Multiple Symmetric Lipomatosis: Madelung’s Disease

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Dear Editor,

Multiple symmetric lipomatosis (MSL), known as Madelung's disease, is characterized by the growth of uncapsulated masses of adipose tissue.1 These abnormal diffuse lipomas grow in different areas of the body, generally in the neck, shoulder, and inguinal areas.2 MSL typically occurs around the neck (buffalo humps), parotid region (hamster cheeks), and cervical region (horse collar).3 The problem is almost entirely cosmetic, but functional problems have been occasionally reported, such as tracheal compression4, difficulty in obtaining clothes that fit, and inability to turn the head from side to side.5

A 52-year-old woman presented to our department complaining of severe neck and back pain and presented with an 11-year history of progressively growing fatty masses in the neck, shoulder, chest, and back regions (Figures 1–3). She had undergone 12 previous surgical operations for the correction of this deformity. Liposuction alone had been performed seven times, and dermolipectomy had been performed five times in the neck area; however, the outcome was only minimal compression relief and repeated relapse of progressive growth. Initial biopsies of the adipose mass confirmed the presence of adipose tissue without malignant transformation. The patient is a nonsmoker and has no alcohol intake history. There was no family history of a similar disorder as well. Physical examination showed multiple giant soft tissue masses involving the neck, shoulders, and back. There were transverse cervical scars observed in the neck area.

Routine preoperative laboratory tests were normal. Superwet conventional liposuction procedure was performed to the five distinct areas on the back under general anesthesia. A total of 3000 mL was aspirated from these areas. After liposuction, dermolipectomy was performed to remove the fat deposits of the right subscapular region (12×6.5 cm) to excise the redundant skin. A closed suction drain was placed. A compression bandage was applied to the operative area that dissipates the dead space. The drain was removed one day after the surgery. The patient was discharged from the hospital on post-operative day 1. In the early post-operative period, there was a serous fluid leakage from the cannula entry point, but this problem was resolved spontaneously in the following four days. Histological examination revealed normal lipomatous tissue.

During the follow-up period, the patient was satisfied with the cosmetic result and the relief from neck and back pain (Figures 4, 5). The patient is in the 1.5th postoperative month, with only minimal complaints of neck and back pain, but the cosmetic problems still persist.

Multiple symmetric lipomatosis is an extremely rare disease, characterized by diffuse, painless, uncapsulated, symmetrical accumulation of fat tissues.6

Figure 1. Typical aspect of the patient with multiple symmetric lipomatosis.
The current treatment alternatives of MSL are generally unsatisfactory. There is no effective medical treatment. Some authors recommend using a beta-2 agonist and cessation of alcohol intake. Surgical treatment is frequently followed by recurrence; nevertheless, it can yield satisfactory functional and cosmetic results. Liposuction is a surgical treatment alternative for reducing the masses. Although liposuction causes less scarring, is less invasive, and has a shorter recovery period, surgical excision is the most effective treatment in patients with larger masses or with severe cosmetic deformities and compression cases. In our patient, liposuction was planned to the five distinct areas on the back where the masses were extremely diffuse, rendering the removal by dermolipectomy difficult. We consider liposuction as the first step, which is combined with lipectomy, and this approach allows for the histological examination of the dystrophic fat with a safer disease management. Our patient

Figure 2. Preoperative appearance (lateral view)

Figure 3. Preoperative appearance (posterior view)

Figure 4. Postoperative appearance (6 weeks) (lateral view)

Figure 5. Postoperative appearance (6 weeks) (posterior view)
did not complain of the outcome of the previous surgical operations, although there had been delayed wound healing and prolonged hospitalization. In these patients, alcohol withdrawal and weight loss should be recommended, although this may not prevent the progression of the course of the disease.8

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