CASE REPORT

Scoliosis in a Patient with Lipodystrophy - Treatment Difficulties and Literature Review

H H Lim, FRCSE MchOrth FAMS, C S Chong, MBBS, Department of Orthopaedics, University of Malaya, Lembah Pantai, Kuala Lumpur

Summary

A 12 year-old Chinese schoolgirl presented with left-sided scoliosis at the age of 9 years. She has a rare defect in lipid metabolism, which is not known to be associated with spinal deformity. Her scoliotic curve deteriorated despite bracing. We report a rare occurrence of scoliosis in patient with lipodystrophy and the difficulty of using instrumented fusion in treating this condition.

Key Words: Adipose tissue, Instrumentation, Lipodystrophy, Scoliosis, Treatment

Introduction

Lipodystrophy is a rare disorder in which there is a loss of subcutaneous fat prominent enough to allow easy recognition at a glance. Clinically, two groups of patients, those with partial lipodystrophy syndrome and those with total lipodystrophy syndrome can be identified.

Mitchell[1] first reported the syndrome of partial lipodystrophy in 1885 describing it as a disorder affecting predominantly female between the age of 5 to 10 years. There is selective regional loss of fat, from the face, neck, arms, abdomen and chest, sparing the lower limb.

Lawrence[2] reported a syndrome of total lipodystrophy in 1946 with a complete loss of adipose tissue over the whole body. The age of onset is either at birth or very soon after. There is a frequent association with hepatomegaly, hyperlipidaemia, and hypermetabolism.

Scoliosis and lipodystrophy has yet to be described in the literature as an associated condition thus the incidence and type of scoliosis are not well defined in these patients. However, many of these patients were said to be tall as compared to their sibling. The absence of subcutaneous fat presents a problem in the treatment of progressive scoliosis. This predicament of insufficient soft tissue padding, is also encountered in scoliotic patients with Arthrogyposis Multiplex Congenita and Marfan Syndrome but in a lesser degree. We wish to highlight the various consequences of this problem.

Case Report

Ms W A L, a 13 year old schoolgirl was noted to have scoliosis by the school health doctors at the age of 9 years old. She was born full term by normal vaginal delivery. Her developmental milestone was normal. Her menarche was at 12 year old. She has an older sister who does not have scoliosis and is not affected by lipodystrophy. Her parents are not affected by lipodystrophy. She was noted to be extremely slim at a very young age and was fully investigated by a paediatrician in her hometown.

Clinical examination showed a thin girl with prominent scoliosis with convexity to the right. There was a right rib prominence and the spinous process was easily palpable (Fig. 1). There was abnormal bony prominence of the condyles of the limbs. There were no symptoms and signs of any endocrine abnormally.

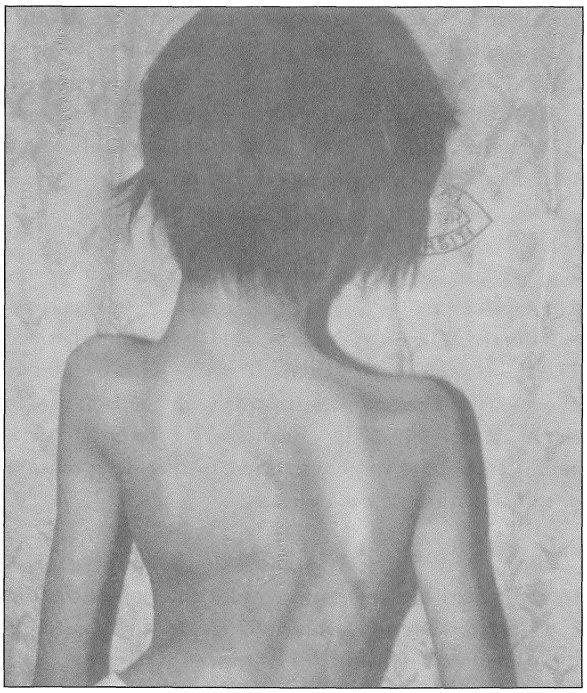

Fig. 1: The back of the patient showing the lack of subcutaneous fat.

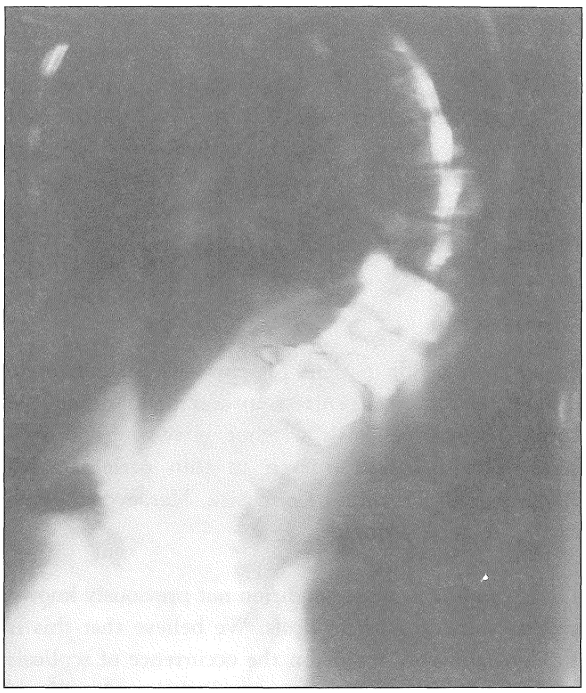

Fig. 2: Radiograph showing the severity of the scoliosis.

Radiological examination revealed a very severe King's II scoliosis with marked rotation of the spine (Fig. 2). At that time she had Risser II iliac apophysis indicating further potential for rapid growth of the spinal column is likely. There were no evidence congenital vertebra abnormalies. Serum biochemistry namely renal function, blood glucose level, serum triglycerides were normal.

At the outset, her scoliosis was thought likely to progress further and was treated on a brace, which she poorly tolerated because of the discomfort of the bony prominence. Numerous attempts to improve the padding of the brace were futile as she developed sores over multiple pressure points. She was non-compliant with bracing and the severity of the curve worsens (Fig. 2). Her respiratory function showed a markedly abnormal values (Table I). A decision to prevent further scoliosis progression by surgical means was made.

Preoperatively, SPECT was performed to confirm the sites and number of hyperactive growth plates along the convexity of the curve that needed to be obliterated. Clinical assessment of the subcutaneous tissue showed that the use of posterior spinal implants was unwise as skin closure and implant prominence would be a problem.

Table I
Preoperative Lung Function Results

	Predicted Value	Patient Value	Percentage-Value
FVC	2.27	0.78	38%
FEVI	4.44	2.74	62%
FIVC	2.06	0.69	34%

Convex epiphysiodesis was performed, from T9 to L1, by using match-stick shaped rib grafts to fused the growth area. During the operation, it was noted that there was a complete absence of subcutaneous fat and spinal muscles were atrophied. Postoperatively, she was allowed to ambulate without spinal brace or jacket. Latest review, a year after surgery, showed solid fusion of the growth plate and no progression of the curve.

Discussion

Scoliosis in a thin patient present with an extremely difficult problem for correction and maintaining the correction with orthosis without it causing pressure sores. This situation is seen in thin patients with Arthrogyposis Multiplex Congenita, Marfan Syndrome and Muscular Dystrophy.

Lipodystrophy is a rare condition not previously known to be associated with scoliosis. We believe that this is the first and only report on the occurrence of scoliosis with this condition. It is not implied that there is an association neither can be conclusively be attributed to the metabolic disorder or to an incidental finding.

In this condition, there is a derangement of fat metabolism of unknown cause with reported metabolic disturbances including hyperlipidaemia and hypermetabolism. Pulmonary stenosis, insulin-resistant diabetes mellitus, nephropathy, hypertension, hirsuitism and acanthosis nigra has also been reported to be associated with this condition. Cardiac involvement may includes cardiomegaly, hypertrophic cardiomyopathy and assymptomatic murmurs. There are a number of patients with hepatomegaly and splenomegaly. This patient had been assessed by a paediatrician to be free of all these complications.

The etiology of the scoliosis in this patient unknown. The type of curve is King type IV and the curve is rigid and rotated. The progression is rapid over a short period. The Cobb angle worsen from 30° to 75° within a period of 6 months. Bracing was difficult with the lack of soft tissue cushion. The respiratory function was compromised and surgery to arrest further deterioration of spinal deformity was indicated.

Various conventional approach to surgery was contemplated but rejected includes anterior discetomy and instrumentation, anterior release and posterior instrumented fusion. Anterior release and instrumentation can only be done, when the length of screw fits the size of the vertebral body and is not excessive long, which was in this case. Anterior release and posterior instrumentation was rejected as implant prominence would have been a major concern. We chose to differential growth arrest procedure to prevent further deterioration of the deformity until the vertebral body is large enough to accommodate vertebral screws.

References

1. Mitchell S W: Singular case of absence of adipose matter in the upper half of the body. Amer-J-Med-Sci 1885; 90: 105.

2. Lawrence RP: Lipodystrophy and hepatomegaly with diabetes, lipaemia and other metabolic disturbance. A case throwing light on the action of insulin. Lancet 1946; 1: 724-73.