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Massive Localized Lymphoedema and Surgical Treatment: Case Report

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Abtract

Lymphedema is the accumulation of lymph fluid in the interstitial space due to damage or absence of lymphatic channels (1). Massive localized lymphedema (MLL) is a rare and non-benign disease that is usually accompanied by obesity (2). MLL was first described as pseudosarcoma by Farshid and Weiss in 1998 as progressively growing pedicled benign masses in 14 morbidly obese patients (2, 3). With the great increase in the prevalence of obesity in the world, there is also an increase in the prevalence of MLL (4). MLL, which is an acquired chronic disease that occurs idiopathically, significantly reduces the quality of life (1).

Keywords: Liposarcoma; Lymphedema; Massive Localized Lymphoedema

Abbreviations: MLL: Massive localized lymphedema; MDM: Murine Double Minute

Case Report

A 40-year-old male patient was admitted to our clinic due to a progressively growing large mass in both groins, larger in the right groin, that had been present for approximately 3 years and occasionally discharged, causing difficulty in walking. The patient had no known disease or medication use other than sleep apnea and had no known history of sexually transmitted diseases. The patient had a history of smoking for 25 pack years. The patient weighed 160 kg and was 173 cm tall. Body mass index was 53.5 kg/m^2. The preoperative appearance of the patient is given in (Figure 1).

Excision of bilateral masses was planned under sedation and spinal anaesthesia. After appropriate anaesthesia, ICG dye was given to the patient and the lymphatic flows of the masses on the right and left were examined with the SPY device. Areas without lymphatic flow were included in the excision area, and the parts with lymphatic flow were planned to be included in the skin flaps to be used to repair the defect.

The patient's larger right mass was excised en-bloc. A tissue sample of the excised mass was sent for frozen pathological examination. The result was reported as lymphoid tissue. During the excision, it was noticed that the anatomical planes were lost, lymphatic fluid was constantly leaking into the surgical field, and the arteries and veins where lymphedema was present were quite enlarged. After excision, haemostasis was achieved and the skin flaps on the medial and lateral sides of the defect were advanced, and the defect was closed primarily. Two active drains were placed in the surgical field. Subsequently, the same procedure was applied to the mass on the left side and the operation was terminated. The masses excised from the patient is given in (Figures 2 & 3). Informed consent is taken from the patient.

Discussion

MLL frequently occurs in postmenopausal women at an average age of 48, is more common in women with a ratio of 2:1 and is seen at a high rate in the lower extremities, inguinal or suprapubic areas (2, 4). MLL is most commonly seen in the upper medial thigh region (s4). When the literature is investigated, there are also MLL cases seen in the upper extremity, popliteal fossa, penis, inguinoscrotal area, vulva, mons pubis, retroperitoneum, and lower abdominal area (4,5).

Lymphogranuloma venerum or penoscrotal filariasis are present in the etiology of scrotal massive lymphedema, also known as scrotal elephantiasis (1). Although the patient mentioned does not have a history of sexually transmitted diseases, obesity

appears as the only risk factor. The most important risk factor for MLL is a body mass index greater than 40 kg/m^2 (2). Metabolic syndrome accompanying obesity, hypothyroidism, and weight loss after bariatric surgery are other risk factors (4,6).

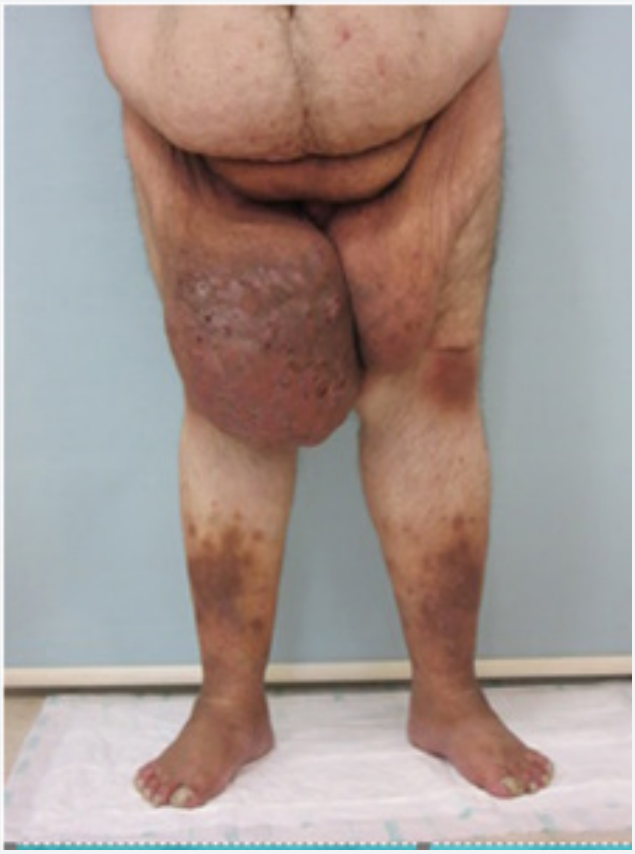

Figure 1: The preoperative appearance of the patient.

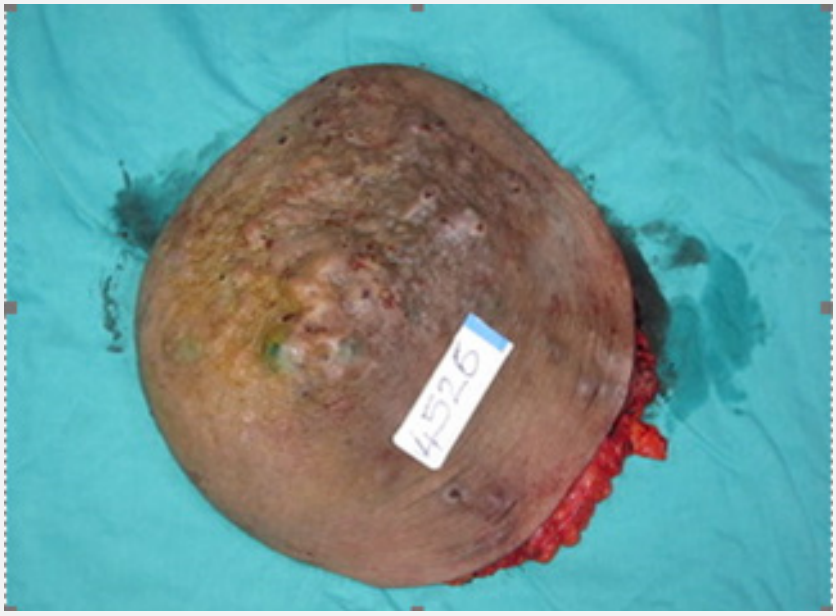

Figure 2: The patient's larger right inguinal mass.

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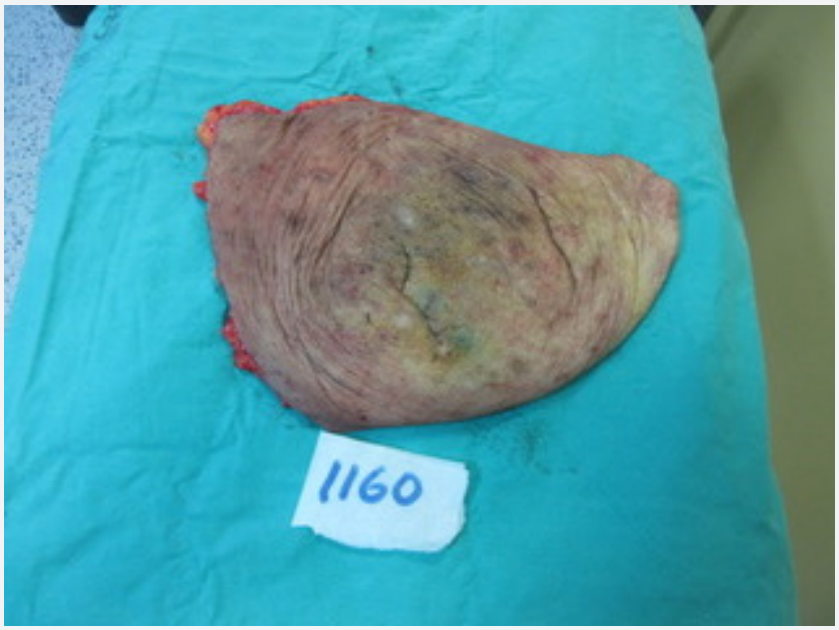

Figure 3: The patient's left inguinal mass.

Clinically, MLL is observed as a large soft tissue mass with ill-defined borders, covered with oedematous, thickened, hyperpigmented and orange-peel skin (2,7). Recurrent cellulitis may also be observed in patients (4). It is possible for this mass to be accompanied by open wounds and lymphorrhea (2). The skin characteristics in mentioned above are similar and are consistent with the literature in terms of edema, hardness, hyperpigmentation, orange peel appearance and tissue defects where lymph fluid leaks. In histopathological examination, there is no accompanying malignancy, dilated lymphatic channels and fibrotic, edematous tissue are found (2). Dermal fibrosis is accompanied by enlarged fibrous septa between fat lobules, increased number of stromal fibroblasts, multinucleated fibroblastic cells, vascular proliferation and lymphangiectasia (2).

MLL is multifactorial (2). Ischemia and chronic lymphedema resulting from localized lymphatic obstruction are thought to be the main pathology (2). Interstitial and lymphatic fluid accumulation stimulates the proliferation of keratinocytes, adipocytes and myofibroblasts due to hydrostatic and oncotic pressure changes (2,7). Interstitial and lymphatic fluid accumulation stimulates the proliferation of keratinocytes, adipocytes and myofibroblasts due to hydrostatic and oncotic pressure changes (2,7). Chronic lymphedema also causes basement membrane thickening and inflammatory cell migration in lymphatic channels (2). Increased tissue weight and the mechanical stretching effect of the mass reduce tissue perfusion and cause a micro-ischemic tissue formation (2). This ischemia causes extensive fibrosis and lobulation of the fatty tissue (2).

The differential diagnosis includes simple lymphedema, well-differentiated liposarcoma and Dercum's Disease (adiposis dolorosa) (4). MLL and well-differentiated liposarcoma are frequently confused. Liposarcoma reveals nuclear atypia and atypical lipoblasts in histopathological examination; also, liposarcoma is distinguished from MLL by the absence of lymphedema (4). In addition, in liposarcomas, MDM-2 (Murine Double Minute-2) and CDK-4 staining are positive, unlike MLL (5,7). In Dercum's Disease, which is seen in similar localization and presentation, the presence of painful lipomas and less presence of pathological skin findings and cellulite help in the differential diagnosis (4).

Angiosarcoma, in which chronic lymphedema is a risk factor, rarely accompanies MLL (2,8). The development of angiosarcoma on the background of chronic lymphedema is defined as Stewart-Treves Syndrome (4). Localized immunosuppression caused by chronic lymphedema is the main factor held responsible for malignant degeneration (4). Angiosarcoma should be kept in mind in cases of dramatic weight loss and lymphadenopathy that raise suspicion of malignancy in MLL patients (4). MRI examination should be performed in patients with suspicion of angiosarcoma, and a biopsy should be performed in the presence of nodules seen in angiosarcoma, although they are not seen in MLL in MR images (4). Since there is a potential for malignancy, long-term follow-up of patients is recommended (7).

The most important reason for patients to administer to the hospital is the limitation of movement caused by the mass. Surgical treatment is far from curative and is performed for symptomatic relief (2). Surgical treatment is indicated if the patient has mobility problems, recurrent infections, and suspicion of malignancy (2).

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While functional operations such as lymph node transplantation or lymphovenular anastomosis may be beneficial in mild cases, excisional treatments such as Charles or Thompson operations are preferred in advanced cases and are considered the gold standard treatment (1).

MLL surgery consists of very difficult operations for surgeons due to the fact that patients apply in the advanced stages of the disease in which they are immobile, anatomical planes are confusing during surgery, lymphedema fluid constantly leaks through the effected tissues, lymphedema tissue is not encapsulated, and its borders cannot be precisely determined (2). Blood and fluid loss due to damage to enlarged arteries, veins and lymphatics during surgery is very critical for patients (2). In scrotal massive lymphedema surgery, the lack of clear surgical plans during resection makes it difficult to protect the genital structures within the mass (1).

Conclusion

Due to the increasing prevalence of obesity, MLL is becoming an increasingly common disease. Diagnosis and treatment of this disease is becoming increasingly important. Excisional surgery options are the most important.

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