Madelung’s disease in a patient with chronic renal insufficiency: a case report and review of literature

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Abstract
Madelung’s disease (benign symmetric lipomatosis, Launois-Bensaude syndrome) was described for the first time in the middle of the 19th century. This disorder concerns mainly men between 30 and 60 years of age. Patients often suffer from coexisting ailments, such as hepatic function disorders, polyneuropathy, diabetes, gynecomastia, hyperuricemia and deviations of lipid management parameters. Treatment of benign symmetric lipomatosis consists mainly in fat tissue sucking or injection lipolysis. Patients often have to wear special clothes correcting the deformed figure. We are presenting a case of a female patient with diagnosed Madelung’s disease, without alcohol abuse. Such abuse is reported in 90% of patients with this disease. Nevertheless, the patient has other typical (and described in the literature) hepatic and pancreatic function impairments, but also glomerular nephritis, which very rarely co-exists with benign symmetric lipomatosis.

Key words: Madelung’s disease, lipomatosis, treatment.

Introduction
Madelung’s disease (benign symmetric lipomatosis, Launois-Bensaude syndrome) was described for the first time in the middle of the 19th century. This disorder concerns mainly men between 30 and 60 years of age. The direct cause of the disease is unknown. Impaired lipid metabolism and inhibition of lipidosis most probably result in adipose tissue proliferation and adipogenesis, mainly around the neck, shoulders and the abdominal area, which leads to considerable deformation of body contours. Due to the dominating fat tissue layout, three types of the disorder have been distinguished: type I – involving the neck (Fetthals), type II – involving shoulders (pseudo-athletic type), and type III – involving the abdomen (gynecoid type) [1].

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Treatment of benign symmetric lipomatosis consists mainly in fat tissue sucking or injection lipolysis. Patients often have to wear special clothes correcting the deformed figure [2, 3]. We present a case of a 54-year-old woman with diagnosed Madelung’s disease and coexisting glomerular nephritis.

Case report
The patient is 54 years old. The first skin lesions, characterized by symmetric adipose tissue proliferation on both arms, appeared 7 years earlier. The changes increased gradually. When the patient was 33 years old, she was diagnosed with common psoriasis. For the last 3 years, she has been a patient of the Nephrology Department due to chronic renal failure, which was diagnosed as a result of pyelonephritis. In 2004, the patient was hospitalised in the Internal and Metabolic Diseases Chair and Clinic, where the diagnosis of benign symmetric lipomatosis was suggested for the first time. The following were also observed: mixed hyperlipidemia, arterial hypertension, hypertrophic cardiomyopathy and chronic circulatory failure. Within 7 years of the disease, the patient gained 22 kg. The medical history of the patient does not reveal alcohol abuse. In 2011, she was admitted to the Dermatology Department of the Medical Univer-
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University of Silesia in Katowice in order to undergo complete diagnostic procedures related to skin lesions.

When admitted, the patient had massive, soft, indolent and symmetric tissue proliferation around both shoulders (pseudo-athletic figure). Physicians also observed adipose tissue proliferation in the abdominal area, around buttocks and thighs. Above elbow joints, on forearms and around the hypogastrium, there were single, confluent lumps of erythematous-exfoliating character. Peripheral lymph nodes were not enlarged. Mucous membranes, nails and the scalp were free of changes (Figures 1–3).

Laboratory tests revealed: erythrocyte sedimentation rate (ESR) 29, γ-glutamyl transpeptidase (GGTP) 118 (normal range up to 24 IU/l), amylase 141 (normal range up to 100 IU/l), and creatinine 118 (normal range up to 96 µmol/l). General urine analysis revealed erythrocyturia and proteinuria. Blood morphology with peripheral blood smear, iron level, electrolytes, aspartate aminotransferase (AST), alanine aminotransferase (ALT), bilirubin, urea, protein, protein electrophoresis, creatine phosphokinase (CPK), aldolase, triglycerides, cholesterol, joint reactions (Latex-R, Waaler-Rose Reaction, ASO) and glucose level were normal. CEA, CA 125 tumour markers fell within the normal range.

A chest X-ray: increased vascular pattern, enlarged left ventricle of the heart, slightly vascular hili. Abdominal ultrasound: an additional spleen, cortical layer of both kidneys with increased echogenicity, kidneys of non-symmetric shape. Ultrasound of soft tissues in shoulders: echogenic lesions reflecting the fat tissue. Nuclear magnetic resonance of the left shoulder: adipose tissue accumulation, fatty focuses within the area of left deltoid attachments. Electromyography (deltoid): the record within the normal range. X-ray of both shoulders: slight densifications of bone structure around the neck of the left humerus and distal part of the right shoulder. Histopathological examination: diffused proliferation of normal fat tissue in subcutaneous tissue with an insignificant increase in the number of vessels. The image reflects lipomatosis. Consultation with a nephrology specialist: the patient with chronic renal failure, stage 3, constantly in the care of the Nephrology Department.

During hospitalization in the Clinic only an external treatment and UVA light therapy were applied on the common psoriasis focuses. The patient is in the care of the Dermatology, Nephrology and Hepatology Departments. The hospital set a date of a surgical consultation in order to establish a surgical fat tissue reduction schedule.

**Discussion**

Benign symmetric lipomatosis is a rare form of an excessive and irregular fat tissue layout. The aetiology remains unclear. However, it is believed that metabolic disorders related to alcohol abuse are as a crucial element of
Masses on both sides of the tongue. A histopathological examination revealed the presence of fat tissue without muscle tissue typical of the tongue.

A 60-year-old female patient described by Zubelewicz-Szkodzińska et al. [15] had fatty deposits around the neck, which considerably limited her mobility. Gdynia et al. [16] described a 55-year-old patient with Madelung’s disease. He developed an axonal neuropathy and hyperlipoproteinemia. The authors suggest the occurrence of decreased COX activity in this patient and disorders within specific regions of mitochondrial tRNA.

Tekin and Ogetman [17] were the first ones to describe a central form of Madelung’s disease, covering the lower part of the body, shoulders and the upper parts of thighs. We observed a quite similar, initial image in our patient. The treatment of benign symmetric lipomatosis consists mainly in fat tissue sucking or injection lipolysis. Hasegawa et al. [18] presented a case of a 42-year-old man with diagnosed benign symmetric lipomatosis. Meso-therapy treatment with phosphatidylcholine injections proved successful in this case. Pharmacological methods still remain controversial.

Zeitler et al. [19] described a 53-year-old male who gained 37 kg within 10 years. The patient had accumulated fatty masses surgically removed. The authors also administered fenofibrates in the dose of 200 mg/day. Fibrates are antagonists of the alpha receptor activating peroxisome proliferation (PRAR-α) and most probably evoke suppression of proteins responsible, among others, for fat tissue architecture [20].

In the case we have described above, the patient had a set of surgical consultations in order to establish a surgical fat tissue reduction schedule.

**Conclusions**

We have presented a case of a female patient with diagnosed Madelung’s disease, without alcohol abuse. Such abuse is reported in 90% of patients with this disease. Nevertheless, the patient has other typical (and described in the literature) hepatic and pancreatic function impairments, but also glomerular nephritis, which very rarely co-exists with benign symmetric lipomatosis.

**Conflict of interest**

*All authors declare no conflict of interest.*
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