CASE REPORT

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Madelung's Disease Leading to Presenile Dementia in a Non-alcoholic Patient

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ABSTRACT

Introduction: Madelung's disease (MD) is a rare disorder of unknown etiology defined as the presence of multiple and symmetrical fatty accumulations most commonly involving the upper trunk, neck, and head. Excessive alcohol ingestion has been linked traditionally to the pathogenesis of the disease. The central and peripheral nervous system could both be affected. Presenile dementia, without alcohol abuse, has been rarely reported in the literature as a complication. **Aim:** The aim of this case report is to highlight that multiple symmetric lipomatosis can be complicated by presenile dementia even if the patient is non-alcoholic. **Case Report:** This case report describes a middle age non-alcoholic woman who presented for increased forgetfulness. Brain CT scan showed cerebral and cerebellar atrophy inappropriate for her age. Despite being started on anticholinergic drug, her MMSE decreased 3 points in 1 year. **Conclusion:** Clinicians should consider early onset dementia as a potential complication of Madelung's disease even in patients with no preceding history of alcoholism. A brain MRI and MMSE can aid with identifying such a complication.

Keywords: Multiple symmetric lipomatosis; presenile dementia; alcoholism.

1. INTRODUCTION

In 1846, Sir Benjamin Brodie was the first to describe multiple symmetrical lipomatosis (MSL) (1). However, Otto Madelung reported the first series of patients in 1888 (2). Madelung's disease (MD) is defined as the presence of multiple and symmetrical fatty accumulations, usually involving the upper trunk, neck, and head (3). Chronic excessive alcohol use has been linked with accelerated progression of lipomatosis (4). Central and peripheral nervous system involvement have been described, with peripheral neuropathy being the most common neurological complication (5).

Herein, we present the case of a 49-year-old female who presented for increased forgetfulness. Her MMSE indicated mild dementia. Brain CT scan without contrast illustrated a cerebral and cerebellar atrophy inappropriate for the patient's age. The MMSE decreased 3 points in 1 year despite being on treatment.

2. AIM

The aim of this report is to shed light on presenile dementia as a complication of MD even in non-alcoholic patients.

3. CASE REPORT

A 49-year-old female with a past medical history of hypertension, type II diabetes, dystonia, and multiple symmetric lipomatosis (Madelung's disease) was brought to emergency department by her husband for increased forgetfulness for the past 3-month period. She had been having increased difficulty with recalling short term information like the place where she placed a cup. However, she was still able to perform daily life activities. She had no events of being lost outdoors. The patient's past surgical history was significant for left knee replacement and multiple lipoma removals. She was not an alcohol consumer.

She was conscious, cooperative, and oriented to time, place, and person. She was able to recall her date of birth, and named correctly 2 out of 3 items she was supposed to remember. She could not recite the numbers in a serial of 7 starting from 100. The patient had no signs of depression. Her MMSE was 21. She had multiple soft lipomas that were palpated around the neck bilaterally, as well as anagen effluvium. The remaining examination was within normal limits. Her laboratory markers includ-

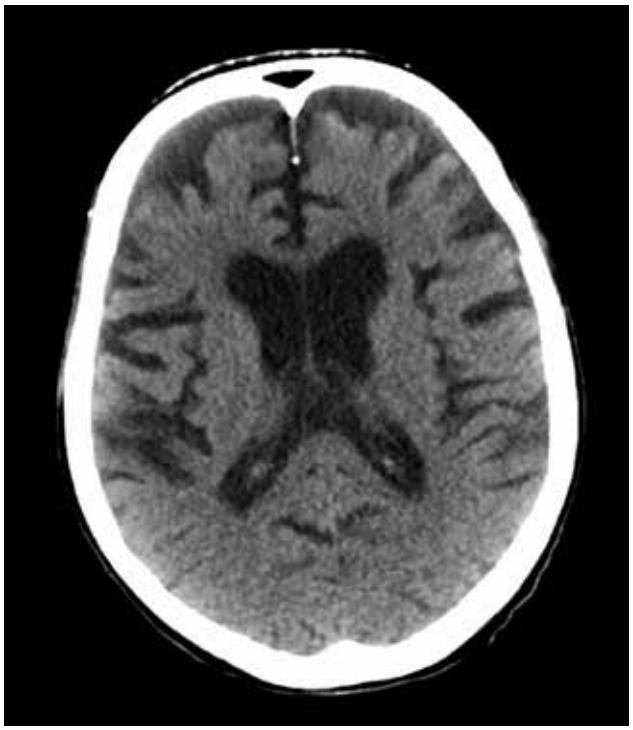

Figure 1. A brain CT without IV contrast showing no evidence of acute intracranial lesion. Dilated ventricles and prominent sulci are evident. Cerebral and cerebellar atrophy inappropriate for the patient's age is revealed.

ing electrolytes, glucose, vitamin B12 levels, and TSH were normal. Urine screening for drugs was negative. A brain CT scan without intravenous (IV) contrast revealed dilated ventricles with prominent sulci. There was a cerebral and cerebellar atrophy inappropriate for the patient's age (Figure 1). The patient refused to perform a brain because of reported anxiety from enclosed spaces. She was discharged on donepezil with follow-up in the neurology clinic.

The patient had several follow-up visits in the neurology clinic. She had no adverse effect of the medication which she was compliant to. However, her MMSE decreased to 19 in a 1 year period.

3. DISCUSSION

MSL has been referred to using multiple eponyms including Madelung's disease (MD), Launois–Bensaude syndrome, and benign symmetric lipomatosis (4). It is characterized by diffuse deposits of fat arranged symmetrically around the neck and shoulder girdle with relative sparing of the lower extremity. The masses are non-encapsulated, and can eventually reach a very large size (6) usually involving the upper trunk, neck and head. Frequently associated findings include diabetes mellitus, hyperlipidemia, liver disease, hypothyroidism and polyneuropathy of unknown origin, but nevertheless, there are published reports of cognitive disorders in patients with MSL. We describe two unusual cases (38-year-old and 45-year-old Greek men. The incidence is reported to be 1:25,000 (7).

MD is commonly associated with men of Mediterranean descent in the third to sixth decade of life who have a history of chronic alcohol use (8). Heavy chronic alcoholism has been described in around 60% to 90% of patients (9). This major risk factor is inconsistent with our patient who has a negative social history of alcohol use.

Many reports in the literature have described the neurological involvement of MD which can be central or peripheral. Neuropathy is the most frequent reported complication (5). MD patients may also have ataxia, pyramidal signs, hearing loss, myopathy, and abnormal visual or somatosensory evoked potentials (5). It has been suggested that since MD occurs with alcoholism, the neurological manifestations can be complications of alcohol ingestion. Naumann and colleagues showed a lack of correlation of conduction velocities with alcohol intake, the presence of neuropathy in abstinent MSL patients, and sural nerve pathology, which tends to be surprisingly normal compared to the axonal degeneration and demyelination typically seen in alcoholic neuropathy (5).

The term early onset dementia refers to dementia that first occurs in a person under age 65. The dementia may be caused by Alzheimer's disease or other diseases and conditions (10). A recent study by Perera and colleagues studied the germline variants in mitochondrial DNA of patients with MD. Out of 10 patients studied, the most consistently associated neurologcial features were numbness and tingling in digits and toes and memory loss in 7 and 4 affected individuals, respectively. A total of 2 patients out of 4 had significant memory loss before the age of 50 without history of alcohol abuse which is similar to our case (11). Triantafyllou and colleagues reported 2 cases of early onset memory loss. However, both patients were moderate to heavy alcohol drinkers unlike our patient (6) usually involving the upper trunk, neck and head. Frequently associated findings include diabetes mellitus, hyperlipidemia, liver disease, hypothyroidism and polyneuropathy of unknown origin, but nevertheless, there are published reports of cognitive disorders in patients with MSL. We describe two unusual cases (38-year-old and 45-year-old Greek men.

4. CONCLUSION

We presented the case of a 49 year-old female with a past medical history of multiple symmetric lipomatosis (or Madelung's disease) who was diagnosed with presenile dementia although she was not an alcohol consumer. Clinicians should consider early onset dementia as a potential complication of MD even in patients with no preceding history of alcoholism.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms.

Author's contribution: Each author gave substantial contribution to the conception or design of the work and in the acquisition, analysis and interpretation of data for the work. Each author had role in drafting the work and revising it critically for important intellectual content. Each gave final approval of the version to be published and they agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy

or integrity of any part of the work are appropriately investigated and resolved.

- **Author contribution**: GEH, RA, SN, and WP gave substantial contributions to the conception or design of the work in acquisition, analysis, or interpretation of data for the work. EOL, JVG, and AK had a part in article preparing for drafting or revising it critically for important intellectual content, and all authors gave final approval of the version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
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