# Madelung's Syndrome – Case Report

**Valdir Frederico Sonni, MD[1]**
**Carlos Eduardo Busch Pires, MD[2]**
**Mari Sugisawa Kay, MD[3]**
**Ivana de Faria Neiva, MD[4]**
**Gabriela Elisete Bier, MD[5]**

1] Associate Member of the Brazilian College of Surgeons. Guest professor of the Plastic Surgery discipline at Pontifícia Universidade Católica do Paraná, Curitiba, Brazil.

2] Senior Member of the Brazilian Society of Plastic Surgery. Chief of the Plastic Surgery Service of Santa Casa de Misericórdia Hospital, Curitiba, Brazil.

3] Senior Member of the Brazilian College of Surgeons. Adjunct Professor of Surgery at Pontifícia Universidade Católica do Paraná, Curitiba, Brazil. Master in surgery by Universidade Federal do Paraná.

4] Gereral surgeon.

5] Clinical specialist.

**Address for correspondence:**

**Valdir Frederico Sonni, MD**

R. Marechal Deodoro, 2908 ap. 02
80050-010 – Curitiba – PR
Brazil

Phone: (55-41) 262-8184

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## *ABSTRACT*

*Madelung's syndrome is a rare type of lipodystrophy afflicting the cervical region, the torso and the region near the upper limbs. The only effective treatment for this disease is surgery.*

*In this paper we describe the case of a patient who has been submitted to periodical surgical procedures at the Plastic Surgery Department of Santa Casa de Misericórdia de Curitiba Hospital since March, 1996. The patient showed noteworthy aesthetical improvement, without recurrence of the lipomatous deposits so far.*

## INTRODUCTION

Madelung's syndrome was first described in 1888 by Otto Madelung. Since then, a little more than 200 cases have been reported on in medical literature.

This condition, of still unknown etiology, is characterized by a symmetrical deposition of adipose tissue along the neck, which causes the classical aspect called "horse

collar" (Otto Madelung[17], 1888), as well as on the parotid, retroauricular and submental regions (hamster cheek), torso, deltoid and supraclavicular regions and in the proximal part of the upper limbs), (pseudoathletic aspect), as well as the posterior cervical region (buffalo hump) (Enzi[8], 1984).

The slow and progressive growth of the lipomatous masses brings about an aesthetical disfigurement, which often causes the patient to isolate himself or to go into depression (Cavalcanti[3], 1995).

## LITERATURE REVIEW

Benign lipomatosis has been divided into 3 (three) clinical groups by Carlsen and Thomnsen (1978) (Table I).

Hugo and Conway (1966) classified Symmetrical Diffuse Lipomatosis (type 2) in two categories:

- predominantly in the torso and the thighs
- predominantly cervical, as described by Madelung.

The lesion originates in the subcutaneous cellular tissue, penetrating into the muscular fascias or in spaces between the organs, apparently following the path of least resistance.

The lipomatous masses are firm and not encapsulated, mixing with the subcutaneous cellular tissue around them, and they are intimately related to the muscles, vessels and nerves. Microscopically, the lipomatous tissue differs from the adipose tissue because it shows more fibrous and vascular elements (Enzi[7,8], 1984).

Growth of lipomatous masses occurs through cellular multiplication similar to a neoplasia and not through hypertrophy of the preexisting cells (Enzi[7], 1977).

It is believed that these patients present a defect in the lipoliytical chain in a phase prior to the formation of intracellular AMPc (Dorigo[5], 1989; Enzi[7], 1977).

Klopstok et al. (1994) reported a common mitochondrial dysfunction in this syndrome. These are associated not inconstant factors: ethylism, tabagism, hyperinsulinemia, hyperlipoproteinemia and hyperuricemia.

Aside from progressive deformity caused by lipomatous tissue, patients don't usually show any symptoms. However, there can be an occurence of mediastinal syndromes deriving from the compression of structures of the mediastinum by the lipomatous tissue, as well as sensorial, motor and autonomical neuropathies (Enzi[8], 1984). The nature of these neuropathies has not been clearly elucidated yet.

Diagnosis of this syndrome can be easily be done by observing the peculiar appearance of the patients; a computerized tomography and magnetic resonance are useful in determining the extent of the masses as well as their relation to the vessels, nerves and muscles.

Differential diagnosis should include the various lesions of the subcutaneous cellular tissue, such as lipomas, angiodysplasias, neurofibromas, sarcomas, goiters, sialoadenitis, obesity and lymphatic tumors.

Several therapeutic methods have been suggested in an attempt to reduce the volume of lipomatous deposits[11, 14, 16, 23, 25, 29].

Leung et al. (1987) reported a case that oral salbutamol reverted a rapidly progressive Madelung's syndrome through an increase in the metabolic rate. The patient, in this case, showed normal lipolytic activities "in vitro" as well as "in viva". However, it is not certain whether lipolytical activitiesremain intact in this disease.

The therapy with salbutamol consists of the ingestion of 12mg/day, divided into three daily doses for six months[14].

Surgical excision, however, is the best treatment (Dorigo[5], 1980; Enzi[8], 1984; Hoehn[9], 1976; Hugo[10] et al., 1966; Schuler[26], 1976; Shugar[27] et al., 1985). Even though surgery comprises only temporary success in improving patient's appearance (once the fat deposits reaccumulate as the metabolic defect remains), the aesthetical and psychological benefits justify the operation.

Excision of the lipomatous masses constitutes a slow surgical procedure, which requires technical skills due to the tissue's extension, fibrous, hypervascular and infiltrative characteristics. Since there is no plan for cleaving the lesions and the normal adipose tissue, total resection of the deposits becomes impracticable. It is thus better to adopt a sculptural technique, in order to restore a relatively normal contouring while removing the largest possible part of the lipomatous tissue (Hugo[10], 1966).

Despite the rigorous hemostasis and the use of post-operative drains, development of seromas and hematomas is rather common (Hugo[10] et al., 1966; Schuler[26], 1976; Shugar[27], 1985).

The use of liposuction in treating this syndrome was described by Carlin and Ratz[1] (1988). They considered this method as an ideal palliative, since it can be carried out under local anesthesia, with smaller cicatricial consequences and in as many phases as necessary. However, relapse of lipomatous masses is probable, just like in the surgical excision procedures. Furthermore, liposuction, according to the authors, presents technical difficulties such as resistance to the cannula penetration (because of the fibrous nature of the tissue), profuse bleeding (because of the tissue's hypervascular nature) and also the need for a conservative approach, because of the close relation of the lipomatous tissue to the vessels and nerves, making it impossible to completely correct the deformity.

## MATERIALS AND METHOD

O.M., male, 53 years old, caucasian, presented no symptoms until four years ago, when he noticed the appearance of masses on the back of his neck (hump) and on his back. Although these masses were painless, they were progressively increasing in volume. At the same time, he noticed a growth of masses on his face, his retroauricular and supraclavicular areas, causing a slight change in his aspect. The patient reported that he had not gained any weight during this period. He was a smoker for forty years (thirty cigarettes per day) and he was used to drinking one dose of alcoholic beverage per day in the last twenty years. No other diseases were reported.

Patient was 1.72 m high and weighed 62 kg. The exam of the segments was normal, except for the presence of firmly consistent masses, with weak adherence to the deep painless planes along the pre, retroauricular, submento and cervical posterior regions, as well as the scapular regions, the lumbar region, the breasts and the scrotum.

For this clinical aspect, Madelung's syndrome was diagnosed and surgical treatment was indicated with a series of resections of the lipomatous masses, in an attempt to give the patient a more harmonious appearance. Table II shows the surgical procedures performed.

## RESULTS

After four operations (as listend in Table II), a significant aesthetical improvement was observed (Figs. 1 - 3).

## DISCUSSION

Although Madelung's syndrome is a rare disease, it is pefectly described by the classical aspects originally reported over one hundred years ago (Madelung[17], 1888), which are the "horse collar", "hamster cheek", "buffalo hump" and "pseudoathletic aspect".

The disease begins in adults with and a previous history of alcoholism and tobaccoism (Enzi[7], 1977; Kodish[11], 1974; Leung[14], 1987; Lyon[16], 1910; Moretti[20] et al., 1973; Tizian[30], 1983). These factors were observed in our patient along with a progressive evolution of the lipomatous masses (Enzi[8], 1984).

Surgical excision was chosen as it is the only effective method of treatment (Dorigo[5], 1980; Enzi[8], 1984; Hoehn[9], 1976; Hugo[10] et al., 1966; Schuler[26], 1976; Shugar[27] et al., 1985).

As referred to by Schuler[26] (1976), the operations were laborious because of the infiltrative, fibrous and

**Table I**

| Benign Lipomatosis | |
|---|---|
| **Type 1** Congenital Diffuse Lipomatosis | Localized mainly in the torso, ill-delimited from the neighboring tissues, non-hereditary. |
| **Type 2** Symmetrical Diffuse Lipomatosis | Develops in adults, primarily localized in the cervical region, hereditary. |
| **Type 3** Multiple Lipomatosis | Numerous small subcutaneous lipomas, encapsulated, localized mainly in the limbs. |

Classification of benign lipomatosis.

**Table II**

| Date | Surgical Procedure |
|---|---|
| March '96 | Posterior cervical lipectomy ("buffalo hump"). |
| May '96 | Bilateral retroauricular lipectomy and also on the suprascapular regions. |
| June '96 | Submento and supraclavicular lipctomies. |
| October '96 | Posterior cervical lipectomy (recurrent "buffalo hump") and on the back (lumbar region). |

Surgical procedures.

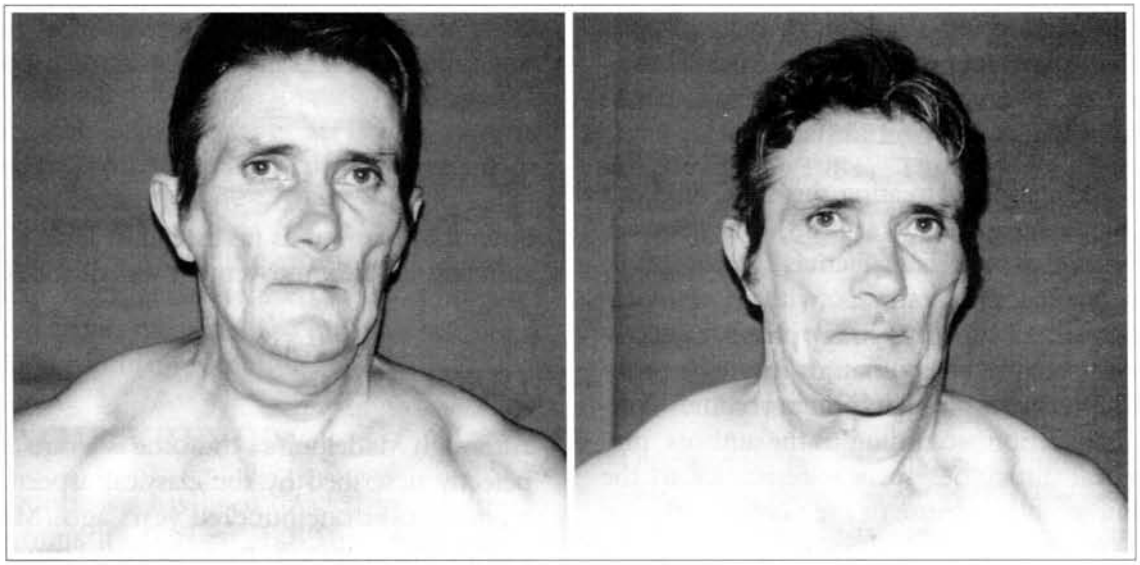

Fig. 1 - Pre and postoperative anterior view.

*Fig. 1 - Pré e pós-operatório (vista anterior).*

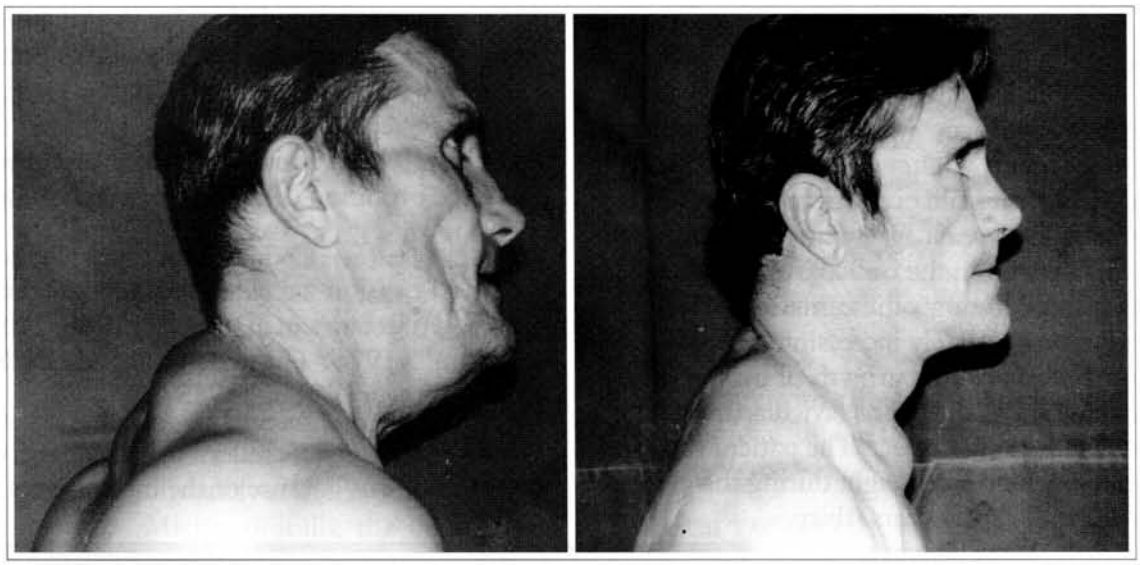

Fig. 2 - Pre and postoperative profile view.

*Fig. 2 - Pré e pós-operatório (vista lateral).*

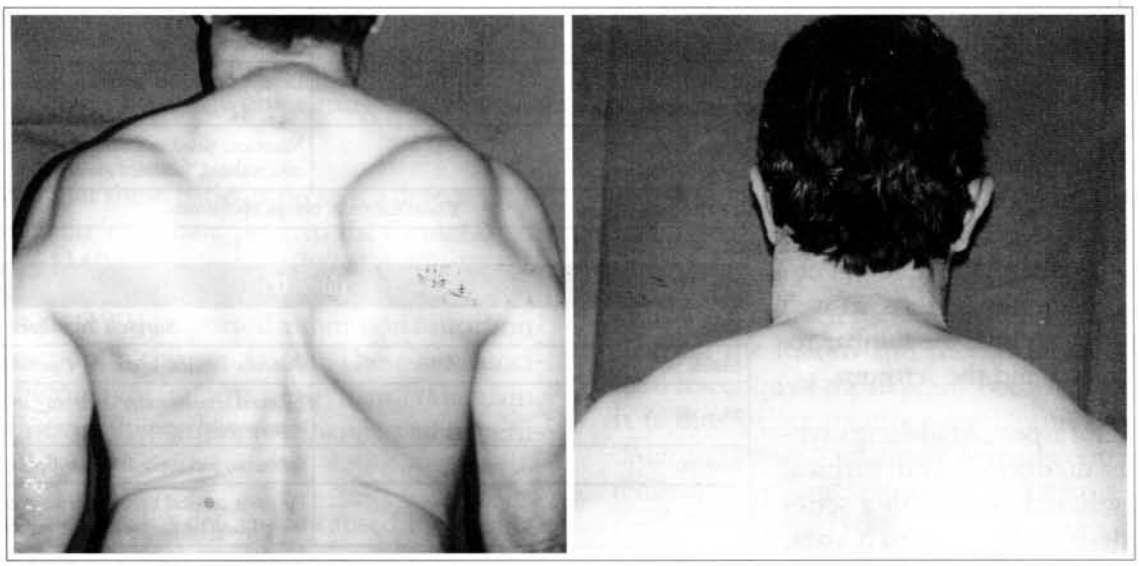

Fig. 3 - Pre and postoperative posterior view.

*Fig. 3 - Pré e pós-operatório (vista posterior).*

hypervascular nature of the lipomatous masses, which showed no plans of cleavage. Even though Penrose drains were used during two days after surgery, all operations formed seromas, requiring aspiration drainage.

The anatomicopathological findings revealed adipose tissue with an increase in fibers and vessels, as reported by Enzi[8], (1984).

Despite the fact that the change in the patient's appearance was subtle, he was extremely satisfied with the results.

## CONCLUSION

The treatment chosen for Benign Symmetrical Lipomatosis is the surgical excision of the tumors. The postoperative result confirms the literature, showing that various interventions in series are necessary, along with patience and skill of the surgeon in order to provide the patient a more harmonious appearance.

Even though the operation entails only temporary success, the aesthetical and psychological benefits obtained justify the operations.

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