Bulletin of Environment, Pharmacology and Life Sciences
Bull. Env. Pharmacol. Life Sci., Vol 12 [12] November 2023 : 437-440

Online ISSN 2277-1808
Journal's URL: http://www.bepls.com
CODEN: BEPLAD

CASE STUDY **OPEN ACCESS**

Madelung's Disease a Rare Disorder with Lipomatous Deposits: A Case Report

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ABSTRACT

This case study explores Madelung's disease, also known as multiple symmetrical lipomatosis (MSL), which is a rare disorder characterized by the abnormal growth of lipomatous deposits in a symmetrical distribution throughout the body. In Ayurveda, the concept of Medoja Granthi is closely related to lipoma. According to Ayurvedic principles, Medoja Granthi is considered to be a localized manifestation of an imbalance in the Kapha dosha and the adipose tissue (Meda dhatu). A 35-year-old male presented with a long-standing history of progressive symmetrical neck swelling over left lateral aspect of neck for 2 years. A preliminary diagnosis of Madelung's disease was suspected based on the clinical presentation and imaging findings. The imaging findings were consistent with the characteristic features of Madelung's disease, confirming the diagnosis. The patient underwent surgical intervention to address the aesthetic and functional concerns associated with the lipomatous mass.The case report focuses on the clinical presentation, diagnostic process, and management strategies employed for a patient diagnosed with Madelung's disease. The study highlights the challenges in diagnosis, the impact on the patient's quality of life, and the importance of a multidisciplinary approach for optimal management.
Keywords*: Madelung's disease, Multiple symmetrical lipomatosis, Granthi*

Received 06.09.2023 Revised 18.10.2023 Accepted 29.11.2023

INTRODUCTION

Madelung's disease, first described by Otto Madelung in 1888, is an exceedingly rare condition characterized by the development of benign, symmetrical, and nonencapsulated lipomatous masses throughout the body.[1,2] Medoja Granthi, a term used in Ayurveda, and lipoma are closely connected. The adipose tissue (Meda dhatu) and the Kapha dosha are both seen to be out of balance, and Medoja Granthi is regarded as a localised manifestation of this imbalance in Ayurvedic theory.[3,4] By Brodie in 1846 and Madelung in 1888, the condition was first identified.[5] There are many terms that can be used to describe MD, such as Launois-Bensaude syndrome, multiple symmetrical lipomatosis, and benign symmetric lipomatosis.[6] Although the exact etiology remains unclear, MD has been associated with alcohol abuse and a genetic predisposition linked to mitochondrial DNA mutations. The reported incidence of MD is 1:25,000, with a male-to-female ratio of lingers between 15:1 to 30:1 and the disorder predominantly affects middle-aged men.[7,8] Ayurveda believes that the accumulation and stagnation of Kapha and Meda mixed with toxins (Ama) in the adipose tissue (Meda) results in formation of Granthi (lipomas). In Ayurveda (Sushruta Samhita), dahan by agnitapta loha shalaka (burning by iron rod) for management of lipomas is advocated. Despite the claims that this method of lipoma management places a lot more emphasis on lipectomy for cosmetic reasons,[9] still Agnikarma has limited scope at sites like neck, face, groin, etc. This case study presents a detailed account of a patient diagnosed with Madelung's disease, highlighting the diagnostic challenges, clinical management, and impact on the patient's quality of life.

Case Presentation

A 35-year-old male presented with a long-standing history of progressive symmetrical neck swelling over lateral aspect of neck since 2 years. The patient had a past medical history significant for alcohol abuse, which had been in remission for the past three years. Physical examination revealed, symmetrical, non-tender fatty masse predominantly located in the lateral cervical and supraclavicular regions. Laboratory investigations, including complete blood count, liver function tests, and lipid profile, were within normal limits. A preliminary diagnosis of Madelung's disease was suspected based on the clinical presentation and imaging findings.

Diagnostic Evaluation Thorough clinical evaluation was carried out to distinguish it from other illnesses such lymphoma, neurofibromatosis, lymphadenopathy, and other lesions originating from muscle, tendon, or vessels in that region. Radiological investigations were conducted to confirm the diagnosis and assess the extent of lipomatous deposits. Ultrasonography (USG) scan revealed the presence of, symmetrical, and confluent fatty mass measuring approx 3×4 cm in left and in right 2×3 cm sized benign lipoma involving the subcutaneous tissue, and supraclavicular areas. The imaging findings were consistent with the characteristic features of Madelung's disease, confirming the diagnosis.

Therapeutic Intervention

A multidisciplinary approach was adopted for the management of Madelung's disease. Patient was referred to a counsellor who advised strict avoidance of alcohol to the patient. As there were no abnormal findings in laboratory investigations of the patient, no other medicine except Tablet Medohara Guggulu (Baidyanath) BID with lukewarm water was prescribed. The patient underwent surgical intervention to address the aesthetic and functional concerns associated with the lipomatous mass. Excision of lipoma at left posterior lateral aspect of neck under local anaesthesia was planned. Approximately 5 cms long incision on swelling at left lateral side of neck was made carefully. After correct identification of important structures, fatty mass between posterior belly of sternocleidomastoid muscle and trapezius muscle with all the extensions was clearly identified. The lipoma was found to be extending deep to anterior margin of trapezius muscle. It was precisely dissected out and completely excised with all the extensions as a single whole mass (Fig. 1). After achieving adequate haemostasis, wound was closed. Patient was discharged without complications after a successful recovery within 7 days.

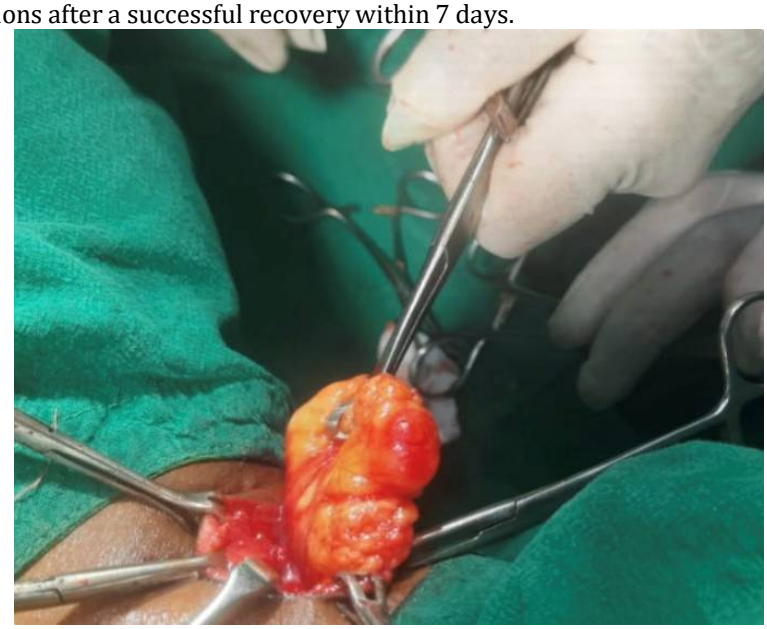

Fig1. Lateral neck excision of lipoma was performed and large fatty mass was completely excised (size: about 3×4 cm).

Follow-up and Outcomes

The patient was called for follow- up after one month of discharge and then after every one month up to six months. During this period no recurrence observed. Patient continued strict avoidance of alcohol consumption.

DISCUSSION

Madelung's disease is a rare disorder that poses diagnostic challenges due to its rarity and diverse clinical presentation.[10] The condition often remains undiagnosed or misdiagnosed for years before appropriate evaluation is initiated. In the literature, MD is categorised in two different ways. Using anatomical fat distribution as a basis, Enzi divided MD into two categories.[11] In type 1, the distribution of fat is symmetrical and concentrated in the neck, shoulders, supraclavicular triangle, and proximal upper limbs; in type 2, however, these areas are unaffected and deposits form in the abdomen and thighs, making this type similar to typical obesity. Imaging modalities such as USG and MRI play a crucial role in confirming the diagnosis and assessing the extent of lipomatous deposits.[12] Type 1 MD was assigned to the instance we reported. It is still unclear what causes MD pathophysiologically. There have been other explanations put out, including deletions in mitochondrial DNA or a malfunction in the lipolytic pathway brought on by

catecholamines. Between 60% and 90% of patients with MD also have persistent alcoholism, yet it is still unknown what causes this. Unlike lipoma, which is enclosed in subcutaneous space, MD fat accumulations are non-capsulated and dispersed along vascular and muscle planes.[13,14] Surgical intervention is usually necessary to address functional impairment and aesthetic concerns associated with Madelung's disease.[15,16,]

"Take -away lesson"

It is important to note that case reports provide observations from individual patients and may not always reflect the general population. Therefore, additional research, including larger studies and clinical trials, is necessary to establish comprehensive guidelines for the diagnosis, management, and understanding of Madelung's disease.

Awareness and Recognition: Madelung's disease is a rare disorder, and its symptoms may resemble other more common conditions. Case reports contribute to raising awareness among healthcare professionals about the existence of this condition, promoting early recognition and accurate diagnosis.

Diagnostic Challenges: The diagnosis of Madelung's disease can be challenging due to its rarity and resemblance to other conditions. Case reports provide valuable insights into the diagnostic process, helping to identify key clinical features, imaging findings, and laboratory tests that can aid in differentiating Madelung's disease from similar disorders.

Management Approaches: Case reports can shed light on different treatment modalities and their outcomes in patients with Madelung's disease. These reports can contribute to the development of evidence-based management strategies, including surgical interventions, non-surgical approaches, and potential future therapeutic options.

Disease Pathogenesis: Studying individual case reports can provide insights into the underlying mechanisms and pathogenesis of Madelung's disease. These reports may contribute to a better understanding of the genetic, metabolic, or hormonal factors involved in the development and progression of the disease.

Patient Support and Education: Case reports can help raise awareness among patients and their families about Madelung's disease, its potential complications, and available treatment options. Sharing such reports can offer support to individuals living with this rare condition and help connect them with appropriate medical resources and communities.

Patient Perspective

Initially, before visiting to this hospital, I visited many health centres for the same disease but I was not satisfied with their management and suggestions. As I met to the consultant here, he discussed entire complaints in details and precise diagnosis was made. I was well assured by the entire team and surgery was completed swiftly without any complications in aforesaid duration. I am very delight to say that I have been discharged within eight day of admission.

ACKNOWLEDGEMENT

The Authors Are Greatful To Dr. Hemant Toshikhane, Dean, Faculty Of Ayurved, Parul University; Dr. Omprakash Dave, Head, Department Of Shalya Tantra Parul Institute Of Ayurved, Parul University For Their Kind Support. We Are Also Thankful To The Staff Of Shalya Tantra Ot And Ipd, Parul Ayurved Hospital.

Conflict Of Interest- None

Informed Consent statement

Informed written consent was obtained from the patient for publication of this report and any accompanying images.

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CITATION OF THIS ARTICLE

Vaibhav Mishra, Parikshit Shirode, Lalita Dushad. Madelung's Disease a Rare Disorder with Lipomatous Deposits: A Case Report . Bull. Env.Pharmacol. Life Sci., Vol 12 [12] November 2023: 437-440