LYMPHEDEMA (NONNE-MILROY TYPE) - A CASE DEMONSTRATION

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Lymphedema (Nonne-Milroy type) has its synonyms as Milroy disease, Nonne-Milroy-type hereditary lymphedema. It is autosomal dominant (1-10).

Clinical manifestations are (a) lymphedema, often of the lower limbs; (b) associated reported findings: congenital chylous ascites, protein-losing lymphangiectasia of the bowel, pleural effusion; (c) susceptibility of affected tissues to infection, acute nephritis.

Radiologic manifestations are (a) hypoplasia (decreased number and/or size of subcutaneous lymphatics) or absence of lymphatic channels; (b) dermal lymphatic filling in feet on visual and roentgenographic lymphangiograms; (c) weakness of the lymphatic wall resulting in extravasation; (d) absence of lymphatic valves.

The figure showed generalized thickening of soft tissue of the lower limbs of both sides with diffuse radiolucent lines due to lymphedema. The bony structures appear normal.

Lymphedema classification (1, 11-18)

1. Genetic lymphedema syndromes.
2. Primary lymphatic dysplasia : (a) association with chylothorax, chylous ascites; (b) other reported abnormalities: splenomegaly, thrombocytopenia, afibrinogenemia, hemangioma, lymphangioma, hydrops fetalis, congenital heart disease.
3. Acquired lymphedema.

TABLE I

Hereditary Lymphedema Syndromes

Type	Inheritance	Age at Onset of L	Associated Features
Milroy disease	AD*	At birth	Pleural effusion
Meige Disease	AD	Puberty	
L + intestinal lymphangiectasia	AD	Infancy	Diarrhea, growth failure
L + yellow nails	AD	Adulthood	Pleural effusion
L + distichiasis	AD	Puberty or later	Ectropion of lower lid, spinal anomalies
L + recurrent lymphangitis	AD	Childhood or puberty	
L + recurrent cholestasis (syndrome)	AR	At birth or childhood	May develop into cirrhosis of the liver
L + cerebrovascular anomaly	AD	Puberty or later	Pulmonary hypertension
L + ptosis	AD	Puberty	
L + microcephaly	AD	At birth	
Noonan syndrome	AD	Puberty	
Turner syndrome	"XO"	At birth	

"Modified after Kaarianen H: Hereditary lymphedema: A new combination of symptoms not fitting into present classifications. Clin. Genet. 26:254, 1984.

L = lymphedema; AD* = autosomal dominant; AR = autosomal recessive.

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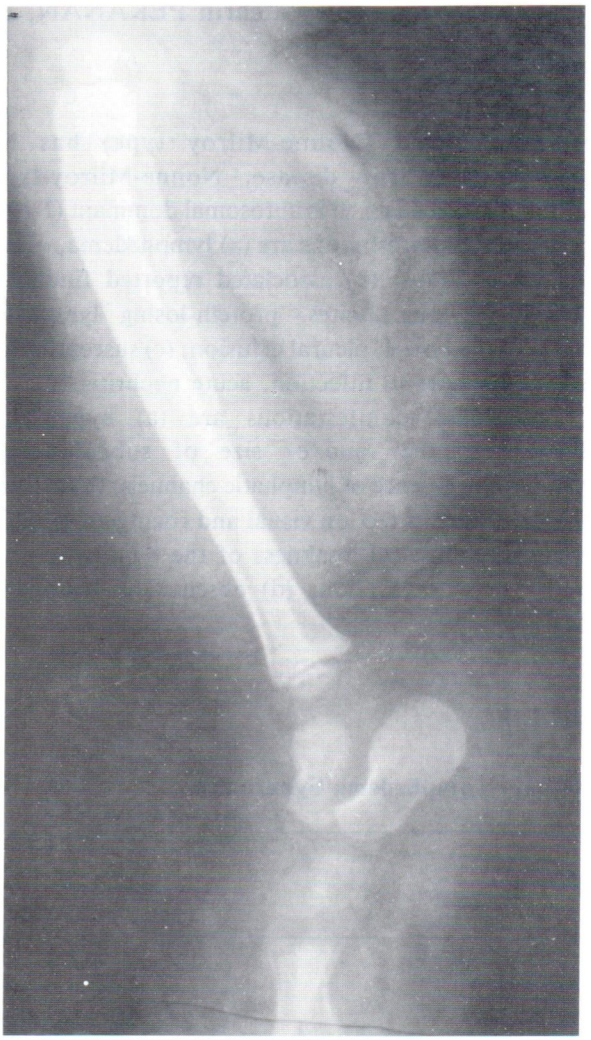

Fig. 1 Generalized thickening of the soft tissue of the lower limb with diffuse radiolucent lines due to lymphedema. The bony structures are normal.