

## Multiple Symmetrical Lipomatosis in a 67-Year Old African Male: First Case Report within a Resource-Limited Setting

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### Abstract

**Introduction:** Multiple Symmetrical Lipomatosis (MSL) is a progressive disease presenting with symmetrically distributed (large) lipomata over the occiput, head, neck, shoulders and trunk resulting in gross deformity. Over 90% of cases are associated with chronic alcohol abuse, and associations with metabolic disorders are also common but malignancy is rare. To date there is only one report of MSL in an African-American patient and no reported cases in Africa

**Case Presentation:** We report the case of a 67-year old African male with a history of gout and chronic heavy drinking presenting with typical features of multiple symmetrical lipomatosis. The lipomata were successfully removed by a visiting surgical mission team.

**Discussion:** Exact pathogenesis and incidence of MSL remain unclear. Misdiagnosis is common and diagnosis in resource-limited settings should be prompted by the history of alcohol abuse and characteristic presenting features of multiple symmetrical lipomatosis. Challenges of surgical interventions in low-resource settings are discussed.

**Conclusion:** Clinicians should be aware of the condition and its association with metabolic disorders and rarely malignancy. In resource-limited settings, ethical and cultural factors should be considered before proceeding to surgery.

**Keywords:** Multiple symmetrical lipomatosis; Alcohol abuse; Surgical treatment; Resource-limited setting

### List of Abbreviations

ALP: Alkaline Phosphatase; ALT: Alanine transaminase; AST: Aspartate Transaminase; GGT: Gamma-glutamyl transferase; HDL: High Density Lipoprotein; LDH: Lactate Dehydrogenase; LDL: Low Density Lipoprotein; MSL: Multiple Symmetrical Lipomatosis; TSH: Thyroid Stimulating Hormone

### Introduction

Madelung's disease, Launois-Bensaude syndrome, otherwise known as Multiple Symmetrical Lipomatosis (MSL) is a slow progressing deforming disease presenting with symmetrical, unencapsulated lipomata. Lipomata typically occur in the occipital fossa, head, neck, shoulder and trunk distribution. The lipomata can grow extremely large but distal arms and legs are rarely affected [1,2]. The etiology remains unknown, but over 90% of cases are associated with chronic alcohol abuse and middle-aged men are the most susceptible [3]. Many other conditions have also been associated with MSL: metabolic syndromes with alterations in glucose tolerance; hypertension and hyperlipidaemia; hyperuricaemia; liver disease; hypothyroidism and

elevated parathormone [4-6]. Malignant transformation is reported in the head and neck region [7]. Clinically, the condition causes apnea, obstruction of vessels and nerves, dysphagia, myopathy and reduced head and neck mobility [8,9]. Due to the unsightly gross deformities patients often experience stigma and isolation. Treatment aims to control risk factors (e.g. alcohol abstinence) and surgical excision of the lipomata [10]. However, recurrences are described, even in patients who abstain from alcohol [11].

The incidence of MSL is unclear but is reported as highest in the Mediterranean with an estimated incidence of 1 in 25,000 Italian males [12]. There are no case reports from the African continent. We report the first case of MSL diagnosed and treated in a 67-year old African male in Madagascar. Pathogenesis, differential diagnosis and the challenges of treatment and follow-up in a resource limited setting are discussed.

### Case Presentation

The patient travelled 300 km from his village to a town in Southern Madagascar where the surgical charity, Mercy Ships was known to be conducting a patient selection process to provide free surgical care to those in need. He presented with multiple large, soft and mobile masses in a symmetric distribution along the spine, hypochondrium,

suprascapular, supraclavicular and occipital regions causing limited neck flexion and extension (Figure 1).

The masses first appeared 3 years ago, initially in the paraspinal region, then ascending to around the face and neck. The patient denied pain, headaches, visual or auditory disturbance, respiratory difficulties or swallowing problems.

He had a past medical history of gout for 5 years and significant alcohol abuse (65 centiliters per day of locally produced rum over the course of 50 years), although he reported cessation of alcohol intake since his last consultation for gout one month previously. Neurological, cardiovascular and respiratory examinations were unremarkable.

The patient was transported for further investigations and treatment in the Mercy Ships hospital located 1.268 km away from the patient selection site.



**Figure 1:** Showing multiple large, soft and mobile masses in a symmetric distribution along the spine, hypochondrium, suprascapular, supraclavicular and occipital regions causing limited neck flexion and extension.

Laboratory results on admission revealed a normal serum glucose (79 mg/dL), elevated total cholesterol of 233 mg/dL with elevated LDL levels (152 mg/dL) and normal HDL (44 mg/dL), elevated triglycerides of 185 mg/dL, strictly normal liver function panel (AST 18 IU/L, ALT 10 IU/L, GGT 41 IU/L, ALP 50 IU/L, LDH 221 IU/L) and normal TSH (1,29 mIU/L). HIV was tested negative. CT scan of the neck and chest demonstrate very extensive symmetric fatty hypertrophic change located intermuscular throughout the neck and spine (Figure 2). No focal masses or calcified deposits can be seen within the hypertrophied fatty areas.



**Figure 2:** CT scan of the neck.

There was no axillary mediastinal or hilar lymphadenopathy nor intrathoracic fatty hypertrophic change. The patient understood the importance of alcohol abstinence and agreed to a surgical procedure.

Six lipomata were excised in a surgery lasting 5 hours with 1200 milliliters blood loss. Many of the lipomata infiltrated deep into the muscle and surrounding tissues. Cytology results confirmed benign lipomata. Postoperative analgesia was provided with simple oral analgesics and the patient required follow up in outpatients for one month (staying in hotel-style accommodation) and was then discharge home to his village. The postoperative results are shown (Figure 3).



**Figure 3:** The postoperative results.

Six months later the patient returned for a second operation to excise ten lipomata from the trunk and shoulders. This procedure also lasted 5 hours but had minimal blood loss (300 milliliters) as these lipomata did not infiltrate the muscle as extensively as those excised during the first procedure. Again the patient required one month of outpatient follow up for wound care management and was then discharged home. Post-surgery results up till 5 months were satisfying and the patient was able to restore his village activities.

## Discussion

We present the first case of MSL in Africa. Our case was associated with chronic alcohol abuse but the exact pathophysiological mechanism for this is unclear although mitochondrial disorders (e.g. the m.8344A>G point mutation) in brown adipocytes have been reported [13,14]. MSL has been described in children but is commonest in middle-aged Caucasian males with a male to female ratio of 15:1 and a peak incidence between 40 and 50 years [15,16]. There is only one report of an African-American patient and therefore the incidence in Africans is largely unknown [9]. In low resource settings misdiagnosis can easily occur. Differential diagnosis needs to be made with obesity, buffalo hump seen in Cushing's disease and even deep vein thrombosis [16,17]. Moreover, since the disease progresses slowly, there is often a delay in diagnosis until extreme deformity becomes apparent. Our patient gave a 3-year history of progressive enlargement of the lipomata, but we suspect a longer prodromal phase with less obvious masses.

Diagnosis remains predominantly clinical with characteristic symmetrical, slow growth of soft, non-tender lipomata in the occipital region, neck, shoulder and trunk. A history of alcohol abuse is an important clue. Since there are no definitive inclusion criteria for clinical diagnosis described in the literature, a histological confirmation showing unencapsulated adipose tissue can be made [2,16]. We did not undertake histological diagnosis due to the lack of facilities and difficulty in transporting human tissue from a remote nature of the surgical setting. However, cytology in our case, confirmed benign lipoma. Compression of vital structures (e.g. trachea compression), extent of the lipomata and possible associated malignant processes are best evaluated by MRI or CT [18]. In our case we used CT which showed extensive symmetric superficial and deep intermuscular lipomatosis compatible with MSL.

Alcohol consumption measured as total adult consumption in litres per capita or pure alcohol per year is very low in Madagascar (2.5 liters). Nonetheless, there may be a higher than expected prevalence of episodic heavy drinking as 10-20% of male drinkers reports this type of behavior [19]. In our case, the man lived in a rum producing village and his high alcohol consumption was likely a contributing factor to the development of MSL. Therefore in resource-low countries, understanding the local cultural context may be a helpful part of the diagnosis. The lipomata may infiltrate deeply into the muscle and surrounding tissues which can lead to extensive blood loss and a longer duration of surgery than expected. Surgery and anesthesia should therefore be prepared for this especially in low-resource settings. Due to the large incision lines postoperative wound management can also be challenging in resource poor environments. In our case this type of surgical, anaesthesia and nursing care was provided by the non-governmental organization, Mercy Ships and including follow up in outpatients for wound care management. Surgery in low resource settings always implies additional risks due to numerous challenges such as post-operative infection control and patient follow-up. Therefore the decision to operate should always be made with sufficient consideration of the patient's circumstances and local possibilities [20,21]. In our case, Mercy Ships was able to offer the patient long-term post-operative hospital admission and rehabilitation with satisfying results 5 months post-surgery.

## Conclusion

We report the first case of Multiple Symmetrical Lipomatosis associated with chronic alcohol abuse on the African continent. Diagnosis is predominantly clinical characterized by a symmetrical distribution of large, soft and non-tender lipomata on the background of alcohol abuse. Clinicians should be aware of its association with metabolic disorders and rarely malignant disease. Social stigma is an important aspect of the disease and therefore esthetic surgery can restore a patient to 'normal' life. In a resource-limited setting, we recommend careful consideration of the ethical and cultural context before undertaking surgery.

## Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

## Competing Interests

The authors declare that they have no competing interests.

## Authors' Contributions

Veronique Suttels was responsible for the literature and primary diagnosis and referral of the patient. Tertius Venter was the patient's surgeon and interpreted further investigations. Michelle White reviewed the manuscript and added valuable comments for the first author.

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