Juvenile Linear Scleroderma: A Case Report

Apurva Khare, Prashant P Choubey
Department of Medicine, People's College of Medical Sciences & Research Centre, Bhanpur, Bhopal - 462037
(Received: June, 2014) (Accepted: July, 2014)

ABSTRACT:
Linear Scleroderma is a rare type of localized scleroderma which occurs most frequently in childhood, but also may develop in adults. We report a 14 year old female, who on basis of thorough clinical examination was diagnosed as case of Linear Scleroderma.

KEY WORDS: Linear scleroderma, localized scleroderma.

INTRODUCTION:
Localized forms of scleroderma are distinguished from Systemic Sclerosis, not only by the absence of vasospasm, structural vascular damage, and involvement of internal organs, but also by the distribution of the skin lesions.\(^\text{[1,2]}\) The three main varieties of localized scleroderma are morphea, linear and coup de sabre. These conditions are characterized by localized inflammation and fibrosis of the skin and the underlying tissue. In this very rare process, sclerotic areas have a linear, bandlike pattern and often follow a dermatomal distribution mostly along limb, less commonly on trunk\(^\text{[3]}\) [Figure 1]. It has incidence rate of 2.5 per million children.\(^\text{[4]}\)

CASE REPORT:
A 14 year old female (age as mentioned by patient’s mother) presented with insidious onset of thickening and depigmentation of skin over left arm since 8 years. Patient gave history of blunt trauma to left index finger\(^\text{[5]}\), after which she noticed thickening of skin at the tip of the finger, which progressed to involve whole of the left index finger over the period of 6 months; radial aspect of left hand, up to level of elbow over 2 years, and present days condition of involvement of radial aspect of whole of left hand, up to anterior chest wall, over 8 years [Figure 2]. There was no history of exposure to any drug or chemical, febrile illness, dysphagia, Raynaud's phenomenon or dyspnea. There was no significant family history of similar episode. On examination, the skin thickening was along the radial aspect in dermatomal pattern. There were linear streaks of depigmentation over effected limb extending up to anterior chest wall [Figure 2]. There was also ‘Claw hand’ deformity in effected limb and flexion contracture at elbow joint [Figure 3].There was no sensory or motor deficit.

On investigation, all routine investigations including complete hemogram, urine routine, renal and liver function tests, chest X-ray, ECG were within normal limits. Specialized test like ANA (indirect immunofloroscence) was negative. Patient and her parents did not give consent for skin biopsy.

Patient was treated with low dose steroids (15mg/day of prednisone), immunosuppressive like...
hydroxychloroquine (200mg/day in OD dose), local application of cream containing vitamin D and physiotherapy for the limb. Surgical corrections were not considered as limb growth was not affected. Methotrexate was not used because of its doubtful

Table 1: Treatment of localized scleroderma in Adults and Children.[6]

<table>
<thead>
<tr>
<th>Pattern of Disease</th>
<th>Clinical Features</th>
<th>Treatment</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plaque morphea</td>
<td>One or a few circumscribed sclerotic plaques with hypopigmentation or hyperpigmentation and an inflamed violaceous border</td>
<td>Often unnecessary. Topical steroids, immunosuppression (e.g., tacrolimus) or phototherapy may be considered. Serial measurement to assess progress</td>
<td>Good prognosis. Lesions less active within 3 yr, but pigmentary changes often persist</td>
</tr>
<tr>
<td>Generalized morphea</td>
<td>Widespread pruritic lesions, often symmetric and following distribution of superficial veins</td>
<td>Suppress inflammatory component using corticosteroids: in children oral doses 15 mg/day have been used. Intravenous infusions often effective. Methotrexate, immunosuppressive maintenance therapy often used, although benefit not proven in controlled trials. Vitamin D–containing creams may be useful. Topical corticosteroids rarely helpful. PUVA has been used</td>
<td>Internal organ pathology or Reynaud’s phenomenon rare. Generally improves within 5 yr of onset, although textural and pigmentary changes may persist</td>
</tr>
<tr>
<td>Linear scleroderma</td>
<td>Sclerotic areas in linear distribution on limbs, as symmetric; in childhood can lead to growth defect. Serial measurements of limb length and girth essential to monitor progression</td>
<td>Suppress inflammatory component using corticosteroids: in children oral doses 15 mg/day have been used. Intravenous infusion has been used. Methotrexate, other immunosuppressive maintenance therapy often used, although benefit not proven in controlled trials. Vitamin D–containing creams may be useful. Physiotherapy and appropriate regular exercise important to minimize growth defect in childhood disease. Surgical correction of limb defects may be considered when disease is inactive</td>
<td>Long-term effects of childhood-onset form are minimized by effective suppression of the inflammatory process and by good physiotherapy. Tends to resolve, but can remain active for years</td>
</tr>
</tbody>
</table>
beneficial role in localized scleroderma (Treatment guidelines as per Table 1). There are few studies, showing increasing role of Methotrexate in the treatment of Localized Scleroderma.

Patient is under treatment for 1 month, and her follow up visit is due after 6 weeks of treatment.

DISCUSSION:

Early localized scleroderma lesions are associated with inflammatory changes in the dermis and violaceous discoloration, and are often accompanied by localized itching or discomfort. With time, the lesions become indurated and sclerotic, and in late stages there may be a pale waxy central area with an erythematous border. Eventually, the skin texture softens, although pigmenatory abnormalities often persist. Linear scleroderma can be associated with growth failure that may be profound; on the face, which leads to hemifacial atrophy, also known as Parry-Romberg syndrome.[1] Progression of localized scleroderma to Systemic Sclerosis almost never occurs. When linear scleroderma lesions cross joint lines, they can be associated with atrophy of the soft tissue, muscle, periosteum and bone. The etiology of localized scleroderma is unclear. Local triggers, such as trauma, are implicated, and associate with infection, including Borrelia burgdorferi, has been described.[5]

CONCLUSION:

Morbidity is high in juvenile variant of localized Scleroderma in form of disfigurement of affected part of the body, even affecting growth of limb in many cases. Hence, it should be diagnosed earliest and treatment initiated. Newer tools in its early diagnosis and newer modalities of its treatment are warranted in future.

REFERENCES:


Source of Support: Nil, Conflict of Interest: None declared.