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LYMPHOSCINTIGRAPHIC EVALUATION OF PRIMARY CONGENITAL LYMPHEDEMA*

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A-four-year-old girl with primary nonfamilial congenital lymphedema was evaluated by lymphoscintigraphy using ^{99m}Tc-Dextran. It was demonstrated that there was no accumulation of radioactivity in the left inguinal and femoral lymph nodes and faint accumulation in the contralateral lymph node. It was concluded that lymphoscintigraphy is a safe, reliable, nonninvasive technique which is helpful in the evaluation of a patient with lymphedema, especially small children in whom conventional contrast lymphangiography offers some challenges. *Key words: primary lymphedema, lymphoscintigraphy, ^{99m}Tc-Dextran..*

Lymphedema is usually difficult to diagnose in childhood. Although contrast lymphangiography offers an excellent morphologic evaluation of the lymphatics, the surgical exposure of the lymphatics that is required may be difficult in an edematous extremity, especially in children, in whom general anesthesia is also necessary. Moreover, the contrast material itself may be irritating, and its administration can lead to pulmonary complications[1]. We present a case of bilateral primary lymphedema in the lower extremity who underwent lymphoscintigraphy, a safe, noninvasive, reliable method which does not require general anesthesia, skin incision, or lymphatic dissection[1].

Case Report

A four-year-old girl with swollen lower extremities extending from the inguinal area to the fingertips was admitted to the Hacettepe University Children's Hospital. A gradual onset of swelling of the left foot which extended to the inguinal area was noticed soon after she was one year old. The patient

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subsequently developed swelling of the right foot. There was gradual swelling of both lower extremities until the age of four, when she was brought to our Hospital (Fig. 1).

Her prenatal, natal and postnatal histories were unremarkable, and there was no history of trauma, infection or familial occurrence of such lower limb asymmetry.

Physical examination on admission revealed a weight of 18.5 kg, height 102 cm, temperature 36° C, heart rate 168 /min, and blood pressure 120/90 mm Hg. Both lower extremities were grossly edematous. There was no pain or color change of the skin. The involvement of the right thigh was less than that of the left. The inguinal lymph nodes were not palpable. Microlymphadenopathy of the right mandibular and jugular were noticed. Laboratory studies revealed a hemoglobin of 12.8 g/dl, hemocrit 35 %, and white blood cell count 6000/mm^3.

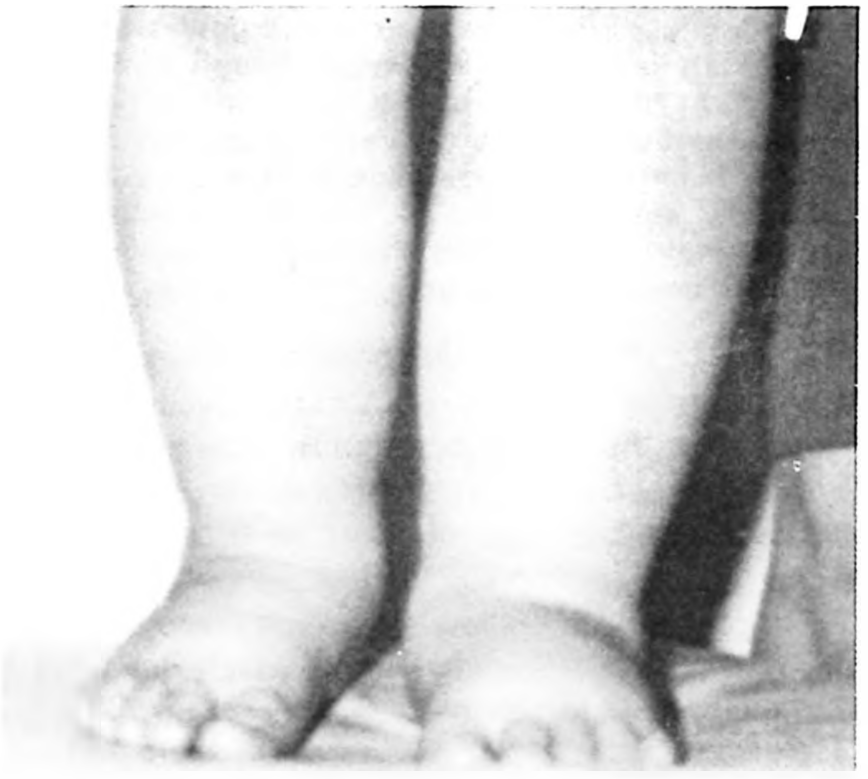

Fig. 1: Photograph of a four-year-old girl with long standing primary lymphedema of the lower extremities.

Since the patient developed a urinary tract infection following admission to the hospital, anesthesia required for contrast lymphangiography could not be given, and consequently, lymphoscintigraphy was performed. 150 µ Ci ^{99m}Tc-Dextran in a volume of 0.05 ml prepared in a previously described manner, was injected intradermally in the first web space of each foot[2]. Scintigrams were taken one, two, three and six hours postinjection, using a large field, all purpose, low energy collimator (Siemens ZLC II). Both legs and lower abdomen including the liver area

Fig. 2: Normal appearance of iliopelvic lymphoscintigraphy. Note intense uptake of inguinal lymph nodes.

Fig. 3: Iliopelvic scintigraphy of patient taken at the sixth hour after bilateral interdigital injection. There is diminished inguinal node accumulation on the right side, whereas, almost no accumulation on the contralateral side (arrows). Note markedly diminished visualization of the liver.

were imaged. Although the scintigrams failed to demonstrate the left inguinal lymph nodes, a faint visualization of the right inguinal lymph nodes was noted one hour postinjection. The external iliac lymph nodes could not be visualized. There was no remarkable change up to six hours postinjection. Almost all the radioactivity remained unchanged on both injection sites (Figs. 2, 3).

In light of all the clinical and laboratory findings, the patient was diagnosed as having primary nonfamilial congenital lymphedema. She was referred to the Department of Cardiovascular Surgery for a possible lympho-venous shunt operation.

Discussion

Primary lymphedema has been classified on the basis of age of onset as: congenital, lymphedema praecox (prior to age 35) and lymphedema tarde (after age 35). Lymphedema is also subdivided into familial and nonfamilial forms. The familial form is known as Milroy's disease, probably an autosomal dominant disorder[3-5]. Congenital lymphedema has been reported in Turner's syndrome (gonadal dysgenesis). A nonfamilial form of lymphedema has also been described. Letessier-Meige disease, a hereditary form, is the later onset form of primary lymphedema[5]. Since there was no familial history of this disease in our case, and the onset occurred at a very early age, the diagnosis of the nonfamilial congenital form was considered.

In order to establish the diagnosis of primary lymphedema it is essential to demonstrate the lymphatic pathways. For this purpose, conventional contrast lymphangiography is the classical imaging modality.

Although contrast lymphangiography can provide much better anatomic delineation of lymphatic channels and nodes, the lymphatic cannulation which is required is usually difficult in an edematous foot. Besides, it is known that lymphangiographic contrast medium may cause further inflammation of already diseased lymphatic vessels[1].

Lymphoscintigraphy plays an especially important role in diagnosing primary lymphedema in children, as was demonstrated in our case. ^{99m}Tc-Dextran is a known radiopharmaceutical for lymphoscintigraphy[6]. ^{99m}Tc-Dextran injected intradermally enters the lymphatic system and is deposited in lymph nodes along the route. After a node is saturated, the radioactivity passes onto further nodes. Some radioactivity bypasses all the nodes, and enters the thoracic duct resulting in visualization of the liver. Poor visualization, cut-off lymphatic channels and/or poor nodal uptake indicate the presence of hypoplastic or aplastic channels, as in our case. The absence of radioactivity in the liver is also a supportive finding.

In our case lymphoscintigraphy performed up to six hours postinjection showed almost a complete absence of radioactivity in the left inguinal lymph node groups, faint uptake in the right inguinal lymph node chain, and almost no visualization of the liver, indicating aplastic lymphatic channels on the left side and hypoplastic channels on the right side. These findings correlated well with the history of the patient, which revealed that the onset of swelling of the left foot preceded that of the right and that the involvement was to a less of a degree. According to the scintigraphic findings, the patient was diagnosed as having primary nonfamilial congenital lymphedema.

In conclusion, lymphoscintigraphy should be the first choice of imaging modality in the evaluation of the patient with lymphedema, children in particular.

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