

A CASE OF PRIMARY LYMPHEDEMA*

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Key words : lymphangiography, lymphatic aplasia, lymphedema, Milroy's disease

Lymphedema is a swelling due to an abnormal collection of lymph in the interstitial tissue resulting from anomalies or functional overload¹. Although lymphedema can be secondary to neoplasia, trauma, irradiation, nodal resection or inflammation, in childhood most cases are of primary lymphedema². The edema is solid and may be complicated by verrucous hypertrophy of the skin and recurrent infections³.

Case Report

M.G., a 12-year-old boy, was admitted to Hacettepe Children's Hospital with the complaint of swelling of the left lower limb. He was born at term after a normal pregnancy and delivery. The swelling first appeared during the third month of life; it did not appear to interfere with his gait. There was no consanguinity in the family.

Physical examination revealed severe edematous swelling of the left foot, leg and thigh. The skin on the involved extremity was smooth and normal in texture and did not pit with pressure (Figure 1). Arterial pulses in the involved extremity were normal. The physical findings of the other systems were unremarkable.

Laboratory studies including a complete blood count, urinalysis and chest roentgenogram were all normal. Comparative bone roentgenograms of the lower limbs revealed no abnormality nor difference in size. A left lower limb lymphangiogram was then carried out. A radio-opaque substance containing 70% iodopyracet (Diodone®) and injected into the web space between the toes, diffused about the dorsum of the foot without filling any identifiable lymphatic vessels, thus indicating lymphatic aplasia (Figure 2).

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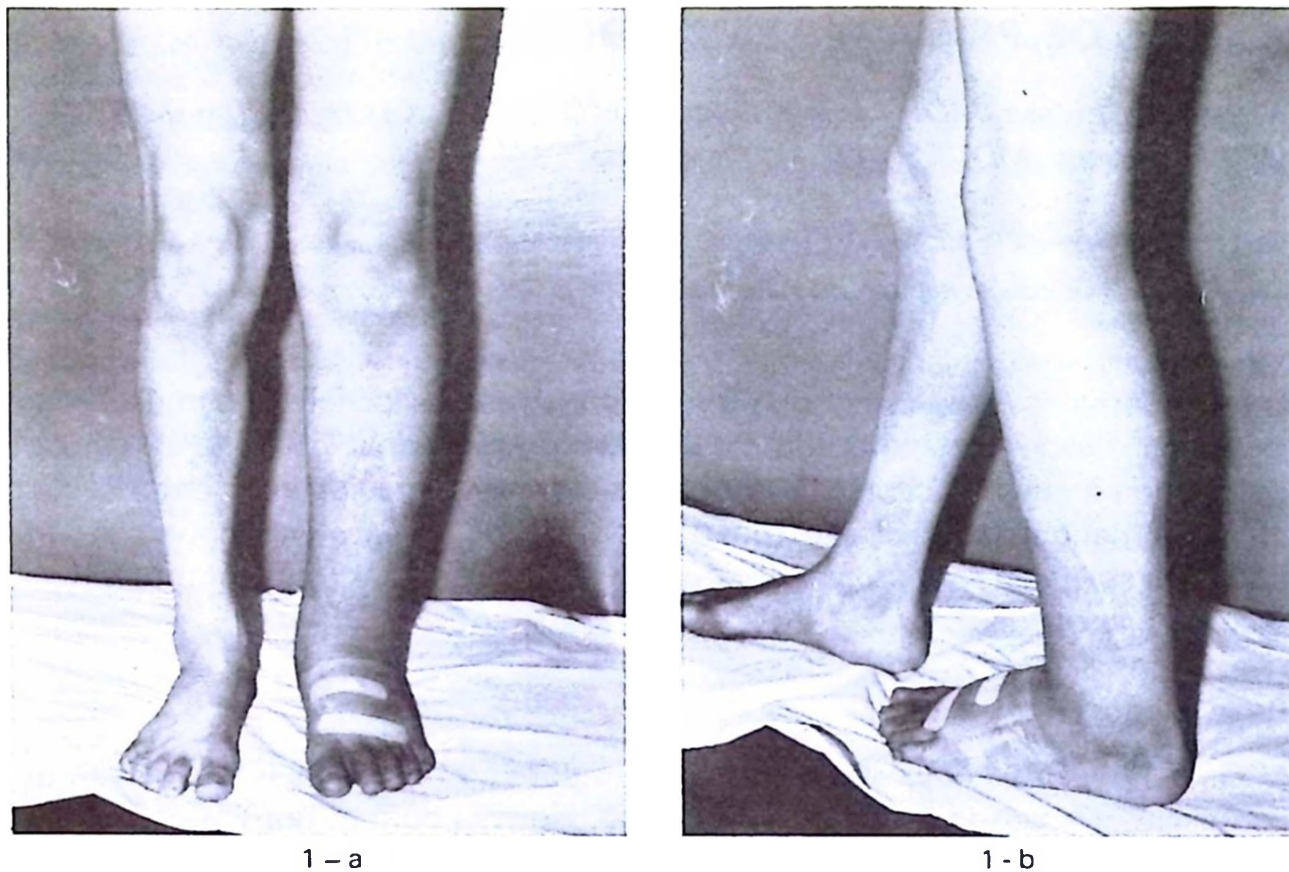


Figure 1 a - b
Severe edematous swelling of the left foot, leg and thigh.



Figure 2
Lymphangiography of the left foot revealed lymphatic aplasia.

Discussion

Three forms of primary lymphedema have been described. Whereas lymphedema precox usually first appears in early adolescence and lymphedema tarda occurs spontaneously in middle age, the more uncommon congenital lymphedema usually has its onset during the first few weeks of life⁴. When congenital lymphedema has a dominant trait, it can also be called Milroy's disease⁵.

In the normal extremity, the tissue fluid collected by the dermal lymphatic channels drains into the subcutaneous lymphatic system that accompanies the main superficial veins to the regional lymph nodes in the groin or axillae⁶. Although the exact pathophysiology of congenital lymphedema has not been clearly described, it is believed that the superficial lymphatic network is either hypoplastic or absent⁴.

It is possible to classify the cases of primary lymphedema into three groups according to lymphangiographic findings : hypoplasia, varicose dilatation and aplasia of the lymphatic vessels. Patients with aplasia or varicosity of the lymphatics tend to have edema from birth or from an earlier age than do those with hypoplasia⁷. In Kinmonth and coworkers'² 87 cases of primary lower limb lymphedema, there were 12 patients with aplasia. The lymphangiographic data for this patient also suggest lymphatic aplasia.

The problem of treatment of lymphedema is an unsolved one. Fortunately, in the case of most of the younger patients, the disease can be managed by nonoperative measures, including weight loss, elastic stocking support, physical activity and diuretics². At the present time, surgical intervention is not curative³.

Summary

A case of primary lymphedema in a lower extremity is presented. Lymphangiograms revealed the lymphatic aplasia of the left lower extremity. Pathogenesis and classification of primary lymphedema are briefly reviewed, and the importance of lymphangiography in diagnosis is emphasized.

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