Journal of Korean Medical Science
Vol 4, No. 2, P 97-101, June 1989

Telangiectatic Osteosarcoma

— A Case Report —

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Telangiectatic osteosarcoma is a rare and special variant of osteogenic sarcoma with distinct radiologic, gross and microscopic features. This tumor is predominantly lytic, destructive tumor without sclerosis on roentgenogram, and is soft and cystic on gross examination. Histologically aneurysmally dilated spaces lined or traversed by stromal cells producing osteoid are noted. This report concerns a case of telangiectatic osteosarcoma occurring in a 7 years old boy. He presented with pathologic fracture of the right distal tibia, followed by a purely lytic lesion on X-ray examination. This lesion recurred five times during a span of one year. Microscopic features of the biopsy specimen was difficult to differentiate from aneurysmal bone cyst because of prominant blood-filled cyst formation. It was finally identified as osteosarcoma from the below-knee amputation specimen through the close examination for anaplastic osteoid-producing stromal cells in the septa that separate the blood cysts.

Key Words: *Telangiectatic osteosarcoma, Bone tumor, Osteosarcoma*

INTRODUCTION

Telangiectatic osteosarcoma was first described by Paget (1854). The term "malignant bone aneurysm" was originally coined in 1903 by Gaylord to denote a bulky, hemorrhagic, poorly ossified telangiectatic osteosarcoma. Ewing (1939), however, considered this sarcoma histologically to be a variant of osteogenic sarcoma. Telangiectatic osteosarcoma is now recognized a specific entity with distinct radiologic, gross and microscopic features (Farr et al. 1974; Matsuno et al. 1976; Unni 1988). Conventional osteosarcoma with telangiectatic foci should be differentiated from this tumor. The differential diagnosis both radiographically and histologically include several benign lesions such as aneurysmal bone cyst and giant cell tumor, among others (Huvos et al. 1982).

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This report concerns a case of telangiectatic osteosarcoma initially thought to be aneurysmal bone cyst in a 7-year-old boy who underwent several operative procedures for removal of recurrent bonelesion during a span of one year.

CASE HISTORY

This 7-year-old boy was admitted to the Department of Orthopedic Surgery of the Seoul National University Hospital because of recurrent bouts of cystic bony lesion in the right distal tibia. This lesion recurred five times and was curetted each time during period of one year from August 8, 1986 to August 3, 1987.

He had a past history of ankle fracture after a blunt trauma on the right knee during a football game on June 30, 1986. At follow-up examination 6 weeks later a well circumscribed radiolucent mass was noticed in the metaphysis of the right distal tibia, for which curettage and bone graft was done in August 1986. In November 1986 intralesional steroid injection was done. He was referred to Seoul National University

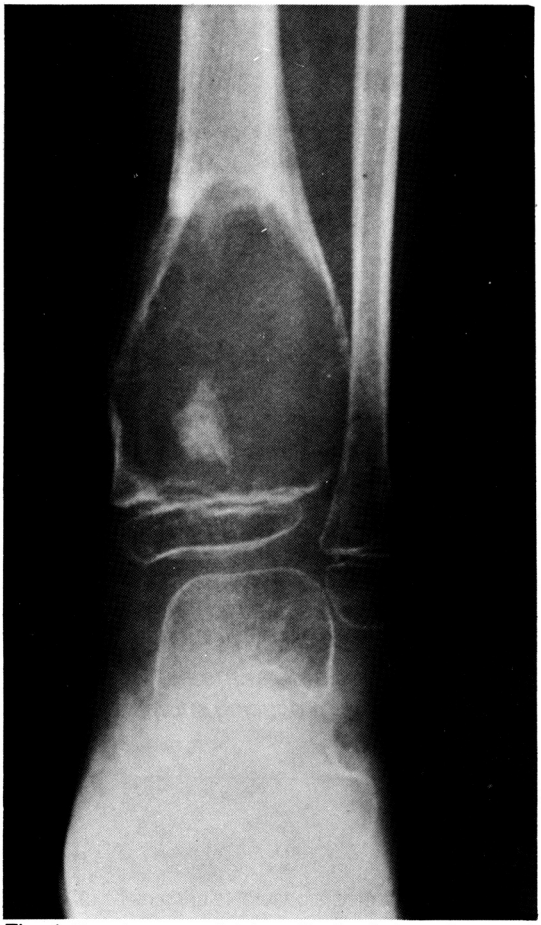

Fig. 1. Roentgenograph taken after the first curettage and bone graft showing a predominantly osteolytic expansile lesion without marginal sclerosis and periosteal bone production.

Children's Hospital on December 10, 1986, where aneurysmal bone cyst was suspected on the basis of radiographic findings (Fig. 1). At the first admission he had no subjective symptoms and steroid injection was given. Two months later he was readmitted because of swelling and tenderness in the right leg. X-ray films of the lesion showed a slight increase of mass in size. Operation revealed a walnut-sized encapsulated cyst which was curetted followed by insertion of bone chip. Three months after the second admission he experienced pain together with skin ulcer and blood oozing from the lesion site. At operation a cavitary lesion filled with dark red blood was found and extended toward the diaphysis as well as the distal epiphysis of the right tibia, with invasion into the surrounding soft tissue. The periosteum appeared intact. It was also curetted. Thereafter, this lesion recurred twice within the next 3 months. Dark blood of 200ml gushed out from the bone lesion during the last operation. Curettage and cement fixation was done. He underwent below-knee amputation because of recurrent lesion associated with intractable pain. After reviewing the entire speicmens and critical re-evaluation of radiographs was done. Finally, he underwent above-knee amputation under the pathologic diagnosis of telangiectatic osteosarcoma.

PATHOLOGIC FINDINGS

Microscopically, sections taken from the first curettage in our hospital revealed blood-filled spaces that were outlined by trabeculae of fibrous tissue or granu-

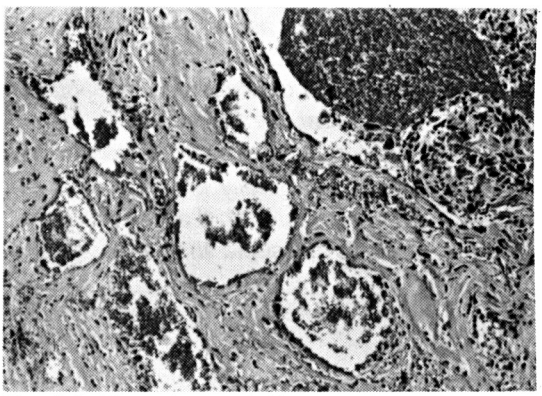

Fig. 2-A. Photomicrograph (the second curettage specimen) showing blood filled spaces separated by less cellular fibrous connective tissue. Cellular anaplasia is absent (H&E ×100).

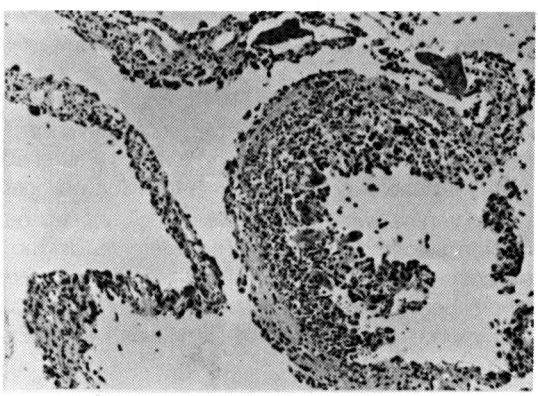

Fig. 2-B. The septa showing spaces lined with mononuclear stromal cells and osteoclastic giant cells. This component appears to be aneurysmal bone cyst (H&E ×100).

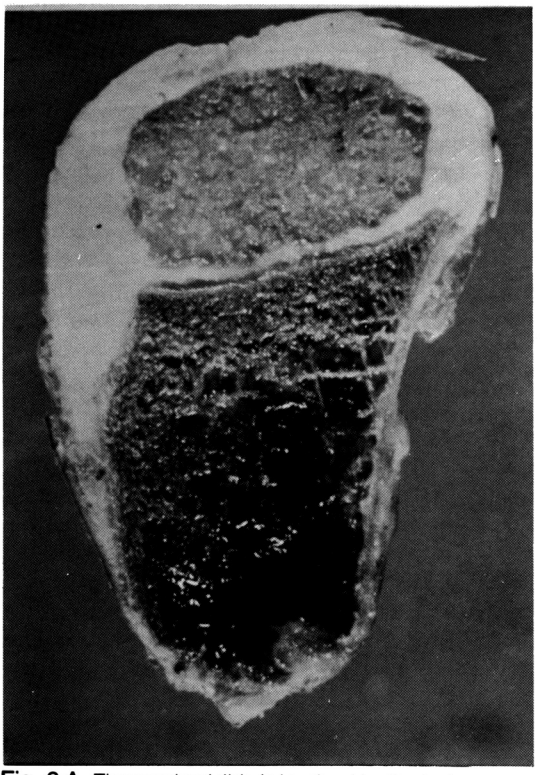

Fig. 3-A. The proximal tibia is involved by the lesion composed of hemorrhagic cystic cavities. This lesion do not extend into the epiphyseal plate.

lation tissue (Fig. 2-A). There were osteoclastic multinucleated giant cells, fibroblastic stromal cells, hemosiderin laden histiocytes and inflammatory cell in the septa (Fig. 2-B). The stroma was loose and vascular as seen in aneurysmal bone cyst. The histologic findings were reported to be consistent with aneurysmal bone cyst. Microscopic examination of tissue obtained from the second to the fifth operation showed cystic structure of fibrous and granulation tissue containing multinuclearted giant cells, benign mononuclear stromal cells and inflammatory cells. These findings were similar to those of the first biopsy. However, among benign stromal cells scattered were focal collections of larger cells with large atypical nuclei and prominent nucleoli. The proportion of these anaplastic cells was increased in subsequent recurrent lesions. No solid areas with bone or cartilage formation were found. Pathologic diagnoses were aneury smal bone cyst with stromal atypia.

Below-knee and above-knee amputated specimens showed basically identical fealures. They exhibited an ill-defined large areas with massive bone destruction.

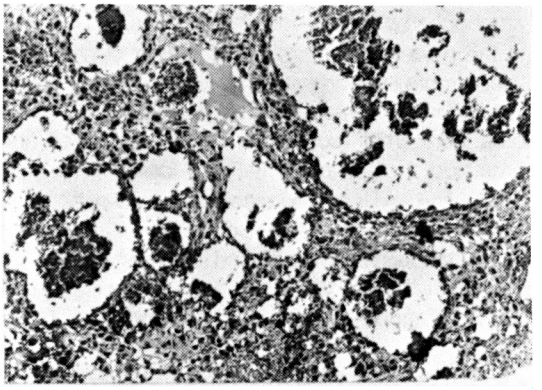

Fig. 3-B. The septa that separate cystic spaces are cellular contained anaplastic stromal cells and benign osteoclastic cells. The cystic spaces are lined by anaplastic stromal cells and occasionally single layer of endothelial cells (H&E ×400).

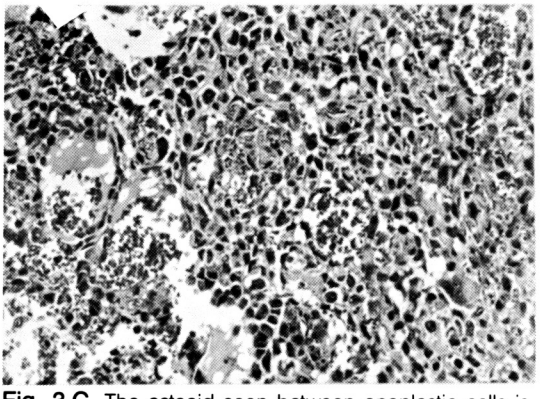

Fig. 3-C. The osteoid seen between anaplastic cells is scanty, fine and lacelike (H&E ×400).

It consisted of cystic cavities filled with clotted blood (Fig. 3-A). The tumor broke through the cortex and bulged into the soft tissue. These changes were widespread and evident in the distal epiphysis as well as in the proximal metaphysis of the tibia, but not extended into the proximal epiphysis. There were no solid mass in the tumor. Microscopic examination showed large blood-filled spaces separated by the thin septa. The cystic spaces were generally lined by anaplastic stromal cells and occasionally lined with a single layer of endothelial cells (Fig. 3-B). The septa were cellular, contained anaplastic stromal cells and benign osteoclastic giant cells. The anaplastic cells were round to oval or spindle, and showed variation in size and shape. The nuclei were large and hyperchromatic, and placed eccentrically in the abundant

amphophilic cytoplasm. There was a marked increase in the nucleo-cytoplasmic ratio. Some of cells contained more than one nucleus. Mitotic figures, including abnormal forms were abundant. The benign giant cells were small (containing five or six nuclei) and simulated thoses found in giant cell tumors. The scanty, fine and lace-like osteoid was present between malignant cells (Fig. 3-C). Both edges of the lesion where the periosteum appeared intact showed permeation of malignant cells in the bone marrow between bony trabeculae with bone destruction.

DISCUSSION

The present case gave us a very difficult problem in differentiating it from the aneurysmal bone cyst because of prominant blood-filled cystic formation. It qave an appearance similar to that of aneurysmal bone cyst, radiologically and pathologically. Although this case mimics aneurysmal bone cyst the obvious anaplasia of the stromal cells within the septa could not be seen in aneurysmal bone cyst. The high degree of cytologic anaplasia may be one of the distinguishing features of the telangiectatic osteosarcoma. Some conventional osteosarcomas have cystic areas simulating telangiectatic osteosarcoma. These should not be classified as telangiectatic osteosarcoma. The criteria for the diagnosis of telangiectatic osteosarcoma are rigid (Matsuno et al., 1976). They are: 1) the roentgenogram shows a lytic, destructive lesion with no appreciable areas of sclerosis; 2) the lesion presents grossly as a cavity with little solid tumor tissue and no areas of sclerosis; and 3) the tumor histologically consists of single or multiple cystic cavities that contain blood or necrotic tumor and are often traversed by septa composed of anaplastic appearing cells. The roentgenographic, gross and microscopic features of the present case fit for these diagnostic criteria of telangiectatic osteosarcoma. Unless the cyst walls are examined closely for anaplastic osteoid producing stromal cells, a diagnosis of aneurysmal bone cyst is possible. The histological features of the telangiectatic osteogenic sarcoma, especially in a limited biopsy specimen may not be always diagnostic. According to the series of 124 cases of Huvos et al. (1982) telan giectatic osteosarcoma showed that age and sex distribution, clinical sign or symptoms and survival rate were not much different from those of conventional osteogenic sarcoma. Howevet, recent reviews state that some clinical findings are different from those of conventional osteogenic sarcoma (Jaffe, 1958; Dahlin, 1967; Spjut et al., 1970). Although similar to osteosarcoma in localization, telangiectatic osteosarcoma has a peculiar tendency to extend into the epiphysis and up to the articular cartilage in patients with closed epiphysis. Another characteristic is the frequency of pathologic fracture that is seen in 29% of patierts with telangiectatic osteosarcoma but in 12% of patients with conventional osteosarcoma (Huvos et al. 1982). The fracture may be caused by the lack of solid areas in the tumor and massive destruction. These clinical features were also seen in this case. Microscopically typical areas of aneurysmal bone cyst may be seen in the peripheral portion of an otherwise typical telangiectatic osteogenic sarcoma (Ruiter et al., 1977). This finding makes one suspect, but does not prove, that the sarcoma arose by spontaneous transformation of a preexistent benign aneurysmal process (Jacobson, 1969). On the other hand, one could also postulate that the aneurysmal bone cyst secondarily became engrafted on the osteosarcoma. The presence of lace-like osteoid in the septa is against the former and the presence of small lesion with no osteoid and the relafive short history are against the latter.

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