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Facelift- and circum-occipital incision placement for fat extirpation of the neck in Madelung's disease – A two-case report

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ABSTRACT

Benign symmetric lipomatosis (Madelung's disease) is a rare disorder of fat metabolism that is characterized by progressive symmetrical formation of unencapsulated and painless excess fat masses around the neck and trunk that result in cosmetic disfiguration and functional impairment. Since the disorder is incompletely understood and causal therapy is unavailable, surgical removal of fatty masses is the mainstay of treatment. In this paper the authors describe their use of the classical facelift incision placement as well as a horizontal circum-occipital incision to approach and excise excess fat of the anterior and posterior neck in two patients. This method yielded satisfying results with a combination of good access to fat masses, smooth trimming and redraping of redundant skin, in addition to fairly inconspicuous scarring post-operatively. After removal of 1.5 kg of fat from each patient and a period of uncomplicated wound healing, both patients showed no signs of relapsing fatty growth. Although more challenging than most conventional approaches, the authors' technique has shown good outcomes in those treated with this condition.

1. Introduction

Benign symmetric lipomatosis (also called Madelung's disease or Launois–Bensaude syndrome) is characterized by the progressive formation of unencapsulated and painless excess fat masses around the neck and trunk. Early stages of the disease are usually asymptomatic (Zhang et al., 2008). Further progression (with fat masses at times reaching dramatic dimensions) leads to significant cosmetic disfiguration, compression of neurovascular, tracheal or oesophageal structures and significant limitations of head and neck mobility and flexibility. While the condition's aetiology and pathological mechanism remain largely unclear, symptom-oriented surgical removal of affected areas is the treatment of choice (von Kemp et al., 1986; Kohan et al., 1993). Different techniques and approaches have been reported, which involve longitudinal incisions across the anterior neck and frequently lead to impaired wound healing and noticeable postoperative scars (Gonzalez-Garcia et al., 2004). We present our approach of applying classical rhytidectomy incisions and a posterior horizontal circum-occipital incision to extirpate large ventral and dorsal fat masses in two patients.

1.1. Patients

Two patients presented to our service with progressively growing cervical fatty masses consistent with Madelung's disease (Table 1).

Patient I, a 53-year-old male, presented with severe limitation of head and neck movement, aesthetic disfiguration, a subjective sensation of constant compression of his neck and a recurring headache, which worsened with head movement (Fig. 1). Using MRI (Fig. 2) and ultrasound, fat masses were identified all around his anterior and posterior neck, infiltrating both the subcutaneous and subplatysmal plane. Secondary findings included arterial hypertension, insulin dependent diabetes mellitus, s/p liver cyst removal, chronic alcohol abuse and smoking of 35 pack years.

Patient II, male, 64-year-old, showed severe head and neck movement restriction due to fatty mass compression (Fig. 3). Having had liposuction on two previous occasions, which failed in terms of only minimal compression relief and repeated relapse of progressive growth, he was not pleased with the aesthetic and functional outcome. On MRI and ultrasound imaging, fat masses

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Table 1
Patient data. BMI: Body Mass Index. IDDM: Insulin-dependant diabetes mellitus.

	Patient I	Patient II
Age	53	64
Sex	M	M
Weight (kg)/BMI (kg/m)	85/28.7	74/24.4
Complaints	Impaired head & neck movement (rotation extension and flexion), compression sensation, movement-dependant headache	Impaired head & neck movement in all directions, aesthetic disfiguration, s/p liposuction (2×) with recurrence of fat masses
Comorbidities	IDDM, psoriasis vulgaris, arterial hypertension, s/p cystectomy of liver, chronic alcohol abuse, smoking: 35 pack years	Duodenal ulcer, irritable bowel disease, irritable bladder, moderate regular alcohol consumption

were identified ventrally and dorsally mainly in a subcutaneous but not subplatysmal plane. Secondary findings included a duodenal ulcer, irritable bowel disease, an irritable bladder and moderate but regular alcohol consumption. Upon assessment of family history, the patient's mother was known to have had fatty enlargement of her neck. The patient's brother (57 y/o) presented with small, early stage, progressively growing submental fat masses.

2. Methods

Both patients were assigned to surgical removal of their extended lipomatosis.

In a first step, anterior fat masses were targeted. Access to the ventral and lateral neck was established through incisions corresponding to the super extended facelift technique: temporal to

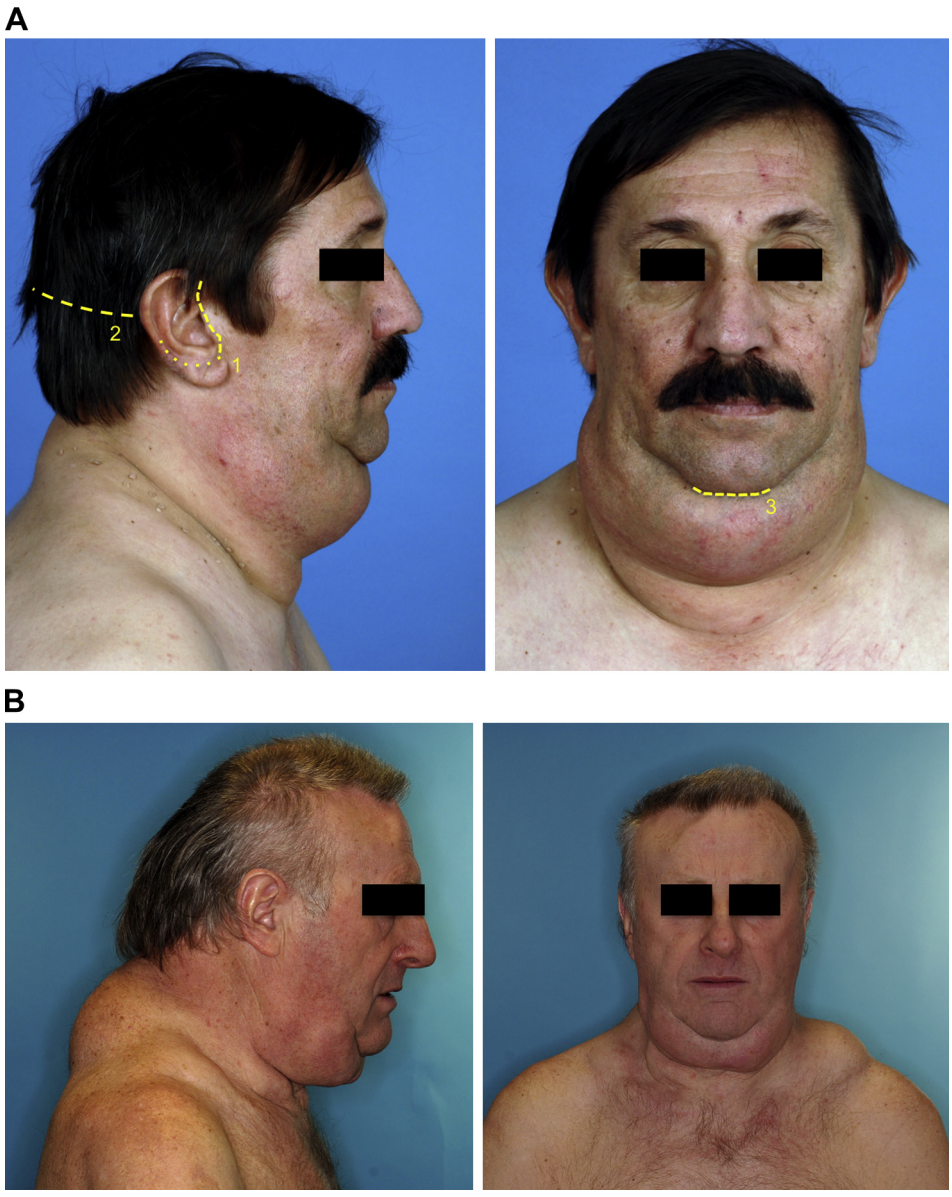

Fig. 1. A: Patient I preoperatively; incision placement. 1: Extended facelift incision. 2: Horizontal occipital incision. 3: Submental incision. B: Patient II preoperatively.