

Lymphangiomatosis of Bone (A Case Report)

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= Abstract = Lymphangioma of bone is extremely rare disease and characteristically involves multiple bones, thus justifying the term lymphangiomatosis. This case report describes a 4 year old boy who came in because of a pathologic fracture after a minor trauma. Cystic rarefactions were found by bone series, in various sites; both proximal humeri, scapula, clavicle, rib, both femora and pelvis. Open biopsy revealed a characteristic cavernous lymphangiomas.

Key words: *Bone, Lymphangioma, Cavernous lymphangioma, Lymphangiectasis, Lymphangiomatosis, Malformation*

INTRODUCTION

Since Bickel and Broders (1947) reported the first case of primary lymphangioma of bone in a 5 year old girl, less than 10 cases have been reported in English literature. These cases had without exception multiple lymphangiomas at the time of diagnosis. Although age ranged from 3 months to 55 years they tended to occur in pediatric age group. The usual sites of involvement were skull, spine, ribs, long bones and pelvis. Only one report (Hayes and Brody 1961) described lesion in the bones of the hand and feet. Recently we have experienced a case who presented a generalized lytic lesions in the bones that were biopsied and proven to be cavernous lymphangiomas.

CASE REPORT

This 4 year old boy was in good health until August, 1986, when he fell down and experienced pain in his right shoulder. He was seen by an osteopath and was managed for 2 weeks without improvement. Then he went to local hospitals, where fractures of right humerus and clavicle were found and was referred to Pediatric Orthopedic Service of Seoul Natinal University Children's Hospital on August 22, 1986.

At admission he was a moderately nourished and developed boy weighing 12 kg. Pulse rate was 100/min. Respiratory rate was 32/min, body temperature 37 C and blood pressure was 95/60 mmHg.

He was complaining of bone pain. Physical examination revealed essentially normal findings except for 1 1/2 finger breadth palpable liver. There was no specific limitation of motion or swelling in any area. There was no residual bruise or scar. X-ray findings of skeleton showed mutiple osteolytic metaphyseal lesions mixed with mild sclerosis in the long bones (Fig. 1). Involved bones were right scapula, right clavicle, ribs, both humeri, both femora and pelvis. A tapered pathologic fracture was noted in proximal metaphysis of right humerus with minimal periosteal reaction (Fig. 2). Massive osteolytic destructive lesions were noted in right scapula, right clavicle and left iliac wing (Fig. 3). Isotope bone scan revealed no increased uptake at the bony lesions except right proximal humerus.

Laboratory studies showed hemogram; hemoglobin 9 ml/dl, hematocrit 43%, WBC 8300/mm³, with blood chemistries of Ca 9.9 mg/dl, P 6.0 mg/dl, Alkaline phsophatase 197 IU/l, SGOT 16 IU/l, SGPT IU/l. RBC morphology was normal. Urinary vanilmandelic acid was 9.78 μ g/mg Cr (normal range 0.7-6.9). 99 mTC-MDP bone scan showed increase uptake in right humerus head and scapula.

Open bone biopsy was on Agust 29, 1986. The cortex and medulla were separately sampled from both proximal humeri.

Microscopically, bony trabeculae were widely separated by fibrous tissue which included numerous cystic spaces. (Fig. 4) These cysts were of

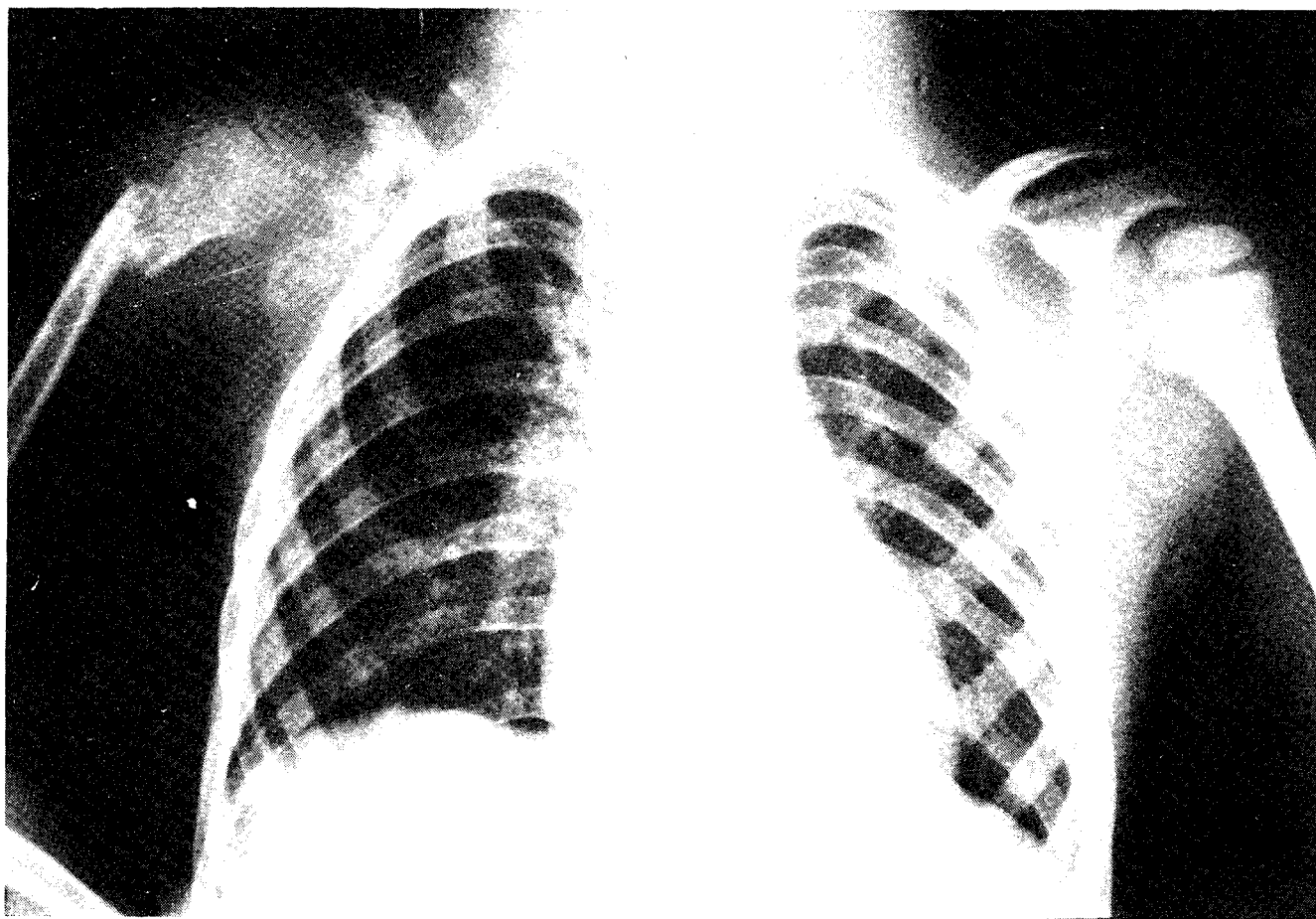


Fig. 1. Multiple osteolytic bone changes with mild sclerosis in metaphysis of both femora. Similar bony lesions in pelvis.

variable size and were scattered or coalescent to form a multiloculated appearance. The cystic cavities were filled with pink serous fluid and were lined by attenuated endothelial cells (Fig. 5). Small vessels containing few red blood cells were also seen in or near these lymphatic spaces. The specimen from the right humerus showed some newly formed bone probably representing prior fracture wound. No inflammatory cells were seen. The bony spicules were thin. Normal marrow element was not seen. The periosteum was slightly thickened in areas where subperiosteal new bone formation was seen. No abnormal vessels were seen here.

DISCUSSION

Lymphangioma of bone has been reported under various names; cystic lymphangiectasis (Hayes and Brody 1961), cystic angiomatosis (Moseley and Starobin 1964), generalized primary lymphangioma (Harris and Prandoni 1948), and multiple lymphangiectases (Cohen and Craig 1955). By this it is clear that many authors tended to think this entity as a non-neoplastic syndrome rather than true

primary tumor of the bone. Some authors advocate hamartomatous nature of the lesion (Moseley and Starobin 1964). As most extraskeletal lymphangiomas are considered as congenital malformation, skeletal lymphangioma could also be considered malformative. It is particularly true in cystic or cavernous varieties of lymphangioma.

Histopathologically the lesion seen in this case could be categorized into cavernous lymphangioma, instead of simple lymphangiectasis or cystic lymphangioma. We reserve the term lymphangiectasis for the dilatation of lymphatics in the tissue where the lymphatics are of indigenous structure. Besides, because the bone and marrow are not normally showing demonstrable lymphatics, it is probably not proper to use the term lymphangiectasis. Cystic lymphangioma is differentiated from cavernous type in extraskeletal tissue by having few larger cysts and involving larger lymphatics and not sharing a common wall of the cystic spaces (Park and Chi 1980). Since lymphangioma of bone involved multiple bones in all cases and sometimes associated with other anomalies such as congenital

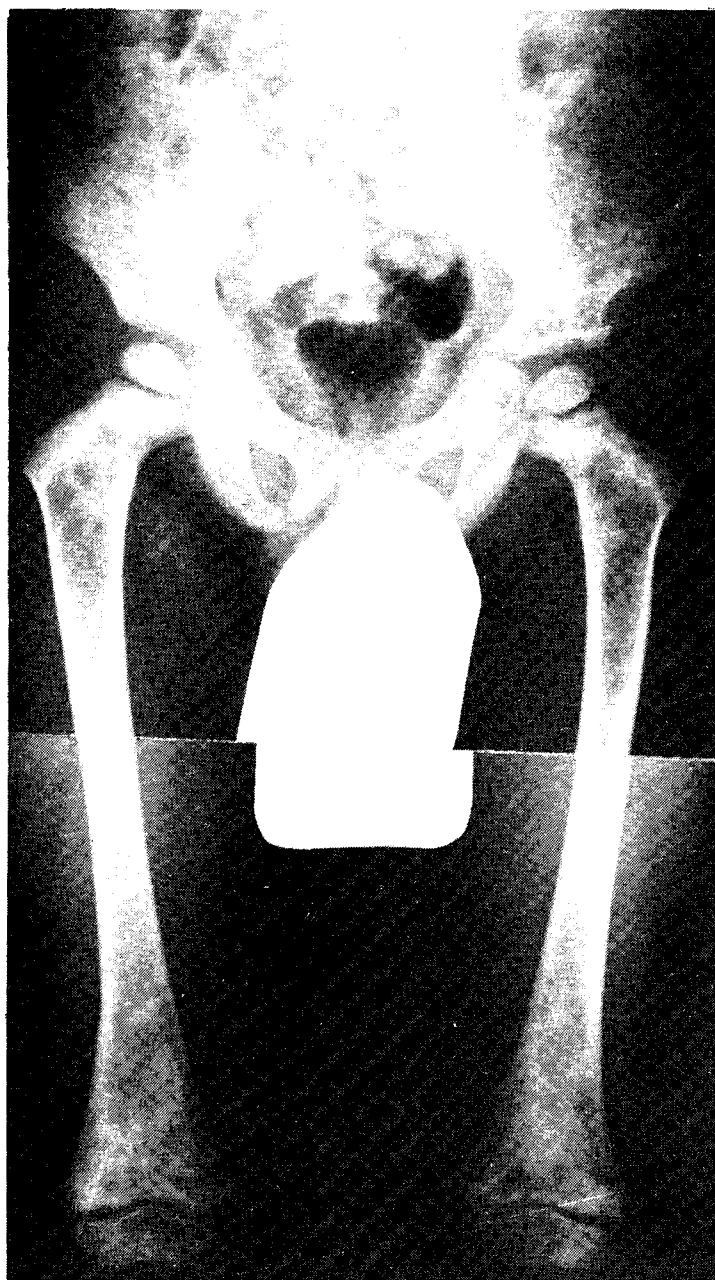


Fig. 2. Tapered fracture in proximal metaphysis in right humerus with underlying osteolytic bony lesions (Left). Left humerus shows same bony lesions as other long bones (Right).

lymphedema or splenic involvement (Cohen and Craig 1955) we think these lesions could best be named as lymphangiomatosis.

Although fibrous element has not been specifically mentioned in the previous reports it was fairly prominent in our case. In fact the first few superficial cuts of the specimen showed only marrow fibrosis and occasionally new bone formation. Only deeper cuts of the block could demonstrate characteristic cavernous lymphangioma. When we reviewed the previous slides again we realized the some lymphatic spaces were actually collapsed



Fig. 3. Massive osteolytic destructive bony lesion in right scapula and right clavicle.

and unnoticed. Therefore careful search for any dilated thin walled vessels appears to be very important for the detection of this lesion.

Among patients who have been followed over a period of years, the course of bone lymphangiomatosis has been quite variable. Several authors (Bickel and Broders 1947; Cohen and Craig 1955) have described progression of the lesions with multiple fractures, while others (Falkmer and Tilling 1956; Harris and Prandoni 1950) have shown no increase in size of the lesions. The present patient is under close follow up and it would be interesting to demonstrate the evolution of the bone defects radiologically in future.



Fig. 4. Photomicrograph of bone lymphangioma. Multiple irregular cavities are seen in medullary spaces. Note fibrous stroma around vascular channels. (H&E x100).



Fig. 5. Bone lymphangioma, showing multiple cavities partly filled with lymph fluid and lined by a single layer of endothelial cell. (H&E x270).

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=국문초록=

골의 림프관종증

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골의 원발성 림프관종을 대단히 드문 질환으로 영문 문헌상 그 보고례가 10예 미만이다. 본 질환은 다발성으로 여러 골을 침범하기 때문에 림프관종증이라고 흔히 불려지고 있다.

본예는 4세 남아로서 경도의 외상으로 인하여 골절을 당한후 입원하여 검사한 결과 양쪽 근위 상박골, 견갑골, 쇄골, 늑골, 양쪽 대퇴골 및 골반골을 침범하고 있는 낭성 변화를 나타내었으며 이 병변은 현미경적으로 동성 림프관종이었다.