

*Prikazi bolesnika/  
Case reports*

**BENIGN SYMMETRIC LIPOMATOSIS -  
MB MADELUNG: *Case report***

**DOBROĆUDNA SIMETRIČNA LIPOMATOZA  
– MB MADELUNG: *Prikaz slučaja***

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*Abstract*

*Key words*

Benign Symmetric Lipomatosis, Morbus  
Madelung, Surgery

*Ključne reči*

dobročudna simetrična lipomatoza,  
Morbus Madelung, hirurgija

Benign symmetric lipomatosis (BSL), or Madelung's disease is a rare illness of local fat distribution and it is rarely reported in the literature. The Madelung disease is presented as a symmetrical deposition of adipose tissue over the head, neck, upper part of the trunk, and proximal part of the upper extremities. The lower part of the body, the trunk and the legs are rarely affected almost not seen. It predominantly affects men between the ages of 30 and 60 years, with a history of alcohol abuse. The disorder can also be associated with hyperlipoproteinemia, hyperuricemia, diabetes mellitus and hypothyroidism. The only effective treatment is surgical removal. A case of bilateral symmetric lipomatosis type I is described, affecting the upper neck region, bilaterally.

*INTRODUCTION*

Bilateral symmetrical lipomatosis (BSL), also known as Madelung's disease or Morbus Madelung, or Launois-Bensaude adenolipomatosis, is a rare benign disease of unknown pathogenesis.<sup>(1,2)</sup>

The disease was first described in the literature by Bordie in 1846. and by Madelung in 1888. Ten years later, 65 more cases were described by Launois-Bensaude, and the disease received his name.<sup>(1,3)</sup>

The male:female ratio of 15:1 up to 30:1 as well as the disease is more commonly observed in white males of Mediterranean origin, aged 30 to 60 years.<sup>(1,2)</sup> Chronic alcoholism is associated in more than 85% of cases,<sup>(1)</sup> in addition to metabolic abnormalities such as hyperuricemia, hyperlipidemia, glucose intolerance, and others such as macrocytic anemia, renal tubular acidosis, and polyneuropathy.<sup>(2,3)</sup>

Clinically it is characterized by the deposition of multiple symmetric masses of noncapsulated fat tissue in the

upper chest and more often neck region, and can be classified into two types.<sup>(3)</sup> Type I presents lipomatous masses in the parotid, cervical, suprascapular, and deltoid regions, and there may be deep involvement. Type II presents diffuse lipomatosis, similar in appearance to common obesity; it is most common morphology in females.<sup>(4)</sup>

*Case Report*

A 59-year-old man came to the University Hospital Insular with a 10-years history of multiple masses mostly on the upper neck, and slighter masses on the upper part of the body.

White (Caucasian) male, without comorbidities, alcoholic for 20 years, presented to this Department with a complaint of bulging in the bilateral upper neck region, painless, which had been progressively growing for the past several years.

At physical examination, he showed good overall status, with bilateral neck mass of soft consistency, painless, meas-

uring 10 × 6 cm to the left and 9 × 7 cm to the right (Fig. 1). The computed tomography (CT) confirmed the presence of a symmetrical upper neck mass, whose density was compatible with fat. The mass was surgically removed (Figs. 2, 3, 4, 5 & 6), and the analysis of the specimen revealed the presence of lipomatosis. Currently, the patient is progressing well without recurrence, and outpatient follow-up has been maintained (Fig.7 & 8).

**DISCUSSION**

Although the etiology of Madelung’s disease is unknown, the present case confirms its strong association with alcoholism. In fact, alcohol intake of more than 80 g per day for at least 10 years, which has been found in up to 90% of cases described in the literature, was also present in our patient. The pathogenesis is believed to be related to the dysfunction in adipocytes and levels of catecholamines responsible for lipolysis.<sup>(3)</sup> Thus alcohol appears to play a role in the adipocyte hyperplasia process <sup>(2)</sup> in genetically susceptible individuals through the pro-lipogenesis, antilipolytic, and decreased lipid oxidation effects. Other studies have also suggested the presence of mitochondrial inheritance through mutation of the maternal gene.<sup>(3,5)</sup>

The diagnosis is achieved by patient history, physical examination, biopsy, and laboratory and imaging tests. Of

utmost importance are complementary tests, in order to discard diseases such as multiple familial lipomatosis, sarcomas, angioliomas, lipoblastomas, neurofibromatosis, Dercum's syndrome, Hanhart syndrome, polydysplasia syndrome, lymphoproliferative disorders, and muscular dystrophies.<sup>(2,3)</sup>

CT/MRI are considered the as best imaging examination for diagnosis, preoperative staging, and postoperative follow-up.<sup>(3)</sup>

The most effective , unique, although palliative treatment is surgical either exeresis or liposuction, with good results, although recurrences are common due to the difficulty of complete tumor excision, as they are not encapsulated.<sup>(2,3)</sup> Some studies have described a clinical treatment with β2-agonist, in order to increase the adrenergic lipolysis and reduce the accumulation of fat, but with questionable efficacy.<sup>(2)</sup> Weight loss and alcohol abstinence appear to have no effect on disease progression, however, these measures associated with surgery can decrease the recurrence rate.<sup>(6)</sup>

The prognosis can be good if if lipomatosis is treated early; morbidity and mortality are more often associated with complications of alcoholism than with the fat deposits directly.<sup>(3)</sup> The lesions never become malignant; however, they may exhibit compression of the larynx and trachea and cause development of neuropathies.<sup>(2)</sup>



**Fig. 1** Preoperative view of bilateral simetric upper neck mass



**Fig. 2** Surgery: incision



**Fig. 3** Intraoperative stage



**Fig. 4** Dislocation of fat mass



**Fig. 5** Postoperative defect



**Fig. 6** Specimen



**Fig. 7** Wound closure



**Fig. 8** Postoperative view

## CONCLUSION

The identification of this rare disease, the appropriate investigation for the exclusion of differential diagnosis, and early surgical approach as seen in the present case, are important to prevent future complications. Regarding the treatment of Madelung's disease, dietary management does not help and abstinence from alcohol may only prevent fur-

ther progression in the size of fat masses. Lipectomy and liposuction are the treatments of choice. Liposuction (although less effective than excision) allows surgery under local anesthesia and avoids the use of general anesthesia in patients with chronic alcoholism and possible liver lesions who are susceptible to hemorrhage.

## Sažetak

Dobročudna simetrična lipomatoza (DSL) ili Madelungova bolest je redak oblik lokalne rasprostranjenosti masti i retko se javlja u literaturi. Madelungova bolest se javlja kao simetrično taloženje masnog tkiva preko srca, vrata, gornjeg dela trupa i proksimalnog dela gornjih ekstremiteta. Donji deo trupa i noge su retko, skoro nikad zahvaćeni. Pretežno pogađa muškarce u dobu od 30 do 60 godina koji preterano konzumiraju alkohol. Ova bolest takođe može da bude u vezi sa hiperlipoproteinemijom, hiperuremijom, dijabetes melitusom i hipotireoidizmom. Jedini efikasan način lečenja je hirurško uklanjanje. Prikazan je slučaj bilateralne simetrične lipomatoze tipa I, gde je zahvaćen region gornjeg dela vrata, bilateralno.

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