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# Lymphangiography in Lymphangiomatosis of Bone

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*Summary*

The lymphographic findings of lymphangiomatosis of bone are presented in a case report. Late radiographs are essential to demonstrate the intraosseous localization of the contrast material. Lymphangiomatosis of bone seems to be due to lymphatic dysplasia with valvular insufficiency and has therefore to be considered as a congenital malformation.

Lymphangiomatosis of bone has first been described as an entity by *Bickel and Broders* in 1947. *Haas and Reichelt* (2) reported a family consisting of several blood relatives with evidence of lymphangiomatosis of bone. However, lymphographic investigations have been published in only four cases. *Kittredge* et al. (3) presented the case of a fifteen-year-old boy with a lymphadenopathy in the left groin, but no lymphedema. The plain roentgenograms showed multiple osteolytic lesions in the pelvis, hip and shoulder. Histologically the defects were caused by dilated lymphchannels. The lymphograms showed a disorganization of the lymphatic vessels in the thigh and pelvis, but the contrast material did not enter the bone lesions.

*Najman* et al. (4) described a boy with a lymphangioma in the inguinal region accompanied by lymphedema. Lytic bone lesions were found in the pelvis, limbs, chest and the skull. Lower extremity lymphography showed dilated lyphatics, but no contrast medium was located in the bone. Contrast material was injected into a bone cyst of the skull from where it passed into adjoining cystic lesions.

*Steiner* et al. (6) performed a lower extremity lymphography on a boy with pleural effusion and lytic bone defects in the humerus and the bony structures of the knee joints, but no lymphedema. Abnormal lymphatic vessels in the mediastinum and thoracic wall were demonstrated by lymphography. Although no contrast filling of the lytic bone lesions was obtained, the histological examination confirmed the presence of abnormal lymphatic channels in the bone.

Recently *Nixon* (5) demonstrated bone lymphangiomatosis lymphographically in a boy with lymphedema and osteolytic lesions in the femur and tibia. Contrast material was found within a cortical lesion of the femur 48 h following lymphography.

## *Case report*

A 5 1/2-year-old Sicilian girl with lymphedema and a naevus flammeus of the right leg was admitted to the Children's Hospital. Physical examination disclosed a difference in length and volume of the right lower leg. Laboratory studies were normal. The radiographs (Fig. 1) showed multiple small, mostly well defined lytic bone lesions without surrounding sclerotic or periostal reaction localized in the meta- and diaphysis of tibia and fibula, in the calcaneus, talus, cuboid of the right leg, in both femurs and in the os pubis and os frontale as well as in both forearms. Angiography did not reveal pathological findings in the arterial or venous system. Lymphangiography (Fig. 2) yielded the es-

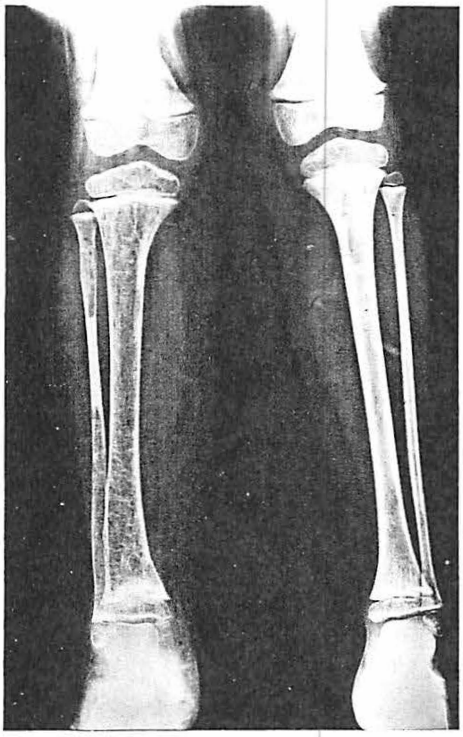

Fig. 1. Plain radiographs of the lower extremities. Multiple osteolytic areas within the right femur, tibia and fibula.

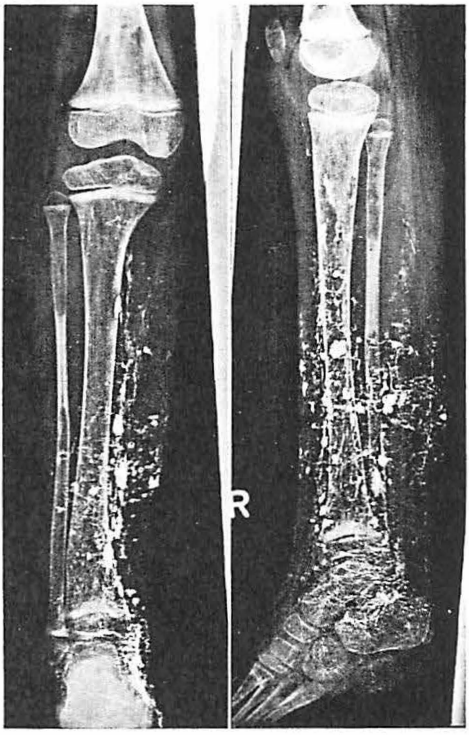


Fig. 2. Contrast filling of dysplastic subcutaneous lymphatics within the right lower limb 1 hour following injection of contrast material.

sential diagnostic information. The subcutaneous lymphchannels were dilated, drainage was delayed and abnormal subcutaneous lymphatics and interstitial staining was obtained due to extravasation. On the roentgenograms taken 6 days following lymphography (Fig. 3 + 4) contrast material was visible within the bone lesions of the calcaneus, tibia and os pubis. There were only a few abdominal lymphnodes filled with contrast medium. The diagnosis of a lymphangiomatosis of bone was established from these data. A biopsy of the fibula confirmed the presence of dilated lymphatic channels within the bone. The histological findings consisted of osteolytic lesions and cavities lined with an endothelium. No inflammatory reactions were observed, no fibrous tissue and no invasive growth nor any atypical cells were present.

## *Discussion*

Lymphangiomatosis of bone is a slowly progressive disease which cannot be challenged neither by radiotherapy nor by surgical treatment. The pathophysiology of the disease might be elucidated by the mechanism of contrast filling of the osteolytic bone areas in lymphography. Insufficiency and agenesia of the valves within the dysplastic subcutaneous and osseous lymph-vessels leads to a lymphatic backflow into the bones similar to the dermal backflow phenomenon. This might also explain the slow continuous progression of the osteolytic changes due to the steadily increased dilatation of the intraosseous lymphatic cavities. Consequently lymphangiomatosis of bone has to be interpreted as a congenital malformation of the lymphatic system similar to

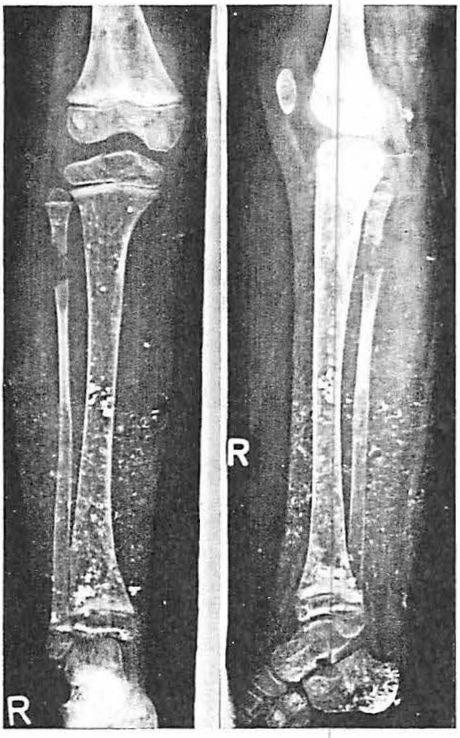


Fig. 3. Storage of contrast material within the cystic bone lesions (tibia, fibula, calcaneus, cuboid, patella) 6 days following lymphangiography. Bone defect within the proximal fibula following biopsy.

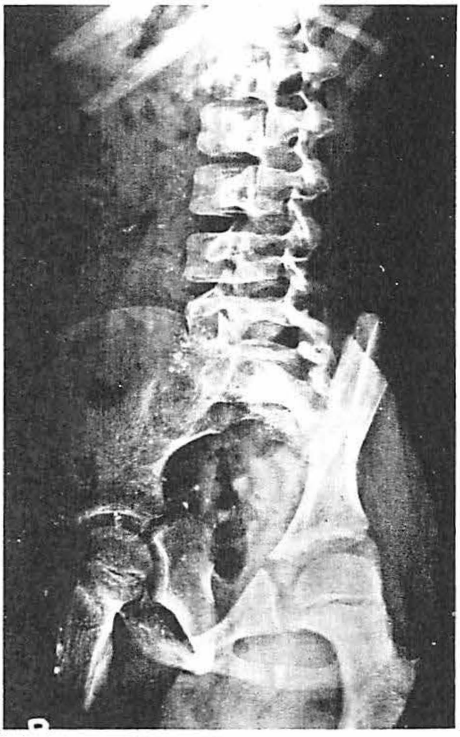

Fig. 4. Contrast filling of the dysplastic aortic lymphnodes 6 days following lymphangiography.

primary lymphedema and congenital lymphfistula. In this respect it is interesting to note that both the case described by *Nixon* and our patient had lymphedema in addition to the bone lesions and lymphography was able to visualize the direct connection between the dysplastic subcutaneous lymphatics and the cystic bone cavities, thereby demonstrating the impairment of the lymphatic circulation.

## *References*

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