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CASE REPORT

Liposuction for lower limb lipodystrophy in congenital analbuminaemia: A case report☆

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Summary Congenital analbuminaemia is a very rare autosomal dominant disorder in which patients have no serum albumin and markedly low serum total protein concentration. Clinically patients present with mild oedema, hypotension, fatigue and lipodystrophy often with abnormal body habitus. With only around 50 reported cases in the literature worldwide, management of the resulting lipodystrophy remains unclear.

A 42-year-old male who was diagnosed with congenital analbuminaemia presented with bilateral lower limb lipodystrophy disproportionately affecting his thighs. This was associated with concerns over appearance, difficulties with mobility and finding clothing. He successfully underwent bilateral lower leg liposuction and has had no recurrence of his symptoms after 12 months.

We have demonstrated that liposuction along with controlled compression therapy is a safe and effective treatment for managing lipodystrophy secondary to congenital analbuminaemia. Although rare, clinicians need to be aware that liposuction is a successful treatment modality, which should be made available to this select group of patients.

Introduction

Congenital analbuminaemia is a very rare autosomal recessive disorder first described in 1954 by Bennhold et al.,[1] with only 50 cases reported worldwide since. In several patients, genetic analyses have revealed point mutations at various regions or nucleotide insertion in the albumin gene leading to albumin fragment production instead of the complete molecule.[2] In homozygotes, only 1/50—1/2000 of physiological albumin concentrations is

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measured whilst only slight hypoalbuminaemia occurs in heterozygotes.[3] Surprisingly, despite the virtual absence of albumin, analbuminaemic patients can obviously survive although there is clear evidence of frequent abortions in the affected families, pointing to the critical role of albumin or the development of compensatory strategies, demonstrated by raised levels of non-albumin proteins.

Clinically patients present with mild oedema, hypotension, fatigue, hypercholesterolaemia and lipodystrophy often with abnormal body habitus. Liposuction in general has been proven to be a low-risk curative procedure for removing extensive lipohypertrophic tissue however with the scarcity of cases of analbuminaemia in the literature, management of the resulting lipodystrophy remains unclear. We present a case in which liposuction along with controlled compression therapy has been shown to be successful in the management of bilateral lower limb lipodystrophy in a patient with congenital analbuminaemia.

Case report

A 42-year-old male was referred to the lymphoedema clinic with a 3-year history of increasing swelling primarily involving the thighs. Following numerous investigations, he was diagnosed as having congenital analbuminaemia and associated lipodystrophy. Previous lymphoscintigraphy revealed lymph drainage abnormalities, however, the most striking finding was an almost unrecordable plasma albumin level. The patient's main complaints were cramps in both legs, difficulty finding clothing and psychological upset. His legs continued to grow in size despite trying a low fat diet. He had also developed subcutaneous lipomas on his torso and right forearm. He had no medical history of note and did not take any regular medications.

On examination, he was found to have significant lipodystrophy affecting the thighs, with the left being worse than the right (Figure 1). The lower legs were essentially normal apart from some pitting oedema although there was a negative Stemmers sign. Lower leg volume measurements were 14,163 ml on the right and 15,674 ml on the left. Following pre-operative counselling the patient was offered liposuction, starting initially on the left thigh.

Liposuction technique

The liposuction technique and post-operative care were identical to that previous described by Brorson.[4] The patient was admitted the day before surgery and routine pre-operative testing was performed. Under spinal anaesthesia, tumescent infiltration of 4 L of Hartmanns Solution (including 1 mg of adrenaline 1:1000 and 40 ml of 0.5% Chirocaine) was administered in the subcutaneous tissue of the left thigh. Liposuction was performed through multiple 5 mm stab incisions and 5 L of aspirate was obtained (supranatant 4500 ml, infranatant 1900 ml). A tubigauze liner and FarrowWrap® compression garment were applied immediately post-operatively followed by 48 h of intravenous antibiotics and elevation. A dressing change was performed after 2 and 4 days. The patient had an uneventful recovery and was discharged home after 4 days. The compression garment was used continuously.

After two weeks, a significant reduction in leg volume had been achieved on the left side which measured 11,207 ml and 14,841 ml on the right. He was advised to continue with FarrowWrap® compression and after three months, his post-operative measurements were 10,420 ml on the left and 15,794 ml on the right. After 6 months he underwent liposuction to the contralateral thigh as

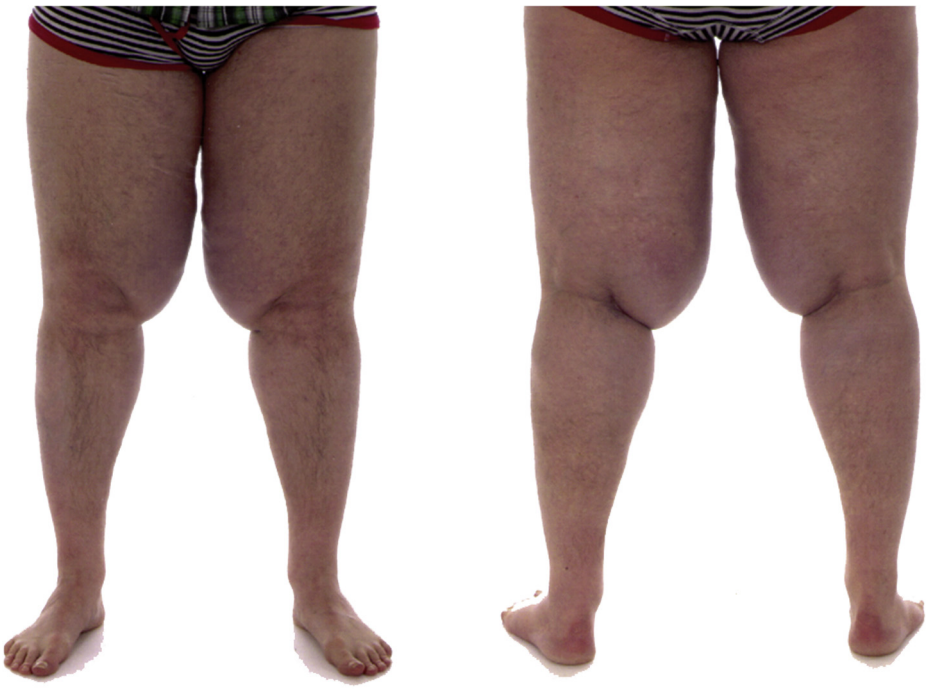

Figure 1 A 42-year-old man with lipodystrophy secondary to congenital analbuminaemia affecting both thighs (pre-op). Leg volumes were 14,163 ml on the right and 15,674 ml on the left.