Benign symmetrical lipomatosis is a rare metabolic disorder characterized by the presence of multiple, symmetric, nonencapsulated fat masses in the face, neck, and shoulders. The clinical course is slow, typically one of slow progressive enlargement with cosmetic and functional sequelae. The authors describe a case in which an open surgical approach was performed to treat this disorder, with good results. There are many aspects of treatment currently lacking a consensus, and the authors discuss these, principally in relation to the location of the fat, the role of liposuction versus surgery, the staging of surgical procedures, and the placement of the incisions.

**Keywords**
rhytidectomy, Madelung disease, facial surgery

Accepted for publication January 30, 2011.

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**CASE REPORT**

A 34-year-old man presented with neck swelling that had persisted after liposuction on three occasions at another hospital. Initial onset of swelling was three years before presentation. The patient’s father had suffered from similar, although less severe, symptoms. The patient’s physical examination revealed fatty swelling around the full circumference of the neck, with the most obvious collections being over the parotid glands and the lower part of the masseter muscles. There was a large posterior cervical fat collection over the cervicothoracic junction, in continuity with lateral masses, along with a separate anterior fatty swelling deep and superficial to the platysma. The patient also showed evidence of some interesting additional features that have not been described in the literature.
Figure 1. (A, C) This 34-year-old man presented with diffuse benign lipomatosis in the face, neck, and shoulders. He had previously undergone three rounds of unsuccessful treatment with liposuction. His physical examination showed fatty swellings around the full circumference of the neck, with the most obvious collections being over the parotid glands and lower part of the masseter muscles. He also had a large posterior cervical fat collection over the cervicothoracic junction (which was in continuity with the lateral masses) and separate anterior fatty swelling, deep and superficial to platysma. He had an indented appearance of the temporal fossae, downward-sloping eyes, and slightly hypoplastic maxillae. (B, D) 18 months after open surgical treatment of the patient’s Madelung disease. The cosmetic results were pleasing to both the patient and the surgeon, and the resultant scars were well hidden.
including an indented appearance of the temporal fossae, downward-sloping eyes, and slightly hypoplastic maxillae (Figure 1).

**SURGICAL TECHNIQUE**

This patient’s surgical treatment was undertaken in three stages. The reduction of neck fat was completed via conventional facelift incisions and supraplatysmal plane dissection. For this step, skin excisions were placed in the usual facelift position, except that the mastoid incisions were placed on the hairline rather than in it, such that they would meet the skin excisions from the posterior neck. In a second stage, the reduction of posterior and posterolateral fat was performed via a posterior midline vertical skin excision—again, in continuity with the excision below the hairline so that it would link with the facelift incision (Figure 2). Last, submental fat excision was then carried out deep and superficial to the platysma.

Through an open approach, it was possible to safely carry out a thorough fat removal. There was good control of the bleeding, avoidance of damage to the facial nerves and their mandibular branches, and easy removal of surplus skin. The patient’s postoperative course was uncomplicated, and he was highly satisfied with the aesthetic improvement (Figure 1). In fact, he spontaneously commented on greater freedom in terms of head mobility.

**DISCUSSION**

**Differential Diagnosis**

Several publications have discussed the differential diagnosis of Madelung deformity and drawn attention to possible associated conditions, principally of an alcohol-related, metabolic, or endocrine nature.³,⁶ In terms of impact on treatment, the only consideration appears to be that absolute abstinence from alcohol may prevent progression in the size of the fatty masses, but this lifestyle change will not induce regression.³,⁶,⁷ At present, the biochemical cause of the condition remains unknown; therefore, the mainstay of treatment is surgery.¹,³,⁶,⁸

**Liposuction Versus Open Surgery**

Before the advent of liposuction, open surgical excision was the only form of treatment for aesthetic deformity or relief of laryngotracheal compression in this condition. Over the last 20 years, there have been a few reports on liposuction for Madelung syndrome.⁷,⁹ One of these reports mentions no recurrence of submandibular swelling after two years of follow-up, but long-term results are generally lacking.

Our patient presented after three previous failed attempts at liposuction; his case can reasonably be considered an example of failure with this method of treatment. While it is perhaps an attractive technique to those not well versed in rhytidectomy surgery, it seems unlikely that liposuction would offer anything other than a temporary and limited improvement, probably because the complications that might ensue from extensive, thorough liposuction of the neck are rather alarming and would discourage anything other than partial removal, from which recurrence would then be expected in view of the progressive nature of the condition. By contrast, dissection in a supraplatysmal plane followed by removal of the abnormal fat from the skin provides a safe and controlled near-total removal of affected superficial fat, also allowing the skin to be redraped, which enhances the cosmetic result (Figure 1C and 1D). The resultant scars are well hidden, with the exception of the vertical scar in the posterior midline. In the case reported here, the scar postoperatively has widened to a degree that was disappointing to the surgeon, but the scar was of no concern to the patient, who could not see it.

There has been little published commentary on the subject of combining liposuction with surgical excision, as almost all cases have been treated by a single method. Constantinidis et al⁹ reported on 11 patients undergoing cervical lipectomy, with liposuction performed during a second procedure eight weeks later. The rationale for this was not clear, although associated metabolic disturbances had led to physicians advising against surgery in some cases. The suggestion in the literature⁷,¹⁰ that liposuction be carried out immediately before surgery seems counterintuitive, since the tissues would be discolored and, therefore, the surface of the platysma muscle would be harder to identify. Preliminary liposuction has also been discouraged because of “the risk of poor wound healing and skin necrosis.”¹⁰

In summary, the role of liposuction is limited to patients with smaller pockets of swelling and those judged to be unfit for more major surgical interventions. While objective data on recurrence are unavailable, recurrence after liposuction seems likely, since the disease is progressive in nature.
Location of the Fat

The second issue for debate with this condition concerns the location of the fat. In this case report, the patient’s dissection was supraplatysmal. This approach effectively removed the fat everywhere except in the submental area, where additional removal had to be carried out deep to the platysma. Previous reports have not always specified the location of the fat, but Civelek et al\(^7\) stated that the fat was “almost always located beneath platysma.” Uglesić et al\(^1\) found fat extending “between muscle sheets into the deeper structures of the neck,” but that appears to have been an extreme case, with a total of 3250 g of fat removed over three operations. Other advocates of a surgical approach\(^7\)\(^9\) have located the fat underneath the platysma.

Incision Placement

The third area for discussion concerns the placement of the incisions. This issue goes hand in hand with the controversy over whether removal of coexistent anterolateral and posterior fat in one procedure is appropriate. The relatively sparse commentary in the literature on surgical incision placement may reflect considerable variation in terms of technique, which is usually individualized for each patient, but this lack of consensus is likely troubling to surgeons who may be undertaking this procedure for the first time.\(^1\)\(^0\)\(^1\)\(^2\) Our report makes the case for a three-stage approach on the basis of minimizing trauma to the patient. There is only a short recovery after each procedure, combined with the advantage of not having to turn the patient midway through the procedure.

The rhytidectomy approach with a short submental scar (2.5 cm) provided adequate access to superficial fat and obviated the need for anterior “chin strap” incisions from one earlobe to the other. These incisions can be cosmetically disfiguring and can interfere with the skin redraping required for a satisfactory cervicomental angle. Upjal et al\(^1\)\(^0\) reported using the circumcision approach in two cases and commented that “the surgery was very protracted and considerable loss of blood necessitated transfusion in both cases.” The alcoholic liver disease in their patients probably compounded the complications.\(^3\)\(^6\)\(^1\)\(^0\)\(^1\)\(^2\) While an anterior incision may be necessary where fatty deposits extend downward around the trachea, this sort of extensive circumcision is unlikely to be required in the majority of cases. In the presence of deeper parasaephalgeal masses, a separate, low, thyroidectomy-type skin incision can provide access to the lower part of the neck, with a better cosmetic result than the “circumcision of the neck” (which is likely complicated by postoperative edema of the skin above the scar).

In short, there is no consensus and little commentary regarding the optimum approach to the posterior neck. The upside-down “fleur-de-lis” design (Figure 2) utilized in our patient has not been described, but it worked well in achieving skin tightening. The vertical midline posterior neck scar is a possible problem but is probably acceptable to most patients if the appropriate disclosures are made and informed consent is obtained preoperatively.

CONCLUSIONS

The biochemical basis of benign symmetrical lipomatosis (Madelung disease) remains unknown. Until the cause can be specified, surgery remains the treatment of choice. After an initial surge of reports on treatment with liposuction, the consensus seems to be reverting to an open surgical approach as the preferred technique. Fat location and incision placement are still matters of debate, but a case can be made for multiple-stage surgery (supine and prone) in which a short submental incision combined with slightly modified rhytidectomy incisions affords access to the anterior and lateral neck, followed by a posterior neck incision to optimize skin redraping and address the posterior neck hump.

Disclosures

The authors declared no conflicts of interest with respect to the authorship and publication of this article.

Funding

The authors received no financial support for the research and authorship of this article.

REFERENCES

