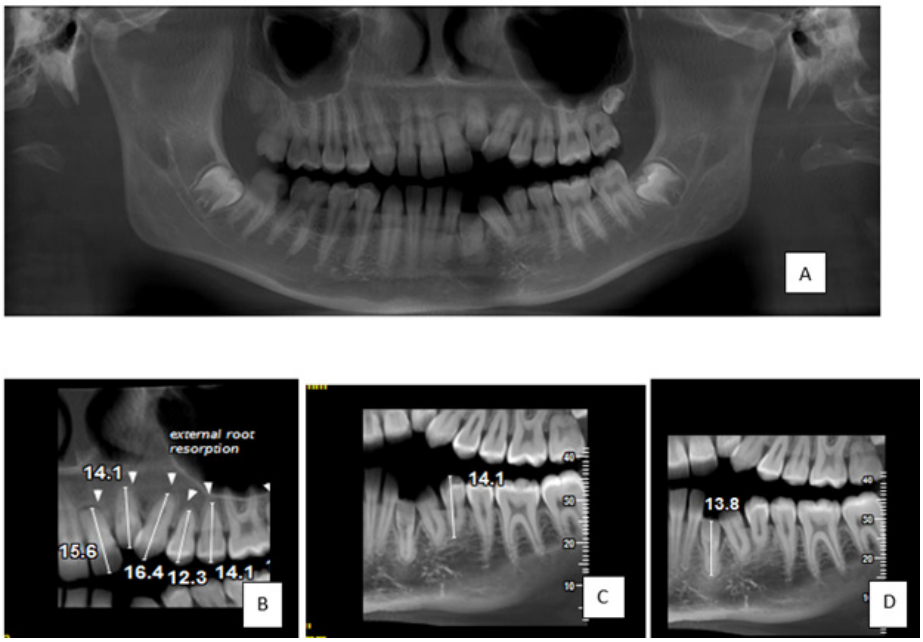

Figure 4. Radiologic findings. A: Bone level of upper and lower left quadrants are deficient. B-D short teeth in line of morphea.

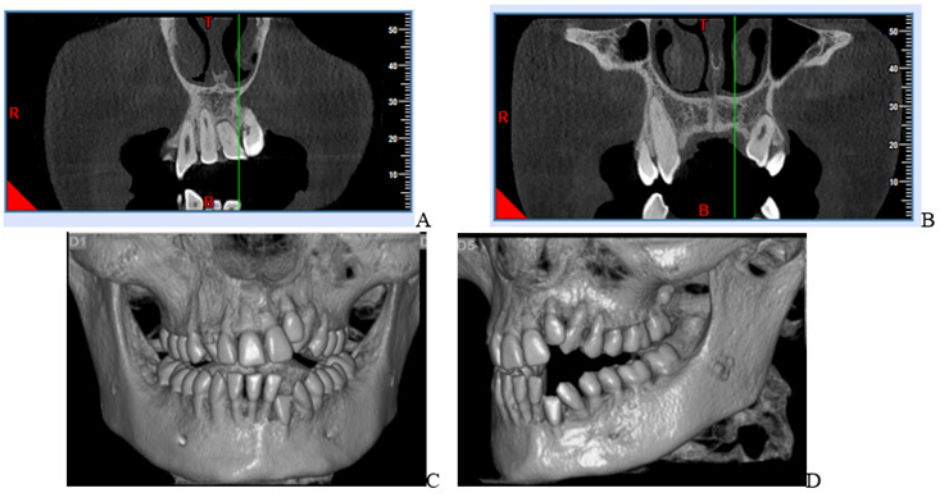

Figure 5. Radiography. A: Morphology of central roots B: Inclination of upper teeth. C: Three-dimensional reconstruction.

Discussion

Scleroderma is a rare connective tissue disease that manifests as cutaneous sclerosis and can have various systemic complications. Scleroderma means "hard skin", which is derived from the Greek words scleros (hard) and derma (skin). Its etiology is unknown; however, several risk factors such as age, sex, genetic background and environmental factors are known as influential factors. Two types of scleroderma are known, including systemic and localized or morphea. The systemic type is associated with skin sclerosis and vascular involvement, and the localized type is classically benign and self-limiting. It is limited to the skin and/or the underlying tissues (2,3) Morphea is a rare inflammatory skin condition that can also affect the subcutaneous tissues. Morphea is usually a clinical diagnosis and can be confirmed by physical examination. However, some patients need a skin biopsy, including subcutaneous fat, to confirm the diagnosis of morphea. A variety of factors, including genetics, environmental factors, such as infections, skin trauma, autoimmune dysregulation with abnormal cytokine production, and/or vascular

dysfunction may play a role in the development of morphea (4).

Morphea is classified into five groups, including plaque, generalized, bullous, linear, and deep. This classification is based on clinical morphological findings, which helps to simplify diagnostic and treatment goals (5,6). The most common site for the linear type of morphea is the face (2). Characteristically, LS is unilateral and does not extend below the eyebrow. The active stage usually lasts two to five years (7).

The age range of the development of the crown and root of the teeth is well established. In this case, due to the smaller effects of the disease on the crown of the permanent teeth at the site of the problem and the clear shortening of the roots, it is clear that the onset of the effects of the disease on the orofacial tissues was around 10-11 years of age when the cutaneous symptoms started.

Oral and dental treatments can have functional or aesthetic goals, or if possible, both. The most important aesthetic goal in orthodontics is to achieve a ‘‘balanced’’ smile (8). A patient’s smile can be examined from different angles in the vertical plane. Occlusal plane canting is one of the parameters affecting smile aesthetics, which can originate from facial asymmetry or asymmetric vertical position of the right or left dental quadrant.

Occlusal plane canting originates from facial asymmetry and/or vertical position asymmetry of the right and/or left quadrants of the dental arches without facial asymmetry (9). What motivated this patient to visit the orthodontics department was her dissatisfaction with the appearance of the orofacial set, especially smile aesthetics.

Before setting a treatment goal, the most important factor is a correct diagnosis; an incorrect diagnosis can easily cause treatment failure, for example, regarding a linear white patch starting along the gingiva and progressing to involve the mucosal surfaces, some dentists may consider it a fungal lesion and decide to treat it with nystatin solution (10). Active morphea lesions are especially important to identify, since they may be responsive to treatment with topical and systemic immunomodulatory drugs (11).

Approximately 67% of the patients with linear scleroderma are diagnosed before age 18 years (2). In the German guideline for the treatment of local sclerosis, it is mentioned that no common treatment has been introduced for this disease yet; however, there are effective methods in the active stage of the disease. The severity and the subtype of the disease are important in selecting a treatment method.

Treatment of LS should be graded according to the clinical subtype and disease severity. Topical corticosteroids (highly potent: once a day for up to 4 weeks, moderately potent: once a day for up to 3 months) are effective for subtypes with limited skin involvement (affecting the dermis).

Methotrexate (adults: 12.5–25 *mg* every week, children: 15 mg/m^2 every week, max: 25 *mg* every week) or systemic corticosteroids (adults: methylprednisolone 500–1,000 *mg* IV daily on three consecutive days every month for at least 3–6 months, children: methylprednisolone 30 *mg/kg* IV daily (maximum 1,000 *mg*) on three consecutive days every month for at least 3–6 months) are useful for subtypes with severe skin and/or musculoskeletal involvement (affecting the adipose tissue, fasciae, muscles, joints, and bones or extensive skin involvement) (12). In the present case, treatment started with EBETREX Tablet 2.5 *mg* for 3 years when she was 13 years old and the lesion was controlled; therefore, there was no pale linear region on her face. However, an atrophic region was obvious that was concave, especially in chin and inferior rim of the mandible. Other patients have such a depression on their chins (13). Our patient did not suffer from any mouth opening restriction; however, this problem may occur in the future as reported in other case reports. In such cases, intraoral examination may be difficult due to microstomia and limited mouth opening (3).

In fact, the most important intervention in such patients is to not start treatment without knowledge, since it will cause more harm to the patient. Therefore, it is necessary to find the best treatment plan with the fewest complications as soon as possible.

To the best of our knowledge, there is no report on the orthodontic treatment of such patients. Since the response of soft and hard tissue to orthodontic force in this patient is still unknown, any treatment should be done with caution. Due to the uncertainty of the outcome of orthodontic treatment, the final condition of the patient was stabilized by improving her oral

and dental hygiene and removing the roots of the deciduous teeth and educating her on the health of her gingivae so that in the future, with the help of other dentist colleagues, we will reach a treatment method to correct her defect.

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Conflict of Interest

The authors declare that there is no conflict of interest.

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