Need To Treat Lymphatic Malformations

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Abstract

Both truncular and extratruncular forms of lymphatic malformations; as described in the Hamburg classification; remain difficult to treat. Most of these malformations are treated adequately or present with complications (recurrent infection, cellulitis, bleeding, skin ooze) increasing the therapeutic challenge. We present a case with complications of a LM highlighting the need for early intervention for these lesions. A 19 year old male presented with watery discharge from the skin over a left thigh swelling since childhood. He was diagnosed to have an extratruncular Lymphatic Malformation (LM). Treatment was deferred. He presented with necrosis of the skin and subcutaneous tissue after 3 months requiring debridement and split thickness skin graft.

Case History

A 19 year old boy had presented with a left thigh swelling with intermittent serous discharge from the skin since childhood. He had episodes of fever with redness and pain of the involved area suggestive of infection. MR imaging revealed multiple cystic spaces – duplex revealed minimal flow, a micro and macrocystic lymphatic malformation of the left leg extending into the pelvis (Figure 1) was diagnosed. He was advised conservative management.

Imaging

He presented 3 months later with necrosis of the skin and subcutaneous tissue. On examination, the left thigh was involved nearly circumferentially with an eschar (Figure 2).

Peripheral pulses were palpable and the calf was supple. He underwent debridement of the left thigh eschar, skin and subcutaneous tissue. Infected lymphatic fluid in the macrocystic lesions in the inguinal area was drained. Blood counts ruled out lymphocytopenia.

Clinical presentation- Infected lymphovenous malformation with necrosis of overlying skin

The culture revealed streptococcus sensitive to penicillin- patient was treated with a week of culture specific antibiotics. The wound was managed with dressings and a split thickness skin graft was applied once the granulation tissue was healthy. Graft take was good. The intermittent discharge from the surrounding skin persisted and the grafted skin also developed this at 18 month follow up (Figure 3).

He was continued on compression garments, manual lymphatic drainage (complex decongestive therapy) and repeated tetracycline sclerotherapy for the remnant lesions and put on oral antibiotic (penicillin) prophylaxis with which the number and severity of the infections of the infections have reduced (one in 3 years).
threaten vital functions, or cause complications (recurrent infection/intratresional bleeding) also warrant early intervention [5,6]. Surgical excision and injection sclerotherapy (bleomycin, tertracycline, OK 432, ethanol) are used in conjunction with compressive garments and complex decongestive therapy [4,7]. CO2, YAG and KTP lasers have been used to treat skin vesicles [8]. Complex debulking/reconstructive surgery is offered in specialized centers [4]. Patients may have alterations of the immune system with poor immunity secondary to lymphocytopenia-long term or peri-procedural antibiotics prophylaxis may be required [9]. Antibiotic prophylaxis helps in reducing the frequency of infection. However, patients must be counseled about the nature of the disease, instructed about how to identify early signs of infection and seek medical care early.

Newer antiangiogenic agents are under study; at present a multidisciplinary team approach consisting of vascular surgeons, interventional radiologists; physiotherapists, plastic and reconstructive surgery is best in planning treatment for these patients [1,10].

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References