A giant congenital cavernous lymphangioma of limb with bony lesions and extension into pelvis and retroperitoneum. Lower limb lymphangiomatosis with soft tissue involvement alone has been described where the age of onset was 11 and 12 years in two cases. In our case, the age of onset was 12 years. At 20 years he presented to us with elephantiasis of the right lower limb. A rare form of giant congenital lymphangiomatosis of the lower limb with ostolytic lesions in femur and tibia on roentgenographic skeletal survey has been reported but without any systemic involvement. In this case, the limb bones were spared. During last 40 years, in our lymphoedema clinic, we have managed a large number of secondary lymphoedema of various grades involving the lower limb. A majority of them were of filarial origin. However, we never found such variation of clinical presentation. The free flow of large volume of lymph from one area to another has never been reported. All the components of clinical features were unique. Even vascular malformation could not be ruled out. Thus, it posed difficulty in diagnosis and management. Since clinical evaluation did not lead to the exact nature of the lesion, we subjected him to detailed investigations that proved to be very useful to ascertain the definitive diagnosis of lymphangiectasis. Since no definitive surgical procedure is described for such a lesion, we planned staged excision. On exploration, the extensive dense lymphatic network soft in consistency was a remarkable feature in contrast to the secondary lymphoedematous tissue that is very firm due to fibrosis following repeated infection. However, it could be easily excised along with excess skin. The primary suturing followed by pressure dressing led to primary wound healing. Encouraged by the result of the first stage, we excised the upper thigh lesion after 6 months that also proved very satisfactory. We have followed the patient for more than 3 years now and the initial good result still persists without any evidence of recurrence.

Most cases of lymphangiectasis have extensive visceral involvement and poor prognosis. In this variant, it is limited almost exclusively to soft tissue of the lower limb and has good prognosis. This rare case posed unpredictable diagnosis with excellent surgical outcome.

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Lymphangiectasis of lower limb

S. B. Gogia
Sanwari Bai Surgical Centre, 28/31 Old Rajinder Nagar, New Delhi - 110 060, India

Address for correspondence: 28/31 Old Rajinder Nagar, New Delhi, 110060, India. E-mail: gogia7@gmail.com

Comments

The authors deserve to be congratulated for their excellent management and overall good result in a very challenging case. However, there is an issue regarding the diagnosis of the patient. Lymph oedema seems to be a neglected problem with overall poor outcomes and any perceived success brings about
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a sense of bewilderment. The flow of arguments to our minds suggests that the authors have been trying to justify the fact that the diagnosis was not lymph oedema because they got a good outcome!!!

To quote -
“Final diagnosis was established by lymphangiography and lymphoscintigraphy, which revealed marked dilatation of lymphatic channels in leg and inguinal region.”

The above is a hallmark of filariasis and occurs early in patients (this patient was 24 years of age), a stage which Pani et al.[1] called reversible oedema. Added to that, the patient was seen in a lymph oedema clinic being run in an endemic area (Eastern UP) with all clinical features, suggestive of lymph oedema. So to my mind, this patient had lymph oedema. Admittedly, such features are found more in Brugia Malayi (found in Kerala) where hydrocoel is not a feature (no mention in the data presented; so presumably, it was not present).

We find no mention of a test for filariasis. Although night blood smear examination is frequently false negative, serum filarial antigen testing has been in vogue since many years and should have been done. The authors themselves state that classic features of lymphangiectasis – in the form of visceral and bone involvement were not present. We would rather take the argument upside down i.e. if it was lymphangiectasis, the stated good results may not have been present.

We have attempted to treat both conditions and inevitably found better results in lymph oedema, especially if it is of filarial origin rather than any other cause. Of late, however, our results in post mastectomy lymph oedema are improving with CDT.[2] Results are worse in primary and other congenital causes of lymph oedema, wherever there is a paucity of lymphatic vessels. Surgery can remove the diseased tissue partly; recurrence is more dependent on the aftercare, notable prevention of infection.[3]

However, the excellent management and results do need to be show cased. We have to re-emphasise that with current management techniques, lymphoedema, especially of filarial origin, is treatable and occasionally curable. Surgery should, however, be used as an adjunct for the more advanced cases like the present one rather than stand alone therapy.

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