



Scleroderma in the common orthopaedic practice: A review

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Abstract

Scleroderma is a chronic, degenerative disease that affects the joints, skin, and internal organs. Scleroderma is also associated with blood vessel abnormalities. It occurs only rarely in children. There are two forms of scleroderma: generalized, that may also be systemic, and localized scleroderma. Localized scleroderma can be seen more frequently in children than the systemic form. The first signs of the disease involve reddish patches of the skin on the trunk, arms, legs, or head. Although the orthopaedic surgeon is rarely involved in the primary treatment of patients with scleroderma, he may be helpful towards making a diagnosis through an early referral to a rheumatologist or dermatologist.

Keywords

Scleroderma; Connective tissue; Sclerodactyly; Telangiectasia; Calcinosis; Linear Morphea

Scleroderma or systemic sclerosis (SSc) is a chronic autoimmune connective tissue disease that may be generalized or localized [1].

The two types of generalized scleroderma are described as diffuse cutaneous SSc (dcSSc) and limited cutaneous SSc (lcSSc). The former is rapidly progressing and affects internal organs. The latter is also called CREST syndrome (Calcinosis, Raynaud's phenomenon, Esophageal dysfunction, Sclerodactyly and Telangiectasia) [2].

Calcinosis is a particularly difficult entity to treat given the paucity of effective options described in the literature. Treatment of finger calcinosis has a wide range of possibilities depending on the extent of calcifications and the involvement of deep structures (Figure 1). From a surgical point of view, whereas simple removal is adequate in minor outpatient cases, a radical debridement in the major and more painful cases seems required. A cover flap is needed particularly in the thumb due to its great functional importance, also if the fingertip is not involved [3].

Localized (circumscribed) scleroderma or morphea (morphoea) has 5 main presentations: (1) circumscribed (few circles on the trunk or limbs); (2) generalized (many circles on the trunk and limbs); (3) linear (lines of involvement on the limbs or head); (4) mixed (combination of circumscribed and linear or generalized and linear); and (5) pansclerotic or deep (involvement of all of the skin) [4]. Four different variants of deep morphea have been described: Eosinophilic fasciitis, subcutaneous morphea, disabling pansclerotic morphea, and morphea profunda (subcutaneous) [5]. Morphea profunda is the most severe type of scleroderma localized on the skin [6].

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Received: November 30, 2016 Accepted: December 21, 2016 Published: December 28, 2016



Figure 1a: A 60-year-old woman was referred for a painful lesion of the tip of her right thumb.



Figure 1b: Radiographs indicated diffuse sclerotic lesions in the soft tissues of the distal phalanx of her thumb.



Figure 1c: She reported an over 15-year-history of Raynaud phenomenon and positive antinuclear antibody titer. The lesion was surgically debrided under local anaesthesia but the borders could not be identified. Biopsy showed localized calcinosis cutis. The patient was referred to rheumatologist where the diagnosis of Crest syndrome was made. Two weeks later the sutures were removed, and the scar was ruptured because of residual calcinosis in the soft tissues. By that time the borders of the lesions were easily identified and removed. No residual clinical lesion was evident 2 months post-operatively.

Solitary morphea profunda is an unusual, distinct, nonprogressive morphological variant [7,8].

Morphea is usually limited to the skin, but it may extend deeper to involve muscle or bone (Figure 2). It usually begins as a red or purple area of skin that then becomes thickened and white. The thick white areas usually thin out over time and turn brown. It is usually asymptomatic, with occasional itch and rarely pain. Morphea may also involve the inside of the mouth, the genitals, and the eyes [4]. MR imaging reveals musculoskeletal involvement in patients



Figure 1d: Radiographic evidence of minimal persistent calcinosis.



Figure 2: A 58-year-old woman was referred for a fractured left lateral malleolus. Scleroderma was diagnosed at 11 years of age from a 5 cm skin lesion of the thigh with skin biopsy. The lesion involved the whole leg 2 years later. She had 2 uneventful pregnancies and reported no disability.



Figure 3a: A healthy 6-year-old girl developed a plaque of linear morphea involving the right buttock, the posterior aspect of the thigh and the calf.

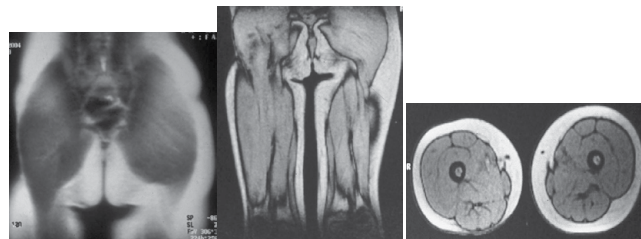


Figure 3b,c,d: Magnetic resonance imaging indicated thinner subcutaneous fat and atrophy of the muscles.

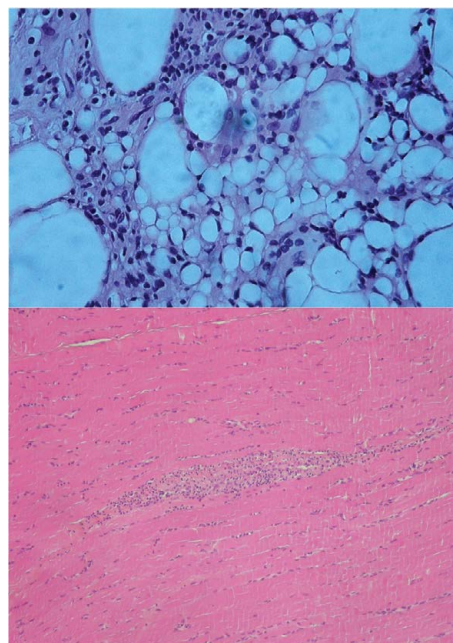


Figure 3e,f: Biopsy indicated inflammation to the subcutaneous tissue and muscle. She had no functional impairment, and the lesion remains stable ten years later.



Figure 4: A healthy 9-month-old girl developed a plaque of linear morphea involving the left groin.

with localized scleroderma [9-11]. Diagnosis is based on findings of skin examination and skin biopsy. Periodic follow-up is necessary to identify systemic evolution [12].

Scleroderma is rare in childhood. Because of the clinical and serological overlap, confusion is possible between dermatomyositis, scleroderma, juvenile rheumatoid arthritis, and systemic lupus erythematosus, particularly when these connective tissue disorders are so uncommon in children. It appears that scleroderma in children may be less severe than in adults and that systemic involvement is less common [13]. The presence of Raynaud's phenomenon, antinuclear factor, lupus erythematosus (LE) cells, and a raised erythrocyte sedimentation rate indicate a poor prognosis [14].

Juvenile localized scleroderma is about 10 times more frequent than juvenile systemic sclerosis. In recent years the time gap between the appearance of symptoms and diagnosis has become significantly shorter [15]. Juvenile localized scleroderma usually has its onset during later childhood.

Linear scleroderma is the most frequent form of scleroderma in childhood. It shows an increased prevalence in Caucasian girls [16]. It generally first appears in young children with different clinical and pathologic features compared with the adult-onset scleroderma. It most often occurs in the extremities and face [17]. It is also most likely to be on just one side of the body. It may cause growth defects and muscle wasting not seen in the adult equivalent (Figure 3). This condition may have devastating effects on growth and development such as limb asymmetry, flexion contractures, and psychological disability [18].

Congenital localized scleroderma is a rare and probably underestimated condition in neonates. The linear subtype is the exclusive manifestation of the disease (Figure 4). It should be included in the differential diagnosis of infants with cutaneous erythematous fibrotic lesions to avoid functional and aesthetic sequelae and to allow prompt therapy [19].

Scleroderma may be under-reported due to the rarity of the disorder in the common orthopaedic clinical practice and smaller morphea plaques may be less often referred to a dermatologist or rheumatologist. Furthermore, the infrequency of scleroderma in the pediatric population plus the fact that this disease is very often self-limiting makes randomized controlled trials very difficult. It is for this reason that most data on treatment modalities for this disease have been extrapolated from studies in adult patients. There is no therapy for systemic sclerosis or localized scleroderma that has proven to be very effective or significantly disease modifying [20].

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