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VIEWPOINT

Surgical Management of Massive Lower Extremity Lymphedema Secondary to Castleman's Disease

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Patients suffering from angiofollicular lymph node hyperplasia, or Castleman's disease (CD), may present with debilitating, chronic lymphedema. As in the case below, treatment by radical excision can produce favorable results.

CASE EXAMPLE

A 54-year-old man presented for management of progressive, debilitating left lower extremity lymphedema due to multicentric Castleman's disease (MCD) (Fig. 1). The patient was confined to a wheelchair and had regularly infected ulcers caused by extreme venous stasis. Chemotherapy and other conservative methods did not slow the progression of the lymphedema. To avoid above-the-knee amputation, the surgical team proceeded with radial excision of the upper left lower extremity with direct closure of his relatively good-quality lateral thigh skin. Over 6 kg of lymphedematous tissue was removed, and debulking to the level of the muscle fascia allowed for tension-free closure. The saphenous vein was preserved to augment the lower extremity venous outflow, and the lower leg ulcer was debrided to healthy bleeding tissue and treated with wet to dry dressings in anticipation of ultimate skin grafting (Fig. 2). Other than a small area of dehiscence on his incision that healed by secondary intension, there were no major postoperative complications. The patient was subsequently able to ambulate and to perform daily tasks upon discharge, and his ulcerations improved significantly. The patient had no further complaints after surgical excision.

DISCUSSION

There are over 6,500 new cases of CD per year in the United States.[1] Unicentric CD is the enlargement of 1 lymph node with no sequelae and is often managed surgically requiring minimal further treatment.[2] MCD is the enlargement of multiple lymph nodes and is more difficult to treat because it can be accompanied by sequelae such as anorexia, anemia, leukopenia, and chronic lymphedema.[2] Chronic, progressive lymphedema results in fibrotic changes to the skin and subcutaneous tissues with accumulation of protein and inflammatory cell–rich lymphatic fluid and also enlargement of blood and lymphatic vessels, making surgical management challenging.[3]

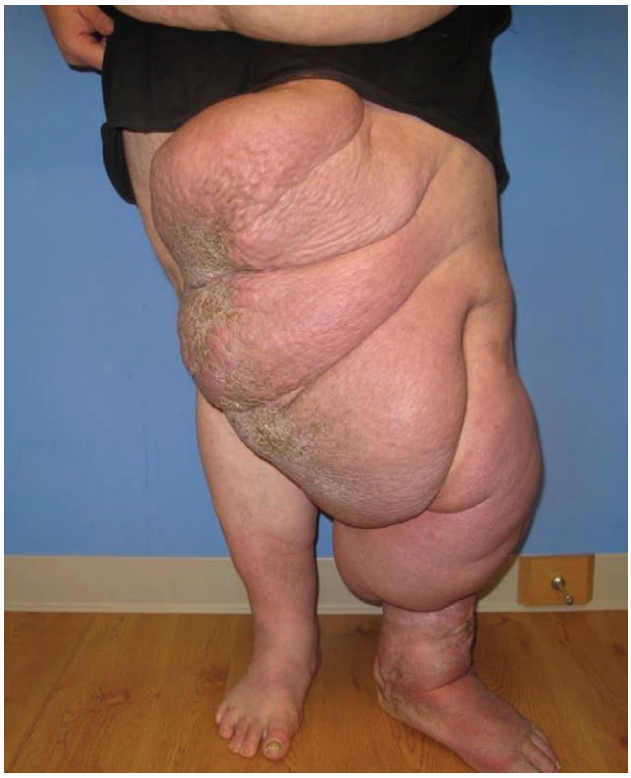

Fig. 1. Preoperative photograph of the patient's lower left extremity lymphedema due to MCD. The patient was unable to ambulate and perform daily tasks and had extreme venous stasis resulting in ulcers. Chemotherapy and other conservative treatments did not slow the progression of MCD.

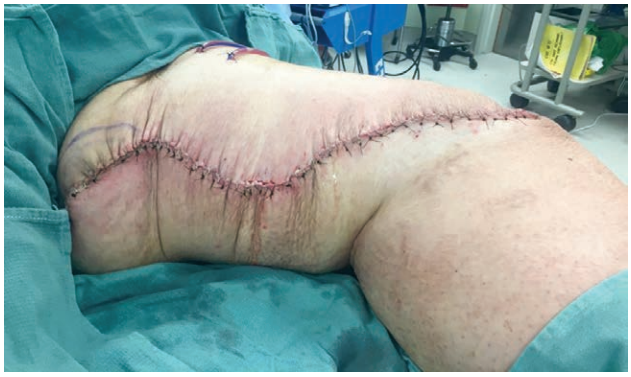

Fig. 2. Postoperative photograph of the patient's lower left extremity. Over 6 kg of tissue was excised, the saphenous vein was preserved, and the surgical wound was closed without tension. There were no complications after surgery, and the patient did not require further treatment.

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Chemotherapy (eg, cyclophosphamide), corticosteroids, radiation, and surgical excision have been used as treatment for MCD with varying success.[4] Documented cases of successful surgical treatment of MCD include excision of excess lymphedematous tissue and splenectomy when splenomegaly is a symptom of MCD.[5] However, recurrence may necessitate further treatment.[5] Because high levels of Interleukin-6 (IL-6) from the overproduction of hyperplastic lymph nodes correlate with the onset of CD, anti–IL-6 monoclonal antibodies (eg, rituximab) can reduce the exaggerated inflammatory response in individuals with MCD especially when paired with chemotherapy.[2]

We recommend a multidisciplinary, multifaceted approach for managing MCD with the intention of avoiding invasive, high-morbidity interventions (eg, amputation) when possible. In the present case, following failed medical management, radical excision was performed before considering amputation. Plastic surgeons should consider the possibility of CD if standard treatment of massive chronic lymphedema does not slow the onset of symptoms. All involved providers for patients with CD should be aware of both surgical and medical treatment options to ensure optimal, individualized management for each patient.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article. The Article Processing Charge was paid for by the authors.

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