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Case Report

Could Madelung Disease be a Cause of Stroke? A Rare Case Report

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Abstract

Multiple symmetric lipomatosis (MLS), also known as Launois-Bensaude syndrome or Madelung disease, is a rare metabolic condition characterized by generally symmetrical, non-encapsulated fat accumulations in the patient's body.

Is there a link between neurological disorders and multiple symmetric lipomatosis, as we demonstrate with our 75-year-old male patient who had an ischemic stroke and was also diagnosed with MLS? Our goal here is to provide an answer to this question.

Keywords

alcohol, lipomatosis, Madelung disease, stroke

CASE REPORT

A 75-year-old male patient was admitted to the emergency department an hour after the sudden onset of speech disorder and loss of strength in his left arm and leg. He had no known previous disease or medication. He had a history of smoking and alcohol use. There was no history of febrile illness or trauma. On physical examination, the patient's general condition was good to moderate, body temperature was normal, and vital signs were stable. On neurological examination, his speech was dysarthric, muscle strength in the left arm and leg was 2/5 hemiparetic according to the Medical Research Council (MRC) scale, his plantar reflex was absent on the left, and the patient's gait was left hemiparetic. The patient was suitable for intravenous thrombolytic (IV tPA) treatment because his National Health Stroke Scale (NIHSS) score was 10, no hemorrhage was detected on brain computed tomography, and his current complaints occurred within the first hour of their onset. Sixty-three mg of IV tPA was administered to the patient. No major artery occlusion was detected on brain-neck CT angiography. The patient was admitted to the neurology intensive care unit. In the patient's repeated examinations, at the end of the 24th hour, the left upper and lower extremities were 5/5 according to the MRC scale, but dysarthria continued.

In the patient's brain magnetic resonance imaging (MRI), there was a diffusion-restricting area in the cortical-subcortical area of the right frontal lobe, extending to the lateral precentral gyrus ***(Fig. 1)****.*

The patient's treatment was arranged as acetylsalicylic acid 300 mg daily, clopidogrel 75 mg and lansoprazole 40 mg daily.

During the physical examination of the patient, rhinophyma and numerous symmetrical swellings were detected in his arms and legs. The same swelling was also present in his right breast and resembled gynecomastia ***(Fig. 2)****.*

Routine blood tests, biochemistry, whole blood, vitamin B12 tests, erythrocyte sedimentation rate, lipid metabolism, thyroid function tests, HbA1c, serum electrophoresis, autoantibody screening (antinuclear antibody, anti-SSA, anti-SSB), antithyroid antibodies, syphilis serology (fluorescent treponemal antibody), and Schirmer test were all normal. ELISA tests and Behçet's pathergy test were negative. Complete urinalysis was within normal limits.

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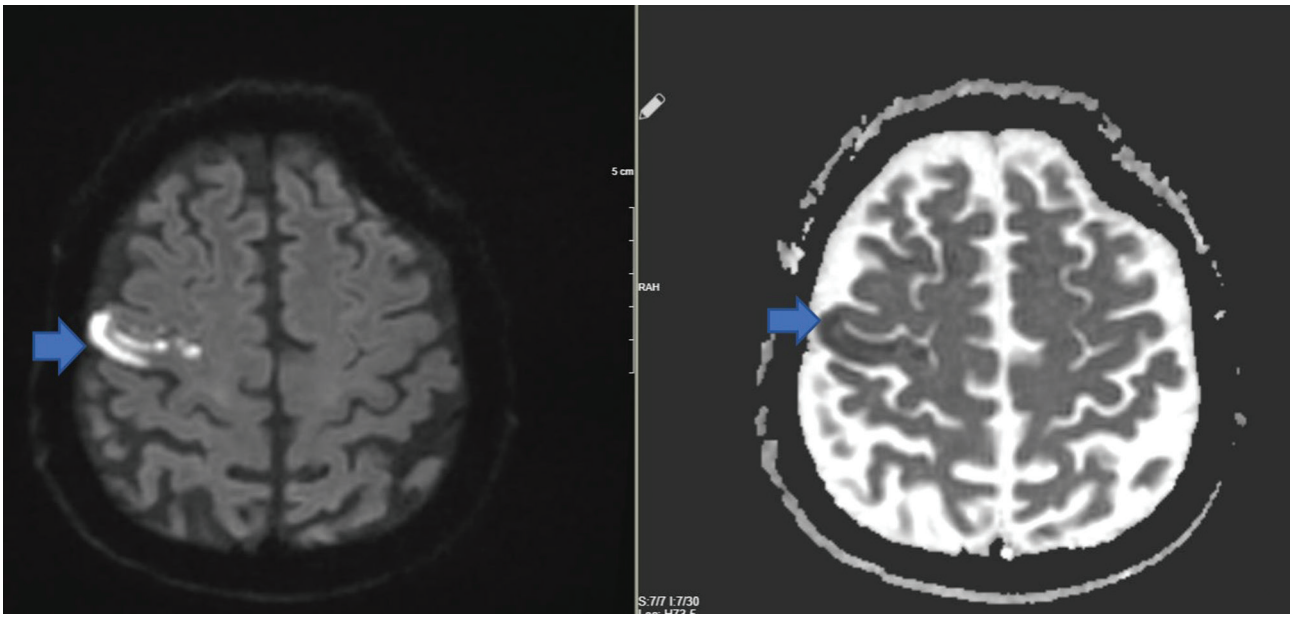

Figure 1. Acute diffusion restriction extending to the lateral side of the precentral gyrus of the right frontal lobe.

Figure 2. Multiple lipomatous lesions on the patient's right chest, arms and legs. It is not found in the head and neck area.

Electrocardiography and echocardiogram were normal. No significant pathology was detected in brain-neck CT angiography.

In the breast and superficial tissue ultrasound (USG), the lesions on his body were evaluated as non-encapsulated lipoma. A biopsy of the lesions was requested, but the patient refused.

The patient was discharged on the eighth day of hospitalization, with minimal dysarthria on neurological examination and a recommendation to continue his current treatment.

DISCUSSION

Multiple symmetric lipomatosis (MSL), also known as Madelung or Launois-Bensaude syndrome, is a rare syndrome with multiple lipomatous lesions, frequently seen in men aged 30-60, and whose etiology has not been fully elucidated. Alcohol use has an important place in the etiopathogenesis of the disease. When we look at the literature, there are cases with genetic transmission. Also, defects in the lipolytic pathway and mitochondrial DNA mutations or deletions have been suggested in response to catecholamines.[1-5]

It has been said that alcohol can affect physiological enzymatic process disorders in the mitochondria, and excess alcohol can disrupt adrenergic lipolysis and cause the appearance of uncontrolled fat deposits in different parts of the body. Cessation of alcohol use may not stop the progression of the disease.[1-5] In addition, our patient had a long history of alcohol consumption.

The condition was first described by Benjamin Brody in 1846, but Otto Madelung in 1888 and Launois and Bensaude in 1898 described another 30 cases of patients with excessive fatty tissue growth around the neck, neck, back and shoulders.[1-5]

We looked at the literature, it can be seen that MSL is classified by Enzi and Donhauser. According to the classification of the Enzi, in type 1, the distribution is symmetrical and is frequently located in the neck, shoulder girdle, back, supraclavicular region and proximal upper extremities, while in type 2, accumulations occur in the abdomen and thigh regions without affecting the upper region.

According to Donhauser, type 1 is the neck, shoulder girdle, back, supraclavicular region and proximals of the upper extremities, type 2 (pseudo-athletic) is the shoulder girdle, deltoid region, chest area and proximals of the upper extremities, type 3 is the lower body, thighs and knees, type 4 is in the form of accumulations in the abdominal area.[2-6]

Considering these classifications, accumulations in our patient were present both in the proximal and distal parts of the upper extremity, in the proximal and distal parts of the lower extremity, and in the right breast. Our patient did not have lipomatous lesions in the neck or back area.

It is well known that mitochondria play an important role in fatty acid production and breakdown in the process related to energy production. The sources in literature had highlighted that brown adipose tissue abnormalities could result from enzymatic mitochondrial defects that could alter the storage and utilization of fatty acids. This can trigger cellular energy deficiency and eventually neuron death. It should also be noted that mitochondrial disorders may be associated with multiple different phenotypes, thus including the development of stroke-like attacks and paralytic ileus, as previously reported.[1,7,8] Although a relationship between mitochondrial disorder and MSL could not be established because biopsy and genetic testing could not be performed in our patient, the hypothesis that it may be a stroke risk factor is also seen in the literature and is intended to be kept in mind in current patients.

Disclosure statement

The authors report no funding or conflicts of interest. The patient provided consent for this case report to be published.

Author contributions

B.T. wrote the first draft. All authors reviewed the article, contributed, and approved its submission.

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Может ли болезнь Маделунга быть причиной инсульта? Редкий случай

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Резюме

Множественный симметричный липоматоз (МСЛ), также известный как синдром Лонуа-Бенсо или болезнь Маделунга, является редким метаболическим заболеванием, характеризующимся в целом симметричными, неинкапсулированными жировыми накоплениями в организме пациента.

Существует ли связь между неврологическими расстройствами и множественным симметричным липоматозом, как мы продемонстрировали на примере нашего 75-летнего пациента-мужчины, у которого был ишемический инсульт и также был диагностирован МСЛ? Нашей целью является дать ответ на этот вопрос.

Ключевые слова

алкоголь, липоматоз, болезнь Маделунга, инсульт