Case report

Post traumatic membranous cystic lipodystrophy: A Case report and review of literature

ABSTRACT

Membranous Cystic Lipodystrophy (MCL) is a rare form of panniculitis diagnosed histologically by the presence of cystic cavities bordered by eosinophilic crenated hyaline membranes, which stain positively with Periodic Acid-Schiff (PAS).
It was first described as a morphological characteristic of Nasu-Hakola disease. Subsequently, this panniculitis has been reported in several other clinical circumstances, predominantly associated with vascular disorders.
We present a rare case of post-traumatic membranous cystic lipodystrophy.

Keywords: Membranous Cystic Lipodystrophy, panniculitis, adipocyte, necrosis, traumatic

1. INTRODUCTION

Membranous cystic lipodystrophy (MCL) is a rare form of panniculitis diagnosed histologically by the presence of cystic cavities bordered by eosinophilic crenated hyaline membranes, which stain positively with Periodic Acid-Schiff (PAS).[1,2]
It was first described in Nasu-Hakola syndrome, which combines dementia, sclerosing leukoencephalopathy, and polykystic bone lesions.[3,4]
Subsequently, membranous cystic lipodystrophy has been reported in various other conditions, including vascular disorders, autoimmune diseases, and, more rarely, post-traumatic cases.[5]

2. CASE PRESENTATION

We report the case of a 19-year-old female patient, born from a non-consanguineous marriage, with no prior medical history, who presented with a slightly painful swelling on the posterior aspect of her right thigh, which had been gradually increasing in size.
Upon questioning, she revealed a history of trauma (a fall down the stairs) that occurred 1 year ago. Additionally, the patient reported no other associated functional signs.
Clinical examination revealed a firm, adherent swelling, measuring 6 centimeters. The overlying skin exhibited mild induration and tenderness.
An ultrasound examination was performed, suggesting an atypical lipomatous tumor. Intraoperative findings revealed an adherent non-encapsulated tumor, from which a whitish fluid was expressed. (Fig. 1)
Histopathological examination confirmed the diagnosis of membranous cystic lipodystrophy. (Fig. 2)

3. DISCUSSION

MCL is a rare form of fat necrosis characterized microscopically by the presence of cystic cavities of fat necrosis lined by hyaline membranes that stain positively with PAS.[2]

It was first described in 1973 as a morphological characteristic of Nasu-Hakola syndrome. It combines a sclerosing leukoencephalopathy and membranocystic degeneration of bones and fat tissue.[3,4]

It is a rare hereditary condition with autosomal recessive transmission, caused by a mutation in the *TYROBP* or *TREM2* genes. These genes are involved in the regulation of immune responses, differentiation of dendritic cells and osteoclasts, and in the phagocytic activity of microglia.[4,6]

It begins typically manifesting during adolescence with pain or recurrent bone fractures in the distal extremities, skin indurations, and later by a presentation of presenile dementia.[6]

Subsequently, this alteration of adipose tissue has been reported in various local or systemic conditions such as venous insufficiency [7–10], diabetes [1,9,11], lupus [7,9,12], Behçet's disease [8], rheumatoid arthritis [13], scleroderma [1,8,12,14], dermatomyositis [15], T-cell lymphoma [16], hemodialysis [17], or following chemotherapy [2]... Furthermore, very few cases of post-traumatic MCL have been reported [2,18,19]. Table 1 summarizes the different conditions associated with MCL reported in literature.

The mechanisms that cause pseudomembrane formation remain unknown.

Machinami, based on an ultrastructural and cytochemical studies of MCL, believes that the pseudomembranous aspect is the result of the deposit of degenerated cell membranes from macrophages and necrotic adipocytes. [5,7,8]
This is suggested by the observation of free fat droplets released from disrupted fat cells, processed by macrophages in connection with a histiocytic infiltrate.[5,7,8,11]
Other possible pathogenic mechanisms include an inappropriate phagocytosis, a disproportioned proliferation of fat cell membranes, fibrinogen deposition, lipid metabolism disorders, interactions between connective tissue and free fat droplets… [5,11]

The mechanisms underlying adipocyte alteration and subsequently pseudomembrane formation are also unclear.
Based on histological examination data, Alegre et al. suggest that MCL can occur as a nonspecific result of compromise in the blood supply.[9]

This hypothesis, supporting the adipose tissue ischemia, is also endorsed by Machinami and sueki al. [8,10].
The role of ischemic injury has also been suggested in infectious, autoimmune and traumatic conditions.[5]

In our case, Nasu-Hakola syndrome was ruled out due to the patient's normal neurological and radiological examination (Fig. 3). Other vascular, autoimmune, and infectious causes were also excluded based on normal clinical and immunological assessments.

Therefore, we conclude a post-traumatic origin of MCL. We consider that the fall down the stairs likely caused a vascular disturbance leading to ischemic injury to the fat tissue which caused at the cellular level, an alteration of adipocyte membrane and metabolism,

interaction between its contents and surrounding tissues, resulting in phagocytosis and pseudomembrane formation.

4. CONCLUSION

Membranous Cystic Lipodystrophy is a rare and nonspecific form of panniculitis that can occur in various clinical conditions, rarely in post-traumatic cases. Its diagnosis relies on histology. The physiopathology remains poorly understood. Ischemic origin is the most widely supported hypothesis

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

I declare on my honor that the ethical approval has been exempted by my establishment.

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APPENDIX

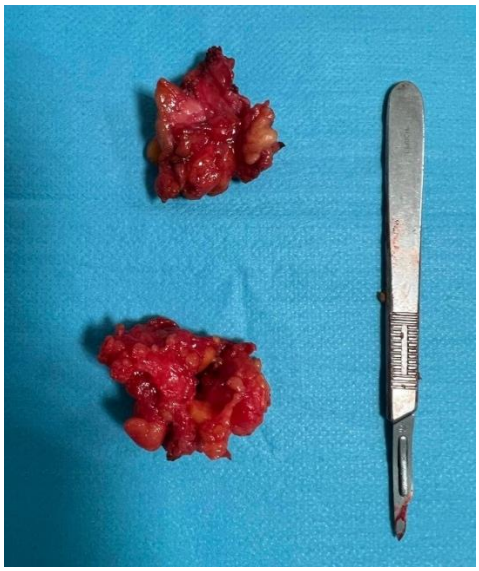

Fig. 1. Resected Specimen

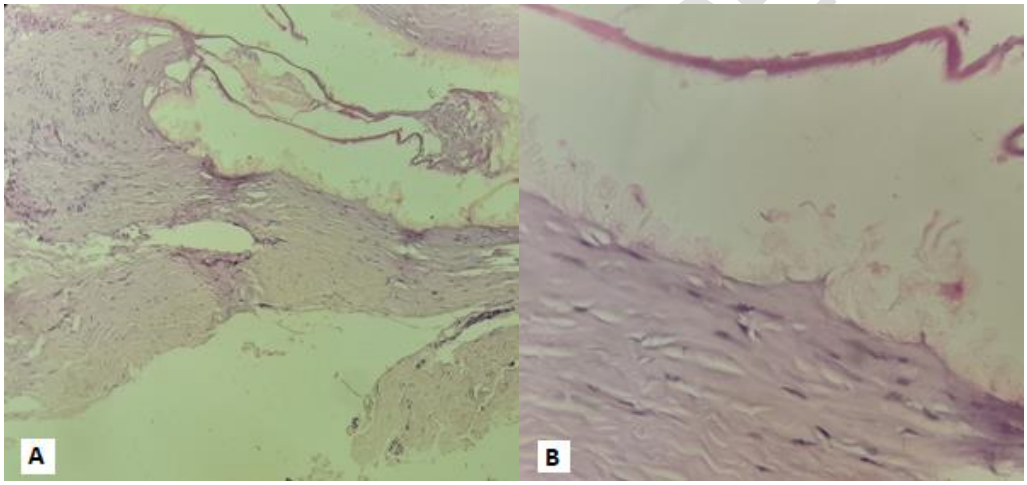

Fig. 2. Hematoxylin and Eosin (H&E) staining. A. (x 10): Fibrous tissue with cystic cavities bordered by hyaline membranes. B (x40) 40: Acellular eosinophilic membranes with crenated appearance lining the cystic cavities.

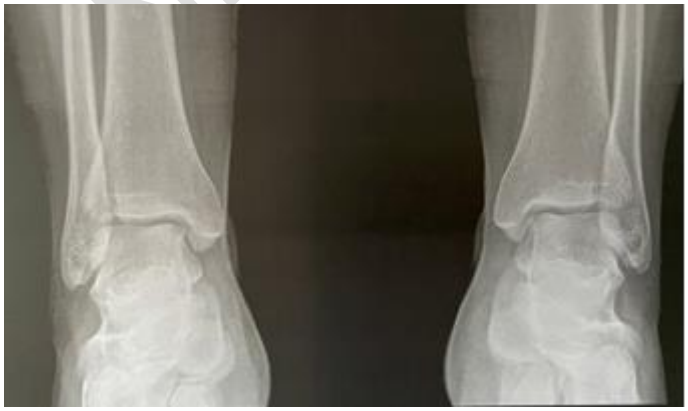

Fig. 3. Absence of polycystic lesions in X-Ray

Table 1. Different conditions associated with MCL

Condition	Source
Vascular	
Venous insufficiency	[7–10]
Arteriosclerosis	[10]
Thromboangiitis obliterans	[10]
Thrombophlebitis	[1,9]
Diabetes	[1,9,11]
Autoimmune	
Lupus	[7,9,12]
Scleroderma / Morphea	[1,10,12,14]
Dermatomyositis	[15]
Vascularitis	[9,14]
Behçet disease	[8]
Rheumatoid arthritis	[13]
Infectious	
Tuberculosis	[5,7,20]
Erysipelas	[7,14]
Neoplastic	
T-cell lymphoma	[16]
Drug-induced	
Chemotherapy	[2]
Post traumatic	[2,18,19]