

Clinical Image

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Idiopathic congenital lymphedema in a neonate

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A 16-day-old male neonate was referred with right lower limb swelling since birth associated with non-hemorrhagic pinkish-red patches on his skin over the thigh. The affected limb also had limited mobility. The baby was born at term through spontaneous vaginal delivery weighing 3400g with no other congenital anomalies. The antenatal and family histories were unremarkable. On examination, his entire right lower

limb was edematous with pinkish-red patches on the skin with no discharge; it was not tender and non-pitting with a palpable femoral pulse; the left lower limb and genitalia were normal (Fig. 1A, 1B). Labs showed CRP of 48 mg/L and a Doppler Ultrasound of the affected limb showed vascular insufficiency and queried vascular malformation (Klippel Trenaunay Syndrome).



Figure 1: A & B) Photographs showing right lower limb edema with a pink-red patch on the lateral thigh.

Primary lymphedema arises from abnormal lymphatic system development, which can be a complete absence of lymphatics or lymphatic hypoplasia. Lymphedema has been found in multiple sites, including the lower limbs, trunk, genitalia, head, neck, and internal organs, but lower limb involvement is the most common. [1] Primary lymphedema has also been associated with several genetic syndromes and cutaneous disorders like Neurofibromatosis type 1, Turner's syndrome, and

Klinefelter's syndrome which may have dysmorphic features or Karyotype confirmation. Congenital lymphedema without any dysmorphic features and unrelated family history is very rare and it occurs in 1:60,000 live births. [2]

The typical presentation of idiopathic congenital lymphedema includes unilateral or bilateral progressive edema of the lower limbs without any dysmorphism or family history of lymphedema. [3]

Differential diagnoses for the idiopathic congenital lower limb edema include deep venous thrombosis, hemangiomas, microcystic/macrocystic lymphatic malformation, noneponymous combined vascular malformation, Klippel-Trenaunay syndrome, post-traumatic swelling, Parkes Weber syndrome and many more. The sonography results of our patient were suggestive of Klippel-Trenaunay syndrome, a capillary-lymphatic-venous malformation of the lower extremity with overgrowth. [4].

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