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Lymphangiomatosis of Bone and Soft Tissue (Results of lymphangiographic examinations)

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Summary

The results of clinical roentgenologic examination of 2 patients with lymphangiomatosis are discussed. The analysis of angiographic findings has shown that arteries and veins remained unchanged in affected area. Lymphography revealed fully changed lymphatics in affected areas. Numerous markedly dilated lymph vessels and cystic cavities of different shape and size were noted. In bony spongiosus substance the fissural spaces were filled with oily contrast medium. Roentgenologically, they appeared as numerous small conglomerates limited to bone contours.

Lymphangiomatosis of bone has been reported under various names: massive osteolysis, essential osteolysis, acute spontaneous bone absorption, primary lymphangioma of bone and others. In 1838 Jackson described first lymphangiomatosis of bone. More recently 39 cases were reported (2).

The etiology of this disease is unknown, but all the authors consider lymphangiomatosis to be a basis of the process. Lymphangiomatosis occurs at the young age. The first symptoms include pathologic bone fracture and deformity. The process progresses as a rule and involves adjacent bones and soft tissues. Lymphangiomatosis is the subject of some case reports which suggest that all the bones of the skeleton can be involved. Bone lesions of the hand and the foot in this disease are rare. Specific roentgenologic signs are as follows: lytic osteopathy, sones of decreased density and cysts, the walls of which have shell-like contours or are eaten. Areas of sclerosis are always absent. The corticalis demonstrates thinning and protrusion. Histologic examination shows lymphatic sinuses limited to epithelium. The diagnosis is more often based on soft tissue histology. Lymphography is of great value as it can reveal considerable changes in lymphatics (4, 5, 6, 7).

The purpose of this study was to demonstrate diagnostic significance of lymphography during examination of patients with lymphangiomatosis of bone and soft tissue.

Case Reports

Case 1. Female patient, N., 31 years. At 12 years of age, pains and swelling in the right knee joint had arised without apparent cause, temperature was found to be elevated. The patient received antiphlogistic therapy. One year after onset of disease a fracture of the neck of the right femur happened. An open biopsy of soft tissues and bones in the upper third of the right femur was performed. Histologically, proliferation of epithelized lymph vessels was noted. Cystic angiomatosis of bones and soft tissues of the right lower extremity was diagnosed. The patient was seen at 29 years of age in the clinic of Research Institute of Medical Radiology. The right lower extremity was shorter by 18 cm than the left. The hip and knee joints showed limited motion. Soft tissues of this limb were swollen. The skin of the lower leg showed numerous blue patches and areas of thinning from which lymph exuded. Survey roentgenograms of the right lower extremity, the pelvis and the vertebral column demonstrated a

marked deformity of the femur, the ischial and pubic bones on the right. The femoral head and a part of the femoral neck were absent and the 5th lumbar vertebra was some what shorter. A deformity of the knee joint and massive marginal bone proliferations were observed. On the background of common osteoporosis the structure of these bones was found to be cellular with cystic changes in some areas. Follow-up roentgenography during 7 years demonstrated marked progression of the earlier changes and involvement of adjacent bones.

Selective angiography of right half of the pelvis and the lower limb was performed. Major arteries and veins appeared normal. Then, direct lymphography was performed using Myodil, which was injected in 7 ml into the lympl vessels on the dorsum of the feet. Lymphograms of the right lower limb showed marked changes in the lymphatics (Fig. 1). In soft tissues, numerous markedly dilated vessels and cystic cavities of different shape and size, where contrast medium persisted, were seen. Inguinal, iliac and lumbar lymphatics and lymph nodes on the right were not opacified because of increased capacity of the leg lymphatics. The lymph vessels of the left limb remained unchanged. Unusual lymphatics deviated from the left inguinal plexus towards the ischial bone. Analogous lymphatics deviated from the iliac plexus towards the upper flaring portion of the ilium and the 4th and 5th lumbar vertebrae. The most part of contrast material has filled fissural spaces, formed in spongy substance of these bones. Radiologically, numerous small conglomerates limited to bone contours were noted (Fig. 2). In contrast to lymphatics the lymph nodes showed milder changes. Their sinusal system was filled irregularly and stained less intensive. Diagnosis on the basis of lymphangiographic findings: lymphangiomatosis of soft tissues and bones of the right lower extremity, the pelvis and lumbar part of the vertebral column.

Case II. Male patient S., 12 years. Two years ago a strong pain in the right half of the pelvis appeared and temperature was higher.

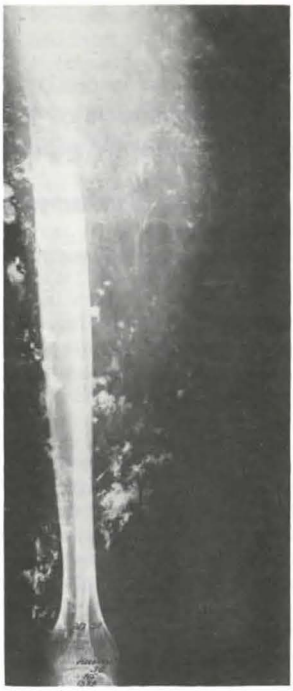

Fig. 1 Lymphogram of the leg shows numerous markedly dilated lymphatics and lymph cysts of different shape and size. Note persistence of contrast material. Capacity of lymphatic bed of the extremity is increased

Roentgenologically, an acute osteomyelitis was diagnosed. The boy underwent conservative treatment. On examination in the clinic of our Institute a slight protrusion of the right iliac bone backwards, swelling in the right inguinal region without clear limits were noted. The skin appeared normal. There was no pain on joint motion. Radiologic findings showed deformity of the pelvic girdle, structural cystic changes in the iliac, ischial and pubic bones on the right. Cysts were irregular in shape and different in size with clear contours. Angiography did not demonstrate any alteration of arteries and veins. Bilateral foot lymphography with the use of Lipiodol UF revealed no changes in limb lymphatics. In the upper inguinal region and the iliac one on the right, numerous deformed varicose

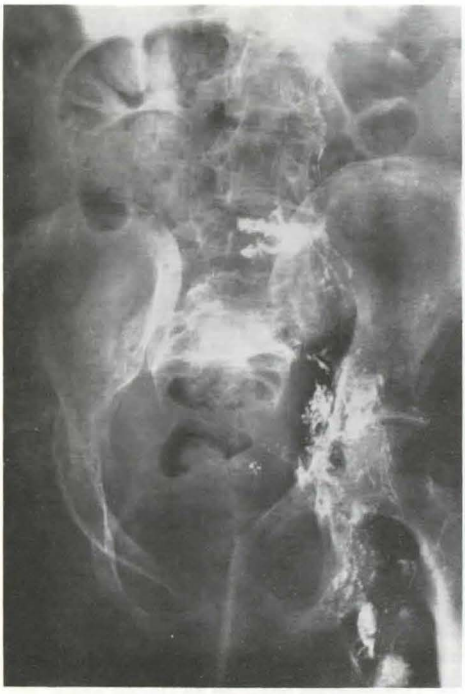

Fig. 2A Lymphogram obtained on filling phase. Contrast material is entering spongy substance of the 5th lumbar vertebra

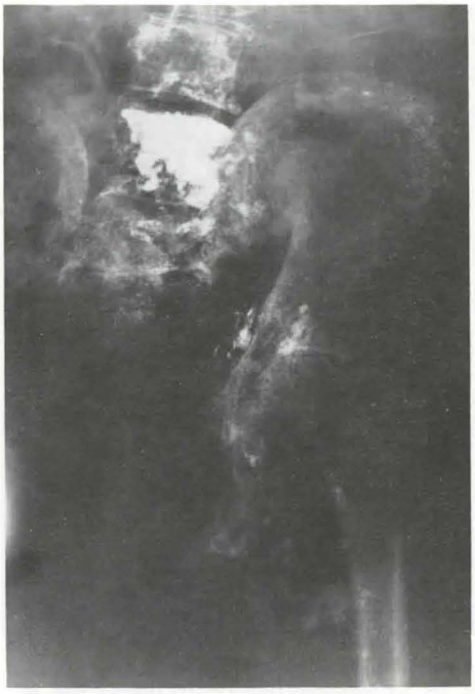

Fig. 2B Lymphogram obtained on storage phase. Note persistence of oily contrast material in the pelvic bones and especially, in the body of the 5th lumbar vertebra

lymphatics and lymph cysts of different size were seen. In dilated lymphatics and cystic cavities oily contrast medium broke down into separate drops (Fig. 3A). After 24 h contrast medium was on the whole removed from the dilated lymphatics of the right upper inguinal and iliac regions. It was accumulated by the acetabular roof, in rami of the ischial and pubic bones as well as in the upper flaring portion of the ilium on the right. In cystic cavities of changed bones storage of contrast medium was noted (Fig. 3B). 4 years later repeat lymphography revealed progression of lymphangiomatous changes: The right upper plexus iliaci communis and the plexus lumbalis were found to be affected.

Discussion

Lymphangiomatosis is a rare, insufficiently explored disease. Little is known about normal and pathologic state of the osteal lymph system. Lymphographic findings suggest deep morphologic and functional changes in the lymph vessels of bones and adjacent soft tissues. Angiography excludes angiomatosis of blood vessels. Follow-up roentgenography for a few years shows progression of the process and involvement of new regions.

At the present time it is discussed, whether the process originates in bone or in soft tissues.

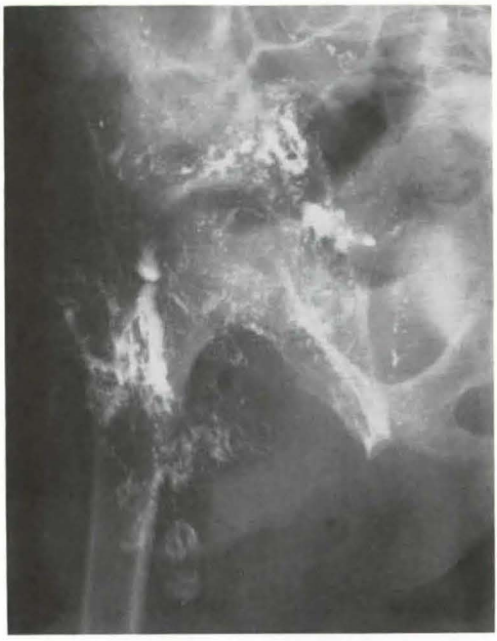

Fig. 3A Lymphogram obtained on filling phase. In the inguinal and iliac region on the righ numerous varicose lymphatics and lymph cysts of different size are seen

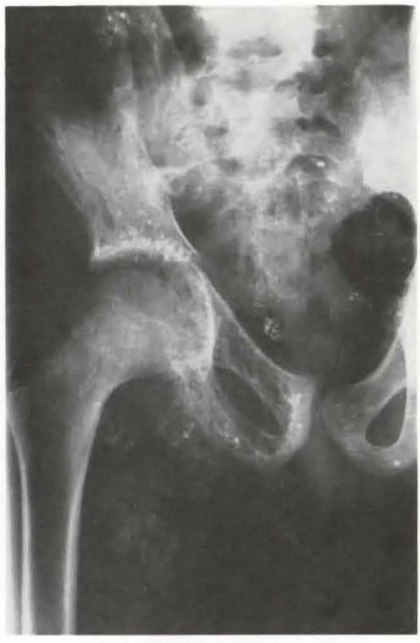

Fig. 3B Lymphogram obtained on storage phase. Note accumulation of oily contrast material by the acetabular roof, in rami of the ischial and pubic bones, as well as in the upper flaring portion of the ilium on the right

Bickel and *Broders* consider lymphangiomatosis as a primary lymphangioma of bone. These authors indicate that histologically, the periosteum of a normal bone contains a dense network of lymphatic capillaries. Therefore, disease originates perhaps from the periosteum. But taking into account the fact that the lymphatics in Haversian canals accompany the blood vessels we suppose that bones are primarily affected.

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