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COMBINED SURGICAL LIPECTOMY AND LIPOSUCTION IN THE TREATMENT OF BENIGN SYMMETRICAL LIPOMATOSIS OF THE HEAD AND NECK

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Abstract. Benign symmetrical lipomatosis (Madelung syndrome) is a rare disease of unknown aetiology, which is characterised by diffuse growth of non-encapsulated lipomas. Between 1995 and 2000 we treated 11 patients with benign symmetrical lipomatosis in the head and neck. The group comprised 10 men and one woman aged 34 to 62 years (mean 47). The most common complaints were reduced range of movement of the head and obstruction when eating or speaking. Combined lipectomy and liposuction were done for all patients, with liposuction being done at a second session. The mean follow-up period was 2.7 years. The functional results were satisfactory in all patients. Nine of the 11 patients were also satisfied with the aesthetic outcome. Two patients developed recurrence 1.5 and 2 years after the operation, respectively. There were no serious complications. We think that combined lipectomy and liposuction is a successful procedure for treating benign symmetrical lipomatosis in the head and neck region. Nevertheless, the advantages and drawbacks of the two techniques should be considered preoperatively.

Key words: benign symmetric lipomatosis, Madelung syndrome, lipectomy, liposuction.

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Benign symmetrical lipomatosis is a rare disorder with diffuse formation of new adipose tissue not only in the head and neck, but also in the shoulder region, the arms, and the upper parts of the trunk. The disease was first described in 1846 by Brodie (4). In 1888 Madelung reported 33 patients, including three of his own cases (17). Since then this type of proliferation of adipose tissue has been termed lipomatosis of the neck or Madelung syndrome, though the synonyms adenolipomatosis, diffuse or multiple symmetrical lipomatosis, lipoma anullare colli have been used, as well as Launois-Bensaude syndrome (15, 19).

The disease is a benign disorder occurring predominantly in middle-aged men (5, 8, 13), and the incidence seems to be the highest in the Mediterranean regions (1). Enzi (7) reported an incidence of 1:25000 in Italian men.

The disease progresses over a period of years and the fat deposits do not reverse spontaneously. Because the cause is not known, removal of the lipomatose tissue remains the method of choice. This can be achieved using two techniques: lipectomy and liposuction.

We report here our experience in the surgical treatment of benign symmetrical lipomatosis of the head and neck.

PATIENTS AND METHODS

Between November 1995 and October 2000 11 patients with benign symmetrical lipomatosis were treated at the Department of Otorhinolaryngology, Head and Neck Surgery of the Saarland University, Homburg/Saar, Germany, and at the Department of Otorhinolaryngology, Head and Neck Surgery of the University of Erlangen-Nuremberg, Germany. There were 10 men and one woman aged 34 to 62 years (mean 47).

In all cases a diffuse accumulation of adipose tissue in the cheek area and in the frontal and lateral parts of the neck was noted which differed in extent (Fig. 1). Two patients had additional deformation in the neck and upper extremity. These two patients also had endopharyngeal growth of adipose tissue on the routine computed tomogram (CT) and magnetic resonance images (MRI) (Fig. 2). The most common complaints



Fig. 1. A 34-year-old patient with pronounced lipomatosis in the cheek and neck.

were reduced range of movement of the head and obstruction when eating or speaking. All patients felt themselves to be severely embarrassed by their appearance.

Seven of 11 patients were overweight, while four were of ideal weight or were underweight. Further details are given in Table I.

Endoscopic ENT findings were inconclusive except in the two patients with endopharyngeal growth of the lipomas. Any other type of tumour in the head and neck was excluded. In five patients the disease remained unaffected by weight loss of up to 12 kg, and in three patients transient abstinence from alcohol did not lead to regression of the adipose deposits.

All patients were being operated on for the first time, and all were treated by combined cervical lipectomy

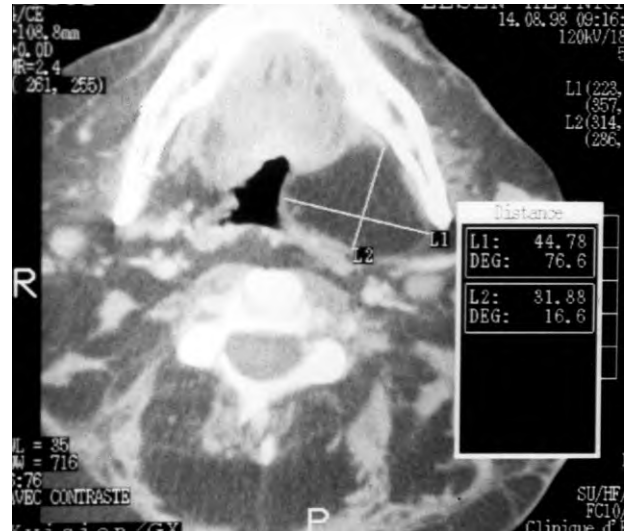


Fig. 2. Deep endopharyngeal growth of adipose tissue. Magnetic resonance scan of the neck (axial slice).

and liposuction. Preparation and preservation of the facial nerve and its caudal branches was necessary in all patients having lipectomy. Preparation was clearly circumscribed and combined with careful haemostasis. Dermectomy became necessary because of excess skin in three cases after lipectomy; in one session up to 2.5 kg of adipose tissue were removed from the neck and parotid region of one of the patients.

Liposuction was done after eight weeks at the earliest at a second session under local anaesthesia and mainly in the submental, submandibular, and posterior cervical region using the tumescent technique (12). An additional second session of liposuction was necessary in four patients. Deformation in the upper extremity was minor and did not disturb the patients. During the medical consultation at the surgical clinic at the time of presentation, patients had been advised not to have an operation.

Table I. Clinical features of 11 patients with benign symmetrical lipomatosis

Case No.	Age (years)	Sex	Duration of disease (years)	Alcohol misuse	Metabolic disturbance
1	45	M	3	Excessive, cirrhosis	Hyperlipidaemia
2	34	M	4	–	Diabetes mellitus
3	48	F	2.5	Excessive, fatty liver	Hyperlipidaemia, hyperuricaemia
4	55	M	5	Heavy	Hyperuricaemia
5	62	M	8	Excessive, fatty liver	Hyperlipidaemia
6	40	M	3	Heavy	Hyperuricaemia
7	44	M	5	–	–
8	43	M	4.5	Excessive	Hyperuricaemia
9	47	M	6	–	Diabetes mellitus
10	48	M	9	Excessive, cirrhosis	Hyperlipidaemia
11	51	M	4	Heavy, cirrhosis	Hyperlipidaemia, hyperuricaemia

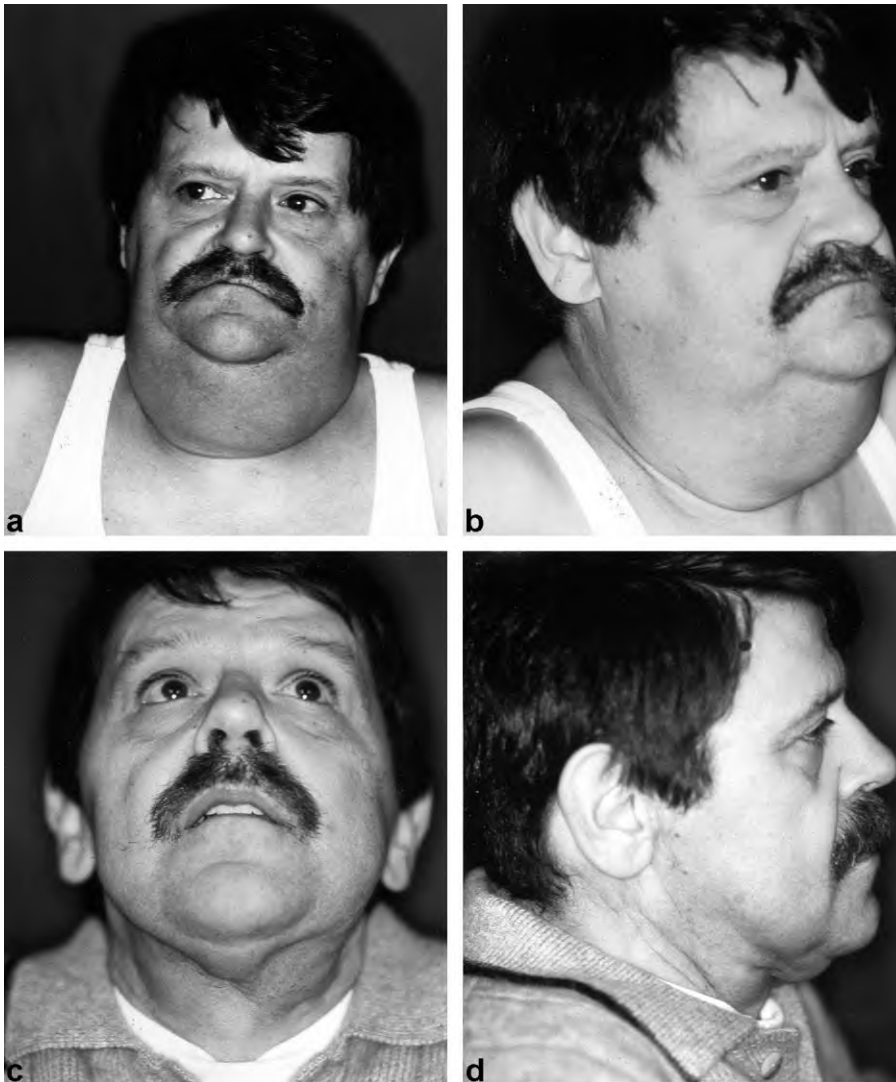


Fig. 3. A 51-year-old patient with benign symmetrical lipomatosis in the head and neck. (*a, b*) Preoperative findings. (*c, d*) Patient two years after lipectomy with skin excision and liposuction. Note the small submental recurrence.

RESULTS

The mean follow-up period was 2.7 years (range three months to 4.8 years). The mean duration of stay in hospital after lipectomy was nine days (range six days to 14 days), and four days (range two days to seven days) after liposuction. All patients were highly satisfied with the functional outcome immediately postoperatively. Mobility of the head had improved distinctly, and obstruction of speech and dysphagia had been noticeably reduced. Nine patients were satisfied as to the aesthetic outcome of treatment, even though small recurrences developed in two of them (Figs. 3–5). Two patients who had had lipectomy followed by liposuction twice expressed only partial satisfaction because of recurrences that occurred 1.5 and 2 years, respectively, after the last operation. They did not want to have further surgery, however.

No severe complications (such as severing of a

motor nerve, skin necrosis or heavy bleeding) occurred. The oral branch of the facial nerve was injured in two patients during lipectomy despite microscopically confirmed preparation. Paresis had largely regressed, however, at seven and 11 months postoperatively.

Two patients developed postoperative haematomas after lipectomy, which were easily treated. One patient with diabetes mellitus developed a wound infection and required continued outpatient treatment over a period of several days after his discharge. There were no complications except a small haematoma and transient paraesthesia in the submental area after liposuction.

DISCUSSION

The aetiology of benign symmetrical lipomatosis is unknown. Increased alcohol consumption is a conspicious

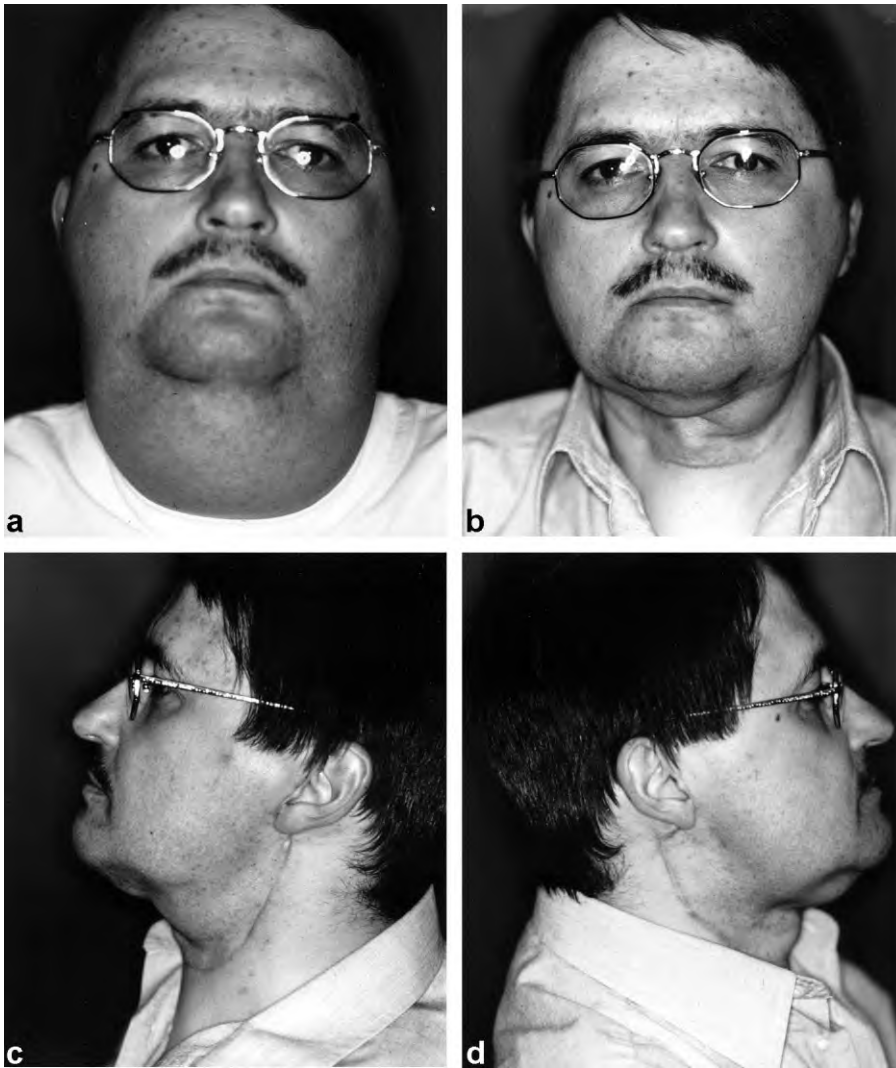


Fig. 4. A 40-year-old man with Madelung syndrome. (a) Preoperative findings. (b, c, d) Pictures taken 2.5 years postoperatively.

uous feature in most patients and is held responsible for the adipose metabolic disorder (9, 23). The lipogenic and antipolytic effect of alcohol can have an important role in the increasing accumulation of fat, but is certainly not its only cause, as the disease also occurs in patients who do not drink alcohol. Hyperuricaemia, diabetes mellitus, hypothyroidism and type I or type IV hyperlipidaemia are also more than usually common in benign symmetrical lipomatosis. A disturbed response of adipocytes caused by catecholamines, which would explain the refractoriness of fat deposits to weight loss (24), as well as mitochondrial dysfunction in brown adipose tissue (3), are being debated as causative factors.

Benign symmetrical lipomatosis is classified into two clinical types (18). In type I the fat deposits are well-demarcated, non-encapsulated, and bulge up symmetrically in the upper parts of the body. In most cases the lipomas develop in the submental and mastoid area and progress upwards and downwards

towards the supraclavicular and neck regions (8). The masses in the neck area cannot be well concealed by clothing, and both aesthetic discomfort and reduced range of movement of the neck are the main complaints that cause the patient to consult a doctor. Less commonly there is extension of the lipoma into the deeper soft parts of the neck and into the mediastinum. In such cases compression of the trachea, or the pharynx, or both cause dyspnoea and dysphagia. The growth of lipomatous foci progresses painlessly and slowly over a period of years, and the consistency of the neck masses can vary (11).

In type II lipomatosis there is diffuse growth of adipose tissue that affects the entire surface of the body, involving the subcutaneous adipose tissue and producing the clinical features of simple obesity. The mediastinum is not affected in such cases (18).

Carlsen and Thomsen (6) described a third type, which is a congenital form of the disease with signs of lipomas around the trunk. The diagnosis is made from

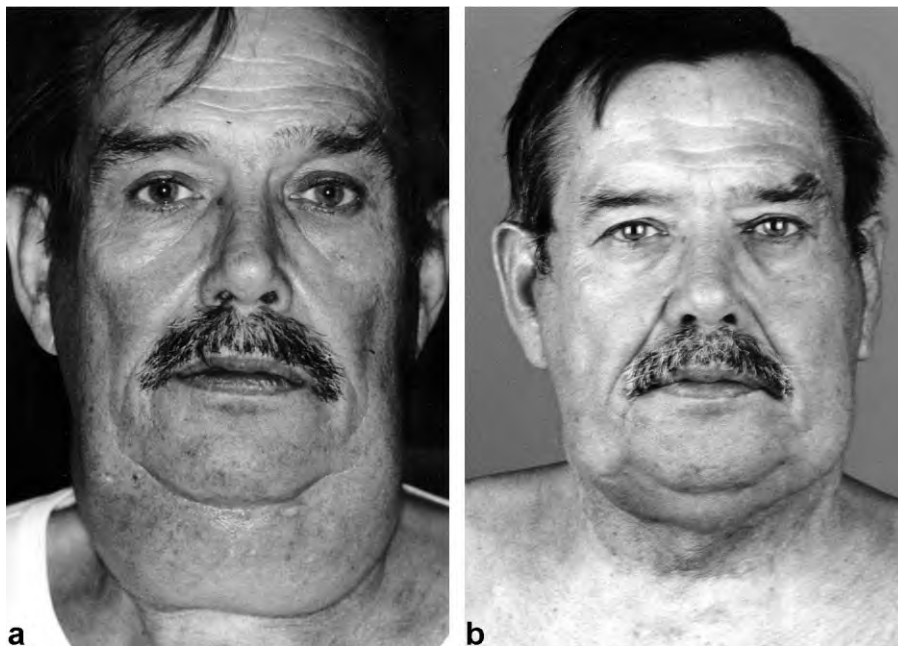


Fig. 5. A 62-year-old patient with Madelung syndrome. (a) Preoperative picture. (b) Patient one year postoperatively. Note the small submental and submandibular recurrence.

the history and clinical examination, and is confirmed by imaging and, if necessary, biopsy. The invariably symmetrical signs on both sides of the body enables a pathognomonic differentiation from other masses. Sonography can be helpful in assessing the growth of adipose tissue, but remains inferior to CT and MRI in calculating the extent and the exact side of the masses relative to other functionally important structures (2).

Histologically, the adipose tissue in benign symmetrical lipomatosis contains relatively large fat cells and is pervaded by septa of vascular connective tissue that have a largely linear course and subdivide the adipose tissue irregularly. In contrast to lipomas, lipomatous tumours are not clearly demarcated from their surroundings and form a kind of pseudocapsule only at certain locations. This “pseudocapsule” is formed by the growth pressure of newly developed adipose tissue, which compresses the surrounding vascular connective tissue or also the perimysium of the surrounding skeletal musculature (11).

Malignant behaviour of the lipomatous tumours can be virtually excluded. Malignant transformation has been reported in only a single case to our knowledge (24).

In contrast, association of the disease with malignant tumours of the upper aerodigestive tract is quite common (19). This may possibly be related to the alcohol and nicotine misuse often found in these patients. A malignant tumour was not detected in any of our patients during the examinations, nor had there been any previous treatment for cancer.

We cannot treat the cause because we do not know what it is. Dietary measures and absolute abstinence

from nicotine and alcohol have no effect on the course of the disease (10, 11, 14, 22). Conservative treatment using the B₂-antagonist salbutamol slowed down the progression of the disease (16), but this treatment is controversial (25).

Removal is the only successful treatment. The two currently established methods of lipectomy (open technique) and liposuction have both advantages and disadvantages. Open lipectomy allows resection of larger amounts of adipose tissue with safe control of the branches of the facial nerve and the vascular nerve sheath. On the other hand, complications such as haematomas, infections, lymphatic fistulas, and the formation of cosmetically-disfiguring scars are common (14).

Intraoperative delineation between pathological fat deposits and the differently developed adipose tissue of the subcutaneous tissue and the cervical triangles is also not possible. This results in incomplete removal of the pathological adipose tissue and leads to recurrence.

Liposuction is a simple technique associated with a low morbidity. With the tumescent technique – which involves infiltration of large amounts of Ringer’s solution together with a vasoconstrictive additive into the area to be suctioned before the liposuction procedure – large amounts of adipose tissue can be removed with only a moderate loss of blood (12). Compared with “dry” liposuction, the tumescent technique ensures less blood and fluid loss, better moulding, and less strain on the circulatory system. By the addition of lignocaine and bicarbonate, liposuction can in many cases be done without full narcosis (12). An insufficiently radical excision in the cheek region

(because of the facial nerve) and problems in patients who have previously been operated on and have hard scars are disadvantages of liposuction (10).

Apart from a few case reports only a few studies involving a larger number of patients treated surgically have been published (10, 11, 14) – though, in an older report by Jaske et al. dating from 1980 (11) lipectomy is recommended as the method of choice, and in a more recent study by Kuipers et al. in 1998 (14) liposuction is preferred as a less traumatising method. Hörl and Biemer (10) reported 18 patients who had a total of 47 operations. (Derma)lipectomy was done 24 times, liposuction 17 times, and combined lipectomy and liposuction six times. A total of 76% of the lipectomies and 72% of the liposuctions were done in the head and neck area, and the remaining interventions in the trunk and the upper extremity.

We recommend wide indications for surgery, even if the operation is desired exclusively for cosmetic reasons, followed by an individualised choice of surgical technique.

In our experience combined lipectomy and liposuction are a successful treatment for benign symmetrical lipomatosis in the head and neck.

On the one hand, extreme findings can be treated adequately by open lipectomy and, on the other hand, liposuction allows better moulding in the case of mainly soft masses of adipose tissue. Masses in the submental, submandibular, and posterior cervical region can particularly be reduced by liposuction during the primary intervention, yielding a cosmetically appealing result.

In contrast, patients with compression of the trachea and pharynx caused by adipose tissue that is giving rise to dyspnoea and dysphagia, cannot be successfully treated by liposuction. The individual anatomical particularities and the consistency of pathological adipose tissue should be considered at the preoperative stage. Unilateral combined lipectomy and liposuction should be avoided because of the risk of poor wound healing and skin necrosis. The course after a surgical intervention can vary considerably. The pathological adipose tissue still remaining can remain unchanged over long periods of time, but can also grow suddenly and fast (20, 21). Although surgical treatment is a symptomatic measure, it can free the patient from possible functional disturbances and can successfully influence the psychological condition of the patient by improving his aesthetic appearance.

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