OPEN FORUM

Body Contouring

Case Report

Infantile Asymmetrical Diffuse Infiltrative Lipomatosis of the Abdomen and Upper Thighs: A Case Report with Long-Term Follow-up

Fawzy Hamza, MD; Tarek Elbanoby, MD; Hazem Dahshan, MD; and Amr Elbatawy, MSc, MD, EBOPRAS, MRCS

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Abstract

The authors present the case of an 11-year-old male patient with a diffuse infiltrative lipomatosis involving the abdomen, flanks, and upper thighs by applying body contouring principles at this early age. Abdominoplasty can be used in children for various purposes, including harvesting a full-thickness skin graft in burns or to treat congenital anomalies involving the pelviabdominal area.

Level of Evidence: 5

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In 1846, Benjamin Brodie described multiple lipomatosis in 2 patients "as large as two oranges."[1] Madelung[2] delineated the lipomatosis in the cervical region and called it "Madelung Syndrome" in 1888. A year after, Launuis and Bensaude defined the diffuse infiltrative lipomatosis (DIL) as the formation of multiple non-encapsulated lipomas with a symmetrical or asymmetrical distribution that may involve any part of the body, including face, trunk, and upper or lower limbs.[3] In this report, we describe the application of principles of body contouring to manage an infantile case of DIL, by abdominoplasty and liposuction, and presenting the long-term outcome of the procedure.

CASE REPORT

An 11-year-old male patient presented with abnormal contour of the abdomen and both flanks in January 2009 (Figures 1-3). The abnormal contour was first noted by the parents at the age of 5 years and gradually increased in size without any additional manifestations or other similar masses in his body. He did not have any other medical abnormalities, had not had any surgeries prior to this, and there was no familial history of obesity nor abdominal tumors. There was no abnormal prenatal nor neonatal history. There was no history of any abdominal trauma, drug administration, or hospitalization. Previous referral to pediatric endocrinology and genetic counseling showed

From the Plastic and Reconstructive Surgery Department, Al Azhar University, Cairo, Egypt.

Corresponding Author:
Dr Amr Elbatawy, Plastic and Reconstructive Surgery Department, Al Azhar University, 74a Belgrave Court, Leeds, UK.
E-mail: amrelbatawy@azhar.edu.eg; Twitter: @dr_3mora

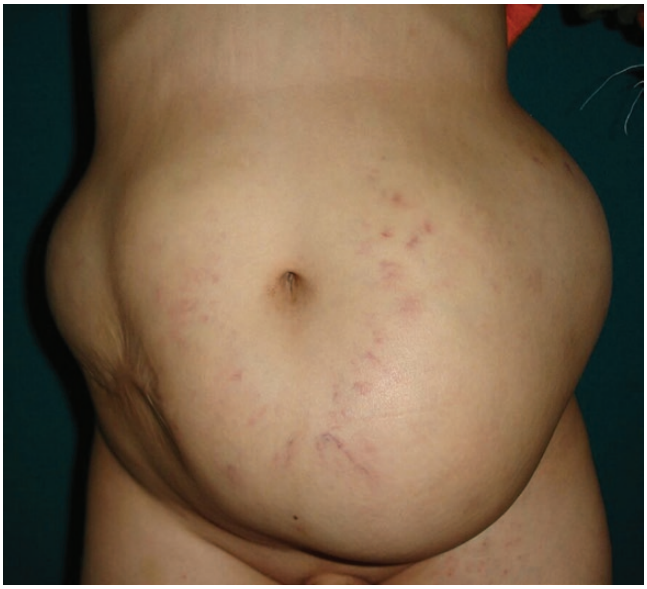

Figure 1. Preoperative frontal view of the 11-year-old male patient. Masses are noted in the abdomen, both flanks, and upper thighs.

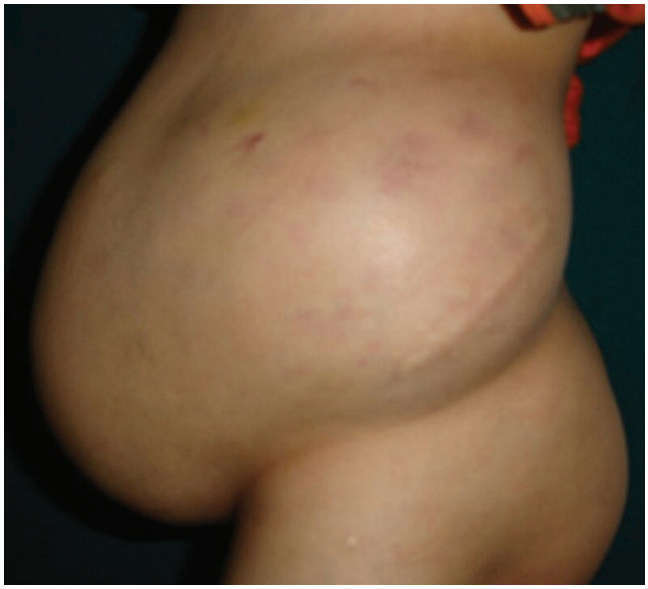

Figure 2. Preoperative views of the 11-year-old male patient, A left lateral view. A scar is located on the left Iliac fossa from the previous biopsy.

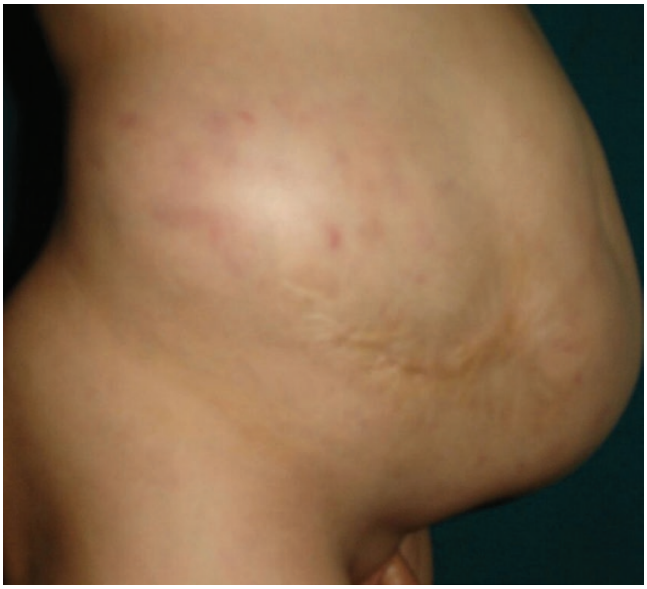

Figure 3. Preoperative views of the 11-year-old male patient, A right lateral view. A scar is located on the right Iliac fossa from the previous biopsy.

normal hormonal profile and no abnormality detected in genetic studies.

Further clinical examination revealed multiple masses of the anterior abdominal wall, both flanks, and upper thighs. The ill-defined masses caused a generalized obesity in the trunk superficial to the muscles. They were not related to the skin, nor did they have any pigmentation or ulceration. There was no lymph node enlargement nor skin sagging. To determine the extent of the masses, a computed tomography (CT) scan was performed. It showed diffuse subcutaneous fatty infiltrations to the anterior abdominal wall, both loins and upper thighs with no bony deformities of the pelvis. According to the CT findings, the masses had infiltrated the subcutaneous tissue with no muscular, intraperitoneal, or retroperitoneal extension and this was confirmed by ultrasound. An incisional biopsy was done preoperatively and revealed subcutaneous lipomatosis with no malignancy. The weight of the boy was 44 kg (80th centile) according to his age.

Considering the patient's age, the extent of involvement, absence of predisposing factors, and normal laboratory examination, he was diagnosed with diffuse abdominal lipomatosis. He did, however, feel weighted down by his bulky trunk and complained of isolation from his peers and difficulties in routine daily activities. After a discussion with the family, surgery was planned for him. We considered an excisional surgery for all the diffuse abdominal lipomatosis with an abdominoplasty Approach.

Operative Procedure

The operation was done in 2 stages; first stage: under general anesthesia, tumescent solution was injected, and a full abdominoplasty incision was made. Dissection of the abdominal flap was performed up until the costal margin with release of the umbilicus. A lobulated mass was found with an incomplete capsule superficial to the rectus sheath

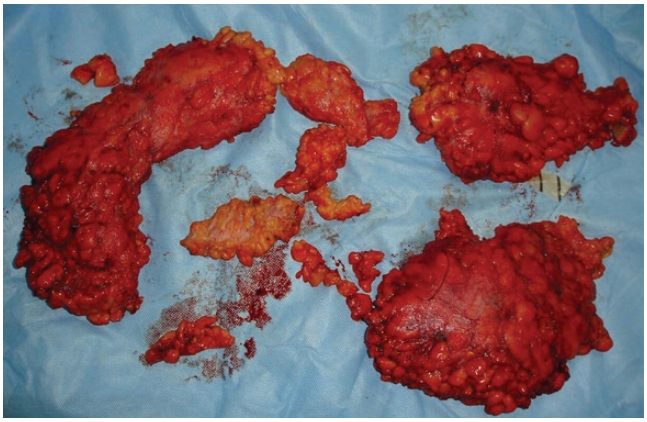

Figure 4. Lobulated masses were found with an incomplete capsule excised completely.

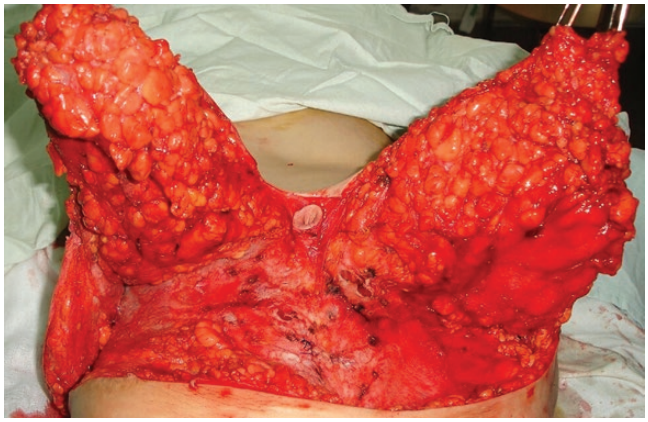

Figure 5. Dissection of the abdominal flap till the costal margin with releasing of the umbilicus.

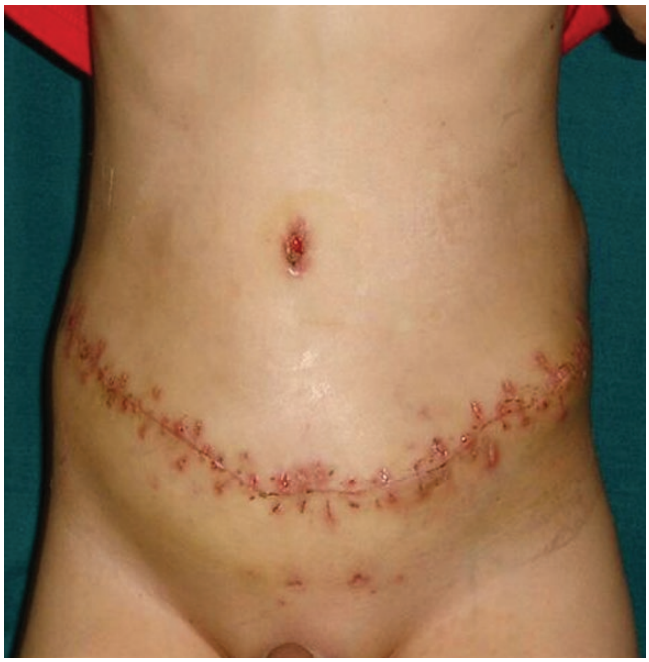

Figure 6. Postoperative frontal view of the 11-year-old male patient. Most of the masses have been removed, and the result shows an appropriate trunk contour.

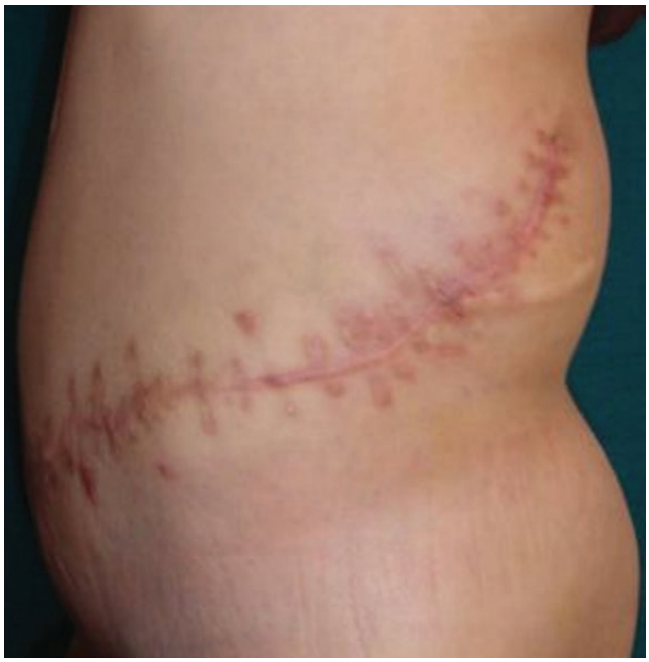

Figure 7. Postoperative lateral view of the 11-year-old male patient, scar shows hypertrophic criteria.

(Figure 4). Removal of most of the anterior and flanks lipomatosis was performed, and followed by abdominoplasty, umbilicoplasty, hemostasis, and wound closure in 2 layers (Figure 5). The total size of the resected masses was 3500 gm, approximately 8% of the patient's total body weight. Pathological evaluation revealed fibrotic dermis overlying diffuse lesions of variable-sized lipomatosis with no atypia or malignancies. The recovery was uneventful in the postoperative period and suture removal was performed after 10 days (Figures 6 and 7).

Three years after the initial surgery, the patient showed no local recurrence. Due to residual masses in the flank area and upper thighs, we planned liposuction on both flanks and thighs with abdominal scar revision. An additional 1300 mL was aspirated in the liposuction and 425 gm excised with the scar revision.

At the 36-month follow-up, the patient showed excellent results, with restored symmetry to the abdomen, flanks, and thighs (Figures 8 and 9). There was no permanent hypesthesia. No areas of growth were evident within

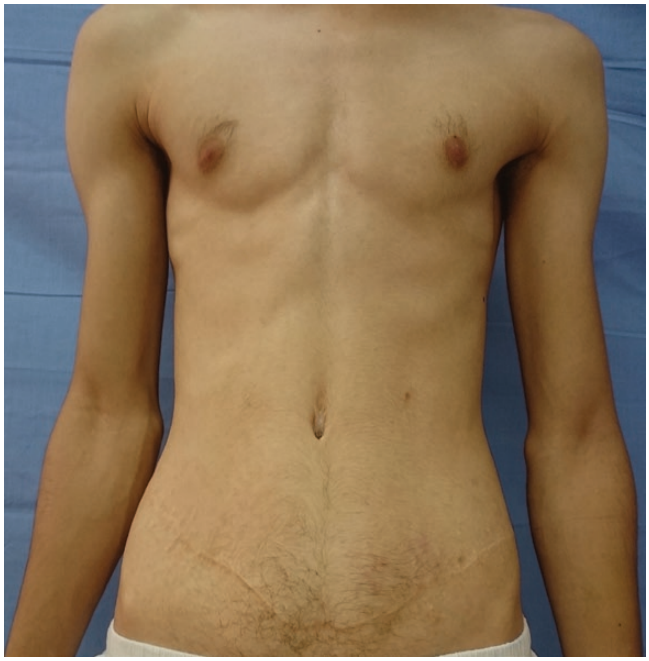

Figure 8. After 3 years, following liposuction and scar revision, the 14-year-old male patient presented with excellent contour and perfect scar.

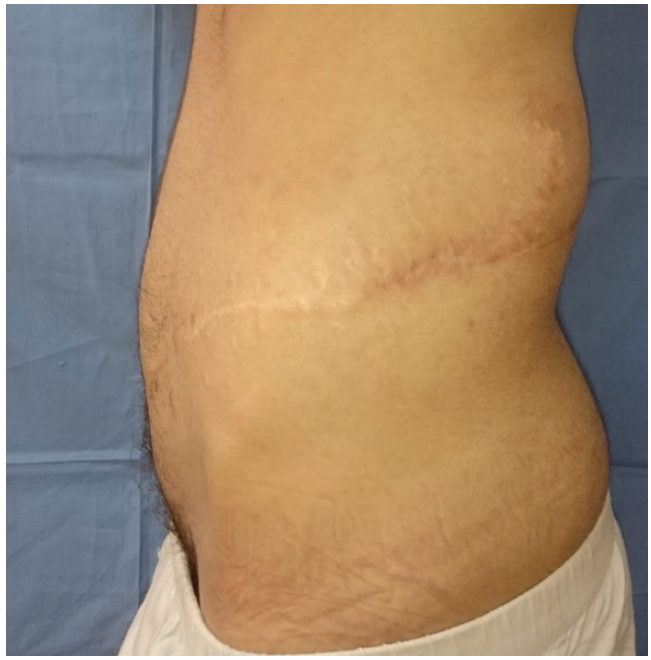

Figure 9. Late postoperative view after the second stage.

and along the margins of resection. The patient started to practice sports within a group and the family was very satisfied. Satisfaction was assessed by our clinical psychologist who interviewed the patient and his parents before and after the procedure especially with the change in the social habits of the patient, being more interested in socializing with his peers.

DISCUSSION

DIL denotes a condition notable for the variable infiltration of benign adipose tissue into the skin, subcutaneous tissues, and muscles in a multitude of anatomic sites.[4-6] DIL describes the overgrowth of mature adipose tissue with a lack of encapsulation. This entity is composed of mature adipocytes, which cannot be histologically differentiated from normal fat.[7] The etiology of the lipomatosis is unknown. A defect in the catecholamine-induced lipolysis has been demonstrated in these patients. They often have reduced glucose tolerance and peripheral insulin resistance. Recent findings show a mitochondrial defect, which may be inherited or due to the toxic effects of alcohol. DIL can be either congenital or acquired and may occur sporadically or in families.[8] Congenital DIL occurs in infancy or early childhood and can lead to prominent aesthetic deformity and functional impairment.[4] Childhood obesity is associated with lower levels of self-esteem, which is reflected by significantly higher rates of sadness, loneliness, and nervousness and are more likely to engage obese teens in high-risk behaviors such as smoking or consuming alcohol.[9] The acquired type affects mostly white men between 25 and 60 years of age. It is usually associated with dyslipidemia (hypertriglyceridemia and paradoxically elevated high-density lipoprotein), impaired glucose tolerance, hyperuricemia, macrocytic anemia, and peripheral neuropathy.[10,11] Lipomatosis has been described in the literature and classified as symmetrical and asymmetrical.[12-15] Several systems of classification were published according to the anatomical distribution of lipomatosis.[16,17] The recent system of classification by Schiltz et al[18] divides the diffuse lipomatosis into the upper part, lower part, and generalized. DIL of the abdomen and lower extremity should be distinguished from simple obesity and other disorders that cause deforming swellings such as neurofibromatosis, vascular malformation, or tumors (ie, simple lipoma, lipogenic liposarcoma, lipoblastomatosis, myxoma, and fibroma). A complete history and physical examination with magnetic resonance imaging or CT make its diagnosis possible.[4,19]

The treatment of DIL is challenging. DIL neither responds to caloric limitation nor exercise or lifestyle changes.[14] Liposuction and lipectomies are versatile options to treat children with diffuse congenital abdominal lipomatosis.[20-24] The lack of a true capsule and increased vascular and fibrous components of the infiltrating lipomatous tissue make surgical management more difficult to completely excise the tumor, and patients need to be aware of the high propensity of recurrence and several surgical treatments.[25]

Abdominoplasty can be selected in cases of diffuse lipomatosis infiltrating the abdominal wall, intraperitoneum, and retro-peritoneum.[12] Abdominoplasty is not a common procedure in pediatric patients. It is usually a treatment for congenital anomalies such as Prune belly syndrome,[26-29] Cantrell's syndrome,[30] or in the reconstruction of patients with burn.[31] Maguina et al[31] reported mini-abdominoplasty to harvest skin for burn reconstruction in 2 young cases, at approximately 13 months of age. Fallat et al[32] described a comprehensive approach in the management of the prune belly syndrome including abdominoplasty, preferring abdominoplasty at 12 months old. Moura et al[33] reported correction of abdominal deformity in a 14-year-old female patient with bladder exstrophy by classic abdominoplasty.

CONCLUSIONS

DIL is one of the causes of obesity in young people. DIL may be syndromic or non-syndromic and symmetrical or asymmetrical. Multi-disciplinary team management is a crucial factor in treatment and includes the plastic surgeon, pediatric endocrinologist, nutritionist, gene therapist, and radiologist. Liposuction and lipectomies are versatile options to treat children with diffuse congenital abdominal lipomatosis.

Disclosures

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