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Localized scleroderma: imaging features

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Abstract. Localized scleroderma is distinct from the diffuse form of scleroderma and does not show Raynaud's phenomenon and visceral involvement. The imaging features in 23 patients ranging from 2 to 17 years of age (mean 11.1 years) were reviewed. Leg length discrepancy and muscle atrophy were the most common findings (five patients), with two patients also showing modelling deformity of the fibula. One patient with lower extremity involvement showed abnormal bone marrow signals on MR. Disabling joint contracture requiring orthopedic intervention was noted in one patient. In two patients with "en coup de sabre" facial deformity, CT and MR scans revealed intracranial calcifications and white matter abnormality in the ipsilateral frontal lobes, with one also showing migrational abnormality. In a third patient, CT revealed white matter abnormality in the ipsilateral parietal lobe. In one patient with progressive facial hemiatrophy, CT and MR scans showed the underlying hypoplastic left maxillary antrum and cheek. Imaging studies of areas of clinical concern revealed positive findings in half our patients.

Localized scleroderma (LS) is distinct from the diffuse form of scleroderma and does not feature Raynaud's phenomenon or visceral involvement. LS can be divided into linear scleroderma, morphea, and generalized morphea. Morphea appears as a circumscribed patch of skin induration, whereas linear scleroderma is characterized by a band of fibrous pigmented skin. LS is a disorder of connective tissues, with primary changes usually confined to skin and subcutaneous tissues, although it may also involve the underlying muscle, bone, synovium, and blood vessels [1, 2]. It is primarily a disease of childhood and is more common than systemic scleroderma in children [3]. As the radiological features have not been well described, we report the imaging features seen in 23 patients attending the rheumatology clinic for our hospital.

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Material and methods

The imaging studies of 23 patients (10 boys, 13 girls) with the clinical diagnosis of LS were reviewed. The patients ranged in age from 2 to 17 years (mean 11.1 years). The age at the time of onset of the LS ranged from 1 to 14 years (mean 7.9 years).

The areas of involvement by LS are shown in Table 1. Five patients had more than one area of involvement. Three patients with chest wall involvement had morphea, and the remaining 20 patients had linear scleroderma.

The studies available for review included plain radiographs, upper gastrointestinal and small bowel follow-through studies, and CT and MR scans of areas of involvement in selected patients.

Results

The most common findings in our series were leg length discrepancy and muscle atrophy, which occurred in five patients with linear scleroderma involving the lower extremities. These findings were evident on plain radiographs and, in one patient, on MR, which reveals atrophy of both the subcutaneous compartments and the underlying muscles of the involved extremities. Another patient with involvement of the left forearm showed similar changes on MR (Fig. 1).

In two of the five patients with leg length discrepancy, a bone modelling deformity of the distal fibular shaft was also seen adjacent to the area of skin involvement (Fig. 2). A sixth patient with involvement of the left lower

Table 1. Areas of involvement by localized scleroderma

Area	Patients
Face and scalp	7
Thorax	4
Upper extremities	6
Lower extremities	12

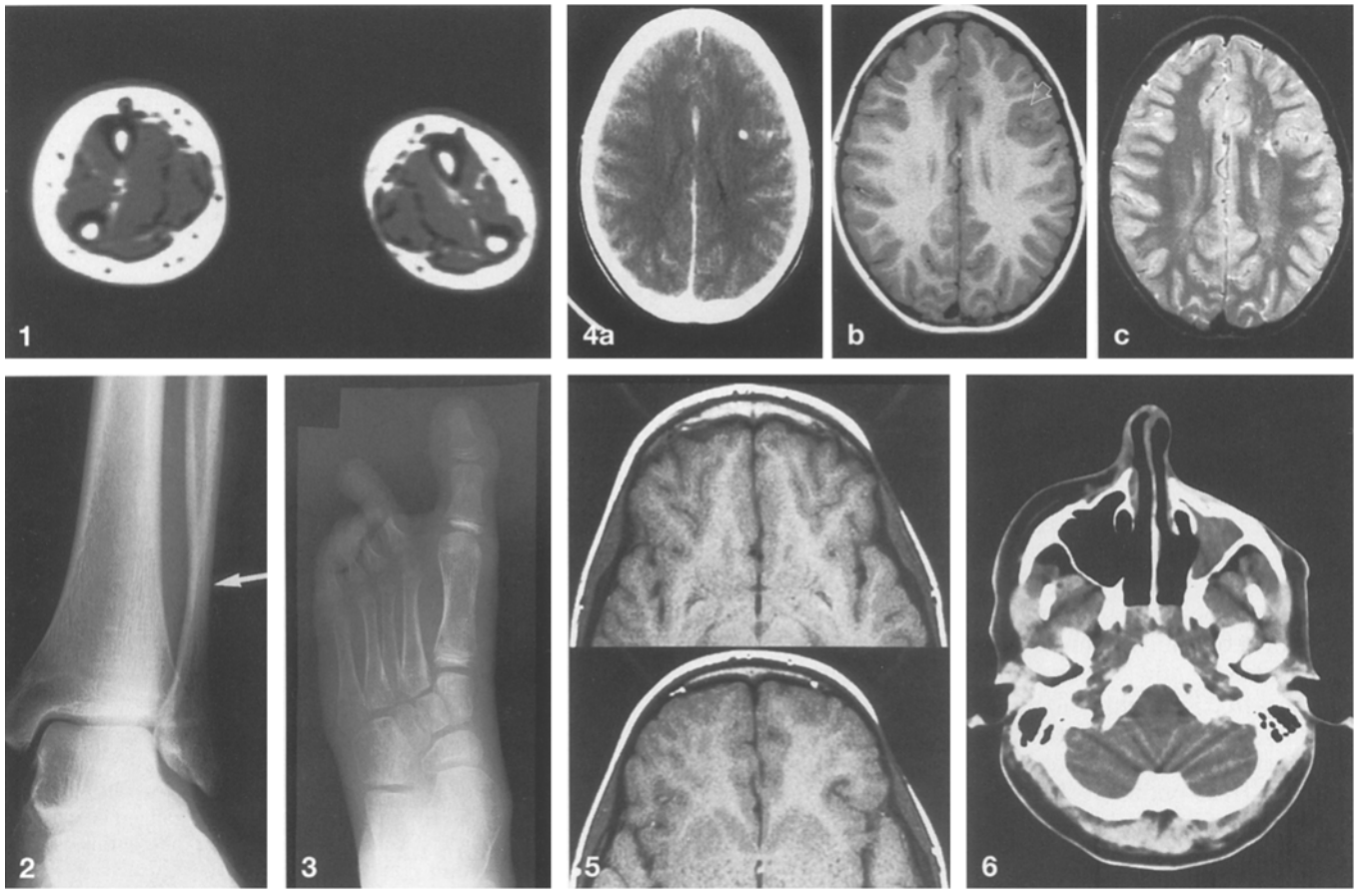

Fig. 1. A 14-year-old boy with a 3-year history of involvement of the left forearm. Axial T1-weighted image (TR 500/TE 11) reveals marked atrophy of the subcutaneous tissues and muscles of the left forearm

Fig. 2. A 16-year-old boy with at least a 3-year history of involvement of the lower leg. Plain radiograph shows bone modelling deformity of the distal fibular shaft, which is locally constricted *(arrow)*

Fig. 3. A 7-year-old girl. Frontal radiograph reveals left foot deformity with clawing of the third, fourth and fifth toes

Fig. 4a–c. A 4-year-old asymptomatic boy with "en coup de sabre" facial lesion. **a** CT reveals intracranial calcifications in the ipsilateral left frontal lobe. **b** Corresponding T1-weighted image (TR 550/TE 11) reveals bulky gray matter in the same region *(open arrow)*. **c** T2-weighted image (TR 2500/TE 90) reveals focal areas of high and low signal intensities, probably representing focal areas of dysmyelination or demyelination and calcification in the area of migrational abnormality

Fig. 5. A 10-year-old boy with linear scleroderma involving the left temple. T1-weighted images (TR 600/TE 20) reveal focal thinning of the subcutaneous tissues in the involved area

Fig. 6. A 15-year-old boy with progressive facial hemiatrophy on the left side. CT reveals marked hypoplasia of the underlying left maxilla. Partial maxillary sinus opacification is also present on this study

extremity showed mild atrophy of subcutaneous tissues and muscles on MR. In addition, MR also showed abnormal bone marrow signals in a patchy fashion in the femur and tibia, with low signal intensity on T1-weighted images, and high signal intensity on T2-weighted images. Linear scleroderma involving the left ankle and crossing the joint line resulted in a severe and disabling joint contracture of the left ankle and foot in a 7-year-old girl (Fig. 3).

Three patients had skin lesions involving the face and scalp, with the "en coup de sabre" deformity. All three were neurologically asymptomatic. Two had CT scans showing intracranial calcifications in the ipsilateral frontal lobe (Fig. 4 a). In one of these two patients, the corresponding MR scan revealed abnormal bulky gray matter in the area of intracranial calcifications seen as areas of signal void on T1- and T2-weighted images, and areas of abnormal white matter seen as high signal intensities on T2-weighted pulse sequences. The MR appearance was consistent with a migrational abnormality, with focal areas of demyelination or dysmyelination and calcification (Fig. 4 b, c). The second patient also showed similar areas of high and low signal intensities on MR, but with possible migrational abnormality as well. The third patient, a 3-year-old boy, had a focal area of low attenuation in the white matter of the ipsilateral right parietal lobe on CT, which was consistent with a focal area of demyelination, but unfortunately MR was not available for this boy.

In two other patients with facial involvement, imaging studies revealed a focal thinning of the subcutaneous tissues in one (Fig. 5), and focal thickening of the subcutaneous tissues in the area of involvement in the other. No underlying bony abnormality was seen. One patient with extensive facial involvement or Parry-Romberg

syndrome (progressive facial hemiatrophy) had marked hypoplasia of the underlying maxilla on the CT and MR scans (Fig. 6).

Four patients underwent upper gastrointestinal studies: two who had occasional abdominal discomfort and two who were asymptomatic but whose physicians were looking for systemic involvement by the sclerodermatous process. One asymptomatic patient had moderate gastroesophageal reflux. One symptomatic patient had a small sliding hiatal hernia. The remaining two patients had normal studies. No radiological abnormality was seen in the three patients with morphea.

Discussion

Linear scleroderma is characterized by the presence of areas of tight, hard, indurated skin with a linear configuration. These lesions are usually found on the skin of the extremities, face and scalp. The onset of linear scleroderma is usually slow and insidious, and a typical patient has the active disease for about 3–4 years [1, 4]. It is usually a superficial and self-limiting disease.

The etiology is unknown, but an immunologic mechanism is most likely. Hypergammaglobulinemia, antinuclear antibodies, and antibodies to single-stranded DNA are present in up to half the patients with linear scleroderma [1]. We have recently reported that levels of serum-soluble interleukin-2 receptor, a marker of immune activation, are elevated in children with active disease [5]. Progress to systemic scleroderma is extremely unusual. In one series of 53 patients, none developed Raynaud's phenomenon or signs or symptoms of systemic lupus erythematosus or progressive systemic sclerosis after a mean follow-up time of 10 years [1]. Unlike systemic scleroderma, linear scleroderma appears benign with regard to life expectancy [1, 4].

Even though linear scleroderma is usually a superficial disease and does not require any imaging investigations, it can give rise to local growth disturbances, as seen in five of our patients. The findings include leg length discrepancy, bone remodelling deformity, and atrophy of the subcutaneous tissues and underlying muscles. In severe cases, these abnormalities can result in significant functional impairment and cosmetic disfigurement even when the linear scleroderma is inactive clinically. Linear scleroderma lesions often cross joint lines, and may lead to disabling joint contracture as a result of the deep extensions of the cutaneous lesions into the muscles and bones. Orthopedic intervention may be required (Fig. 3). This is in contrast to the reported findings in systemic scleroderma, where localized bone resorption of unknown etiology is a prime radiologic finding [6, 7].

When linear scleroderma occurs on the face or scalp, it is referred to as scleroderma "en coup de sabre" [3]. More extensive involvement, especially of the lower face, is known as progressive facial hemiatrophy or Parry-Romberg syndrome. Occasional reports of epilepsy and hemiparesis in patients with facial or scalp involvement appear in the literature. Ipsilateral cerebral calcifications have also been reported in at least seven patients with linear scleroderma involving the face or scalp [8, 9]. The etiology of intracranial calcifications is unknown, although some researchers have postulated that they may be due to calcified hemangiomas [10, 11]. The 10-year-old girl we describe, with ipsilateral calcification in the left frontal lobe, also demonstrated a migrational abnormality with small areas of abnormal myelination in the same region. To our knowledge, this has not been reported previously and may possibly be associated with some of the intracranial calcifications previously described.

MR, which is a more sensitive modality than CT, was able to show increased signals in the ipsilateral white matter on T2-weighted images in five out of six patients with progressive facial hemiatrophy [8]. However, there was no correlation of CT or MR findings with the clinical or neurological signs and symptoms. Most patients were asymptomatic apart from cosmetic or functional problems.

In summary, this short review revealed positive findings in half our patients. The peripheral findings included leg length discrepancy, muscle atrophy, bone modelling deformity, abnormal bone marrow signals on MR, and joint contracture. The central nervous system findings included inracranial calcifications, white matter abnormalities, and migrational abnormality. Even though LS is usually a superficial disease, our study concurs with previous reports that it may sometimes lead to significant functional impairment and cosmetic disfigurement. LS is an entity distinct from systemic scleroderma.

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