Case Report

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Scleroderma in the Pediatric Patient. A Clinical Case Report of Systemic Sclerosis

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ABSTRACT

Scleroderma is an autoimmune disease affecting the skin and sometimes internal organs of the body. There are two types of scleroderma, localized scleroderma or systemic. The most common type of scleroderma in children it is localized scleroderma. Systemic Scleroderma it is rare in children. It is reported that of all children with scleroderma, less than 10% have the systemic kind. The clinical history and anamnestic research are essential for the diagnosis and treatment plan. Scleroderma can affect oral and perioral tissues that can lead to difficulty in swallowing, reduction of mouth opening, xerostomia, limitation of masticatory function and difficulty in healing.

Abbreviations: SS: Systemic Scleroderma; VEDOSS: Very Early Diagnosis of Systemic Sclerosis; NSAIDs: Nonsteroidal Anti-Inflammatory Drugs; JSSc: Juvenile Systemic Sclerosis

Introduction

Definition

Scleroderma is an autoimmune disease affecting the skin and in some cases other organs of the body. In Localized scleroderma only the skin is involved with thickening and tightening of the skin [1]. When it involves internal organs it can affect the lungs, kidneys, heart, intestinal system and other areas, with inflammation and scarring.

Localised Scleroderma

There are different types of localised scleroderma. The most common form in children is Morphea scleroderma. It is characterized by irregular or round patches that start out small and pink in the skin. Linear Scleroderma is characterized of lines of tight, shiny and often darkened skin often in the arm or leg and extend to the hands or feet. This type of scleroderma can also affect the underlying muscle, fat and bone, leaving a scarred appearance. It is important to get this treated early in growing children, especially if a patch crosses over a joint, as normal growth and development can be affected. Linear scleroderma en coup de sabre is when children have linear scleroderma on their face and/or scalp [2]. When it involves the whole side of the face and in some cases the tongue, it is denominated Parry Romberg syndrome with additional difficulties because the facial bones may not grow properly.

Systemic Sclerosis

In Systemic sclerosis there is internal organs involvement such as the heart, lungs or kidneys, and can lead to difficulty in swallowing, heartburn, shortness of breath, high blood pressure and pulmonary hypertension. Children with systemic sclerosis usually have Raynaud's syndrome that it is characterized of paleness or blue of extremities and sometimes face caused by spasm of arteries that reduces blood blow. Systemic sclerosis is different in children from that in adults and is one of the most severe rheumatologic conditions diagnosed in children (Figure 2).

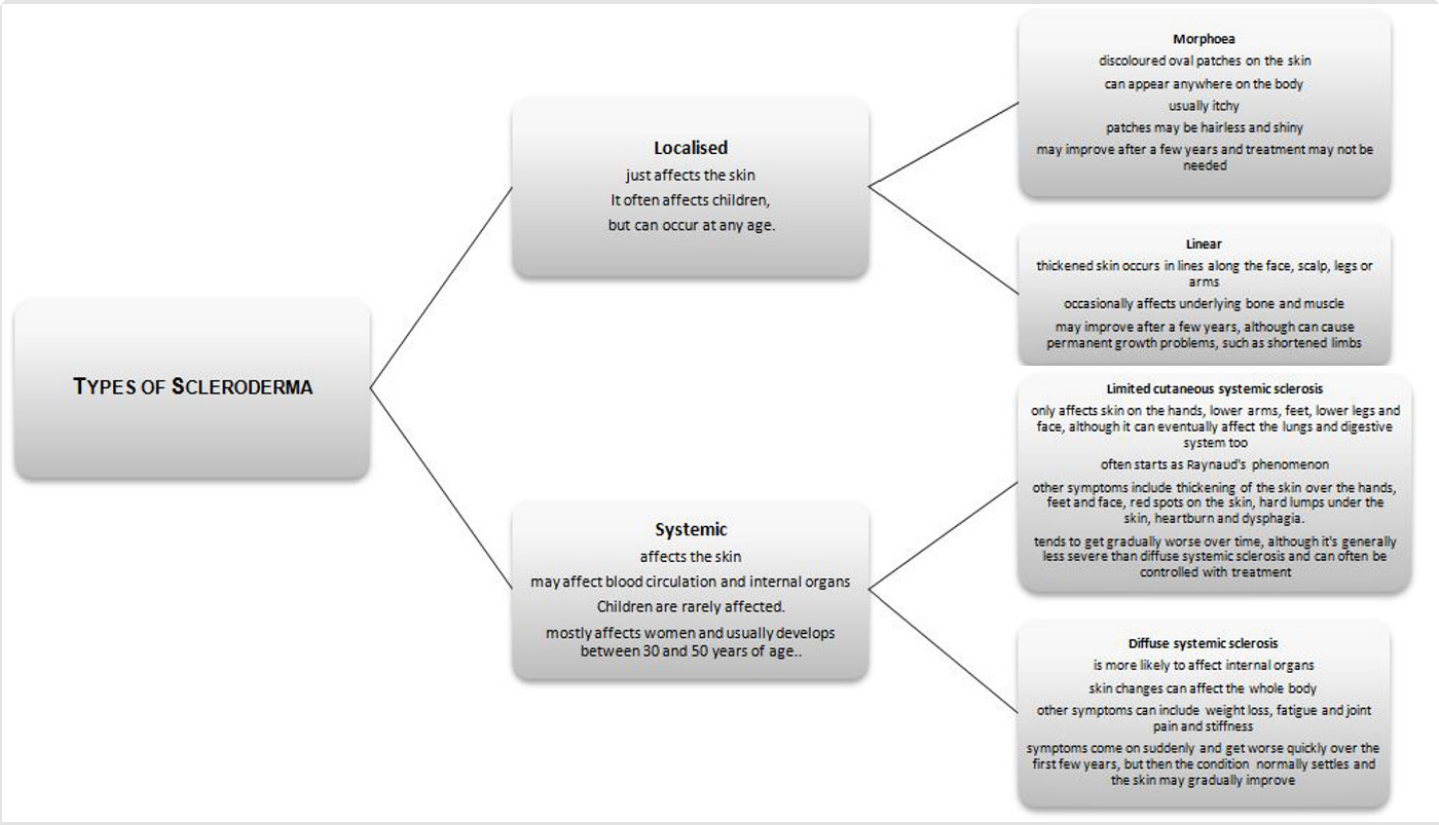

Figure 2

Aetiology

The cause of scleroderma is not known. There some evidence that certain genes are important hereditary factors, but the environment seems to also play a role. The result is activation of the immune system in a susceptible individual, causing damage to the inner lining of tiny blood vessels and injury to tissues that result in scar tissue formation and the accumulation of excess collagen [3]. Some evidence for the role genes may play in leading to the development of scleroderma comes from the study of Choctaw Native Americans who are the group with the highest reported prevalence of the disease.

Epidemiology

Most children who get the condition are diagnosed with localised scleroderma. A very small number of children are diagnosed with systemic sclerodermia [4]. SS it is rare in children. Of all children with scleroderma, it's less than 10% that have the systemic kind. Before puberty it can occur equally in boys and girls. After puberty, it occurs more often in girls.

Clinical Case Report

Presentation

A 5 - year-old girl attended with her parents to dental consultation. The parents motive for dental consultation was the concern about the lingual eruption of lower incisive (41) with absence of space for its regular eruption.

History

In the anamnestic research parents refer medication with methrotrexate and that the child has systemic sclerosis.

Examination

In the clinical examination it is observed blue paleness of the face and limitation of mouth opening.

Diagnosis

The clinical history highlights the existence of systemic scleroderma (SS). In the clinical examination it is observed the Raynaud's syndrome with blue paleness on the face and extremities.

Treatment

Selective extractions of 52 and 53 were performed to allow alignment of lower incisive (41 and 42). A lower removable appliance was used for dental alignment [5]. Mouth care instructions were provided. Preventive caries treatments were made. Oral hygiene techniques and motivation for correct oral hygiene were taught. The child was reviewed at 1 month intervals where it were carried out the activation of the removable appliance and showed significant resolution.

Investigations

Orthopantomography indicated for a full assessment of the teeth and bone. In the Orthopantomography it´s observed that despite the disease there are no bone changes (Figure 1). It is described that 10 to 20% of ES patients have varying degrees of posterior resorption of the mandible branch, coronoid process and condyle [6]. Dental resorptions may still occur and periodic radiographic monitoring is also required.

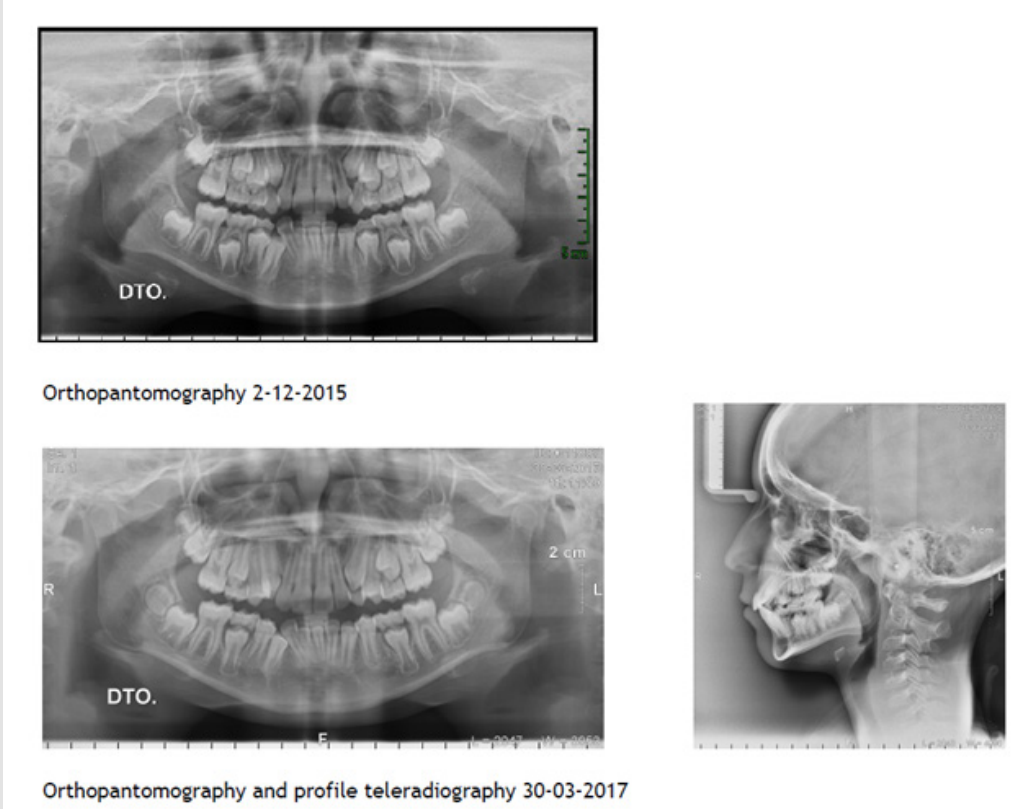

Figure 1

Discussion

Systemic scleroderma can affect oral and perioral tissues. The orofacial manifestations that may arise include increased periodontal ligament space, reduction of mouth opening, xerostomia, limitation of masticatory function and difficulty in healing, among others [7]. The higher incidence of caries in patients with this type of disease also leads to preventive treatments and periodic consultations. It is indicated that patients with SS have regular dental appointments [8]. The knowledge that the patient has Systemic scleroderma (SS) disease makes the pediatric dentist to act accordingly, as for example with the difficulty of mouth opening to make molds, because patients with SS have difficulty in stretching and keeping their mouths open which can make treatments extremely difficult [9].

Diagnosis

It´s important an Early diagnosis for the better prognosis of the disease [10]. The Very Early Diagnosis of Systemic Sclerosis (VEDOSS) initiative in Europe identified key features to diagnosing SSc in the very early stage, such as antinuclear antibodies Scleroderma-specific, antibodies SSc, pattern on nailfold capillaroscopy and Puffy fingers in Raynaud's syndrome patients.

Treatment

The therapeutic approach depends on the presentation of the disease and complexity of symptoms. There is topical treatment for the skin that may improve pain and ulceration [11]. To ease painful symptoms nonsteroidal anti-inflammatory drugs (NSAIDs) can be used, and/or steroids such as prednisone with limited benefit. Calcium channel blockers like nifedipine or dual endothelin-receptor antagonist bosentan may be beneficial for Raynaud's phenomenon, severe digital ulceration may respond to prostacyclin analogue iloprost. In children with Raynauds syndrome it's also important to keep the whole body warm, especially fingers and toes. It's also important to protect fingertips and other skin areas from injury, which can happen even during normal daily activities [12]. The skin tightness may be treated systemically with methotrexate and ciclosporin. And the skin thickness with penicillamine. It´s important to define the subtype (local or diffuse) as management differs. In particular, those with early diffuse disease should be considered for early immunosuppressive therapy.

Prognosis

Prognosis is dependent on the type and extent of the disease. The prognosis of limited forms of the disease (eg, morphea) in children is good. Often the skin patches will soften and fade leaving no serious long-term consequences [13]. Linear scleroderma in children also have often a good prognosis. The survival of juvenile systemic sclerosis (JSSc) at 5 years, 10 years, 15 years, and 20 years after diagnosis is 89%, 80–87.4%, 74–87.4%, and 69–82.5%, respectively. These survival rates are significantly higher than those reported in patients with adult-onset systemic sclerosis [14]. Progressive systemic sclerosis is a slowly developing, progressive and rare condition, which, when it involves internal organs, can lead to death. The most common causes of death are secondary to complications of cardiac, renal, or pulmonary involvement and failure.

Conclusion

The Knowledge of the definition, different types and clinical manifestations of scleroderma by the paediatric Dentist is extremely important to the management of the paediatric patient [15]. The pediatric dentist must ensure that a thorough and careful history is obtained to aid diagnosis of the paediatric patient, since anamnestic research may help [16]. From the knowledge of the disease, procedures should be performed so that the symptoms of the disease are minimized [17]. Patients with scleroderma have difficulty stretching and staying with their mouths open which can difficult dental treatments. Preventive treatments are also advised to decrease the incidence of caries and periodontal disease. Systemic scleroderma can affect oral and perioral tissues. The orofacial manifestations that may arise include increased periodontal ligament space, reduction of mouth opening, xerostomia, limitation of masticatory function and difficulty in healing.

It is described that 10 to 20% of patients with scleroderma have varying degrees of posterior resorption of the mandible branch, coronoid process and condyle. Dental resorptions may still occur and periodic radiographic monitoring is also required [18]. Scleroderma can have a strong impact on families and children. It may affect a child's ability to do physical activities and might mean numerous hospital visits with many hours out of school. Children with systemic sclerosis will need expert medical care and long-term support into adulthood. Children who died reported in studies were diagnosed almost 2 years later than the average child with juvenile systemic sclerosis. Early diagnosis is important as early treatment is thought to influence outcome [19]. The pediatric dentist play an important role in the earlier diagnosis identifying features in the child as being key to diagnosing scleroderma in the very early stage that lead to a better prognosis and well being of the child.

Conflict of Interest

The author declare no conflict of interest.

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