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늑골에서 발생한 혈관확장골육종

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혈관확장골육종은 골육종의 드문 아형이다. 늑골은 일반적인 골육종뿐만 아니라 혈관확장골육종에서도 흔치 않은 발생부위이다. 이 종양은 경화된 부분이 없이 혈액이나 괴사된 종양조직을 포함하는 단일성 또는 다발성 낭성 공간으로 구성되므로 다른 양성 낭성 병변과의 감별을 위해 주의 깊은 관찰이 필요하다. 그러므로 늑골이 혈관확장골육종의 호발 부위는 아니지만 일차적인 감별진단에 혈관확장골육종을 포함시키는 것이 중요하다. 늑골에서 발생한 혈관확장골육종과 일차성 골육종에 관한 기존 보고, 혈관성골육종의 감별진단과 드문 부위에 발생한 골육종의 임상적 특징에 관한 문헌고찰과 함께, 늑골에서 발생한 혈관확장골육종 1예를 보고하는 바이다.

색인 단어: 혈관확장골육종, 늑골

Telangiectatic osteosarcoma is an uncommon histopathologic subtype that represents less than 5% of all osteosarcomas[1]. Telangiectatic osteosarcoma is a type of osteosarcoma that resembles aneurysmal bone cyst radiographically and histologically[2]. Typical osteosarcomas may contain areas of telangiectatic changes, however, the diagnosis of telangiectatic osteosarcoma should be confined to pure, lytic lesions. Large portions of telangiectatic osteosarcomas are cystic and lytic lesions containing hemorrhage and necrosis rather than diagnostic anaplastic cells. An important differential diagnosis, the aneurysmal bone cyst, is more common than this tumor. Small foci of malignancy might be missed by a pathologist, especially in a biopsied specimen and when this high grade osteosarcoma is obtained from a rare location. We report a case of telangiectatic osteosarcoma involving the left 7th rib in a 36-year-old man. This is an unusual presentation of a telangiectatic osteosarcoma.

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Case Report

A 36 year-old male complained of left sided chest pain and swelling beginning 10 months ago. Physical examination revealed an ill-defined mass in the left anterolateral chest wall. Alkaline phosphatase was increased to more than 4000 IU/L. Chest radiograph showed a well-defined, about 10cm in diameter, soft tissue mass in left lower thorax with destruction of the anterior arc of the left 7th rib (Fig. 1A). The chest computed tomography (CT) also showed the well-circumscribed, soft tissue mass on the left 7th rib. The anterior arc of the rib was destroyed, and the mass showed heterogeneous enhancement with multifocal cystic portions (Fig. 1B). On anterolateral thoracotomy, a 10×6×6 cm sized firm mass originating from the 7th rib without invasion into the adjacent rib, lung and diaphragm was seen. En bloc resection of the mass along with the 6th and 8th ribs, and serratus anterior muscle was performed. The extent of the en bloc resection included the anterior half of left 6th and 8th ribs and intercostal and serratus anterior muscle of the area. Pathologic examination revealed a well circumscribed mass, measuring 10.5×5.6×6 cm. On sectioning, the mass showed destructed rib with hemorrhagic, cystic and necrotic

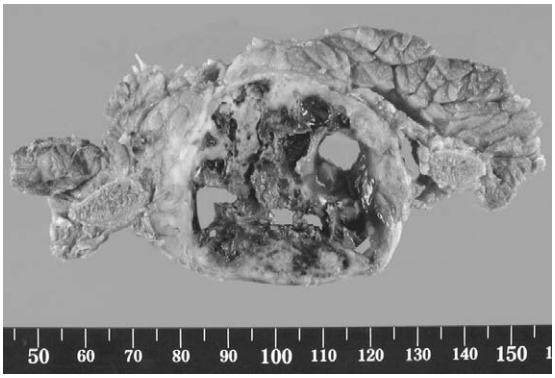

Fig. 2. The cut surface of the mass showed destructed rib with hemorrhagic, cystic and necrotic surface.

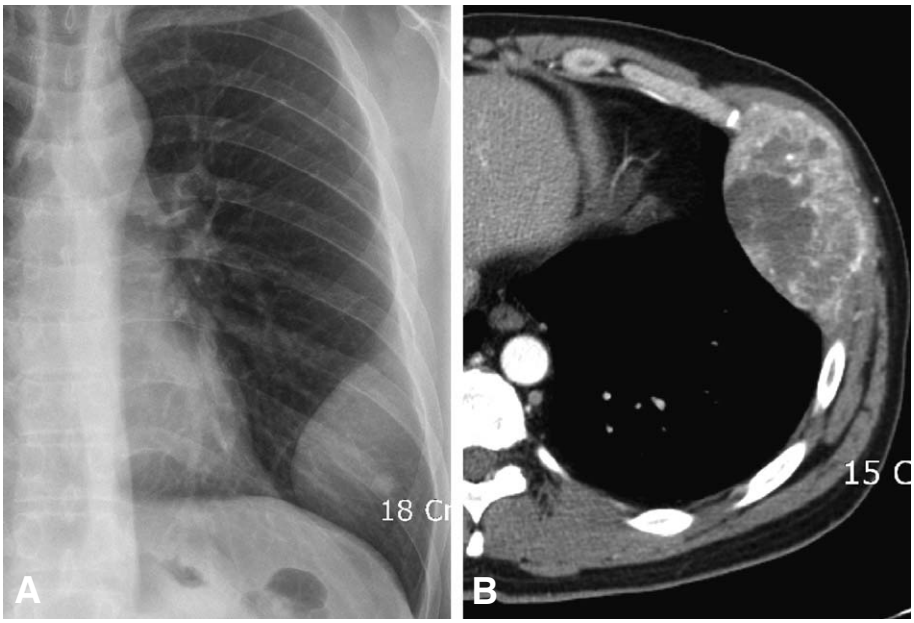

Fig. 1. (**A**) Frontal chest radiograph reveals a well-circumscribed, soft tissue mass in the left lower thorax, (**B**) Chest CT shows soft tissue mass with bone destruction of the left 7th rib. The mass showed multiple cystic areas and heterogeneous enhancement.

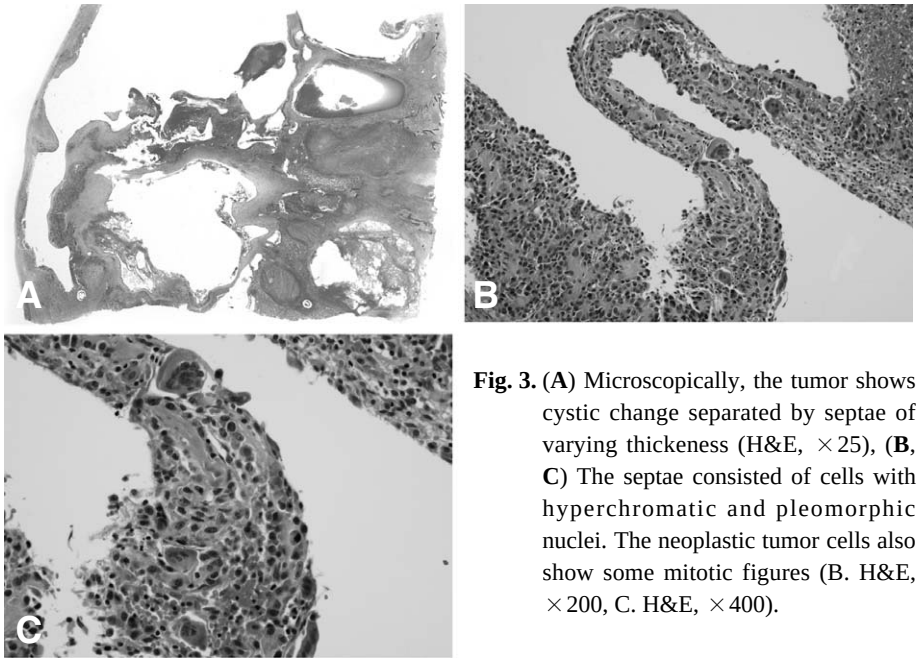

Fig. 3. (**A**) Microscopically, the tumor shows cystic change separated by septae of varying thickeness (H&E, ×25), (**B**, **C**) The septae consisted of cells with hyperchromatic and pleomorphic nuclei. The neoplastic tumor cells also show some mitotic figures (B. H&E, ×200, C. H&E, ×400).

surface (Fig. 2). Microscopic findings disclosed septae of varing thickness separating irregular cystic structures which are filled with blood clots (Fig. 3A). Osteoid formation with little mineralization was seen. The septae were composed of malignant pleomorphic cells with some atypical mitotic figures (Figs. 3B, C). The patient was given adjuvant chemotherapy with high-dose methotrexate. He is currently alive and free of the disease 26 months after the diagnosis and initiation of therapy.

Discussion

Telangiectatic osteosarcoma is a rare variant of osteosarcoma, representing less than 3% of all osteosarcomas[1]. Most of these lesions have been described in the distal femur, proximal tibia and proximal humerus, with other sites being rare[3]. Among 605 cases of osteosarcomas, the cases arising in ribs were only 18 cases (0.03%)[4]. Farr et al[3]. and Murphey et al[5]. reported twenty-eight and forty cases of telangiectatic osteosarcomas, respectively. However, rib involvement was not present in their study.

Radiologically, geographic bone destruction and expansile remodeling of the involved bone is common in plain X-ray films of telangiectatic osteosarcomas. However, for cases with a chest wall mass, chest radiograph has limitations. There is suboptimal visualization of any calcifications within the mass, rib destruction, and periosteal reaction in a plain chest film which are otherwise readily visible in the plain film of a lesion presenting in the long bones. CT provides excellent assessment for the characteristic features of primary osteosarcoma and other common lesions[6]. CT is necessary for proper approach to the diagnosis, especially in chest wall masses.

Ewing sarcoma, chondrosarcoma and osteosarcoma including benign lesions such as fibrous dysplasia and aneurysmal bone cyst are radiologic differential diagnoses to consider in a primary tumor of the rib[7,8].

Ewing sarcomas occur in younger (mean age 16 yrs) patients with systemic symptoms like fever and malaise. More extensive mass with bone involvement and no matrix calcification are noted in imaging studies of these tumors. Most chondrosarcomas originate from the costochondral junction and show chondroid matrix calcification. In contrast, the characteristic radiologic images of telangiectatic osteosarcomas reflect the largely hemorrhagic and/or necrotic composition of most of the lesion with frequent finding of fluid levels. However, after enhancement, nodular viable sarcomatous cells appear as a solid thick nodular enhancing rim of tissue that contains subtle mineralization.

Among many differential diagnoses of telangiectatic osteosarcoma arising in a rib, the aneurysmal bone cyst is the most dangerous pitfall in bone pathology and radiology. Aneurysmal bone cysts have only thin rims with well defined inner margins and septae surrounding cystic spaces[5]. These are differentiated from telangiectatic osteosarcomas which show aggressive growth with non-encapsulated soft tissue components.

Aneurysmal bone cyst is also included in the histologic differential diagnosis of telangiectatic osteosarcoma. The differentiation of a telangiectatic osteosarcoma from an aneurysmal bone cyst must ultimately be made histologically by the presence of obviously anaplastic cells with frequent atypical mitotic figures in the trabeculae of the former[2]. Another histologic differential diagnostic point to consider is a secondary aneurysmal bone cyst. Secondary aneurysmal bone cysts are seen mostly in association with osteoblastomas, giant-cell tumors and osteosarcomas[9]. Particularly, osteosarcomas with secondary aneurysmal bone cysts look very similar to telangiectatic osteosarcomas. Using the same diagnostic criteria suggested earlier, areas of secondary aneurysmal bone cysts have stroma showing benign cells, as opposed to the presence of malignant cells in the fibrous septae of telangiectatic osteosarcomas.

The prognosis of telangiectatic osteosarcoma has engendered a lot of controversy. In 1976, before the institution of neoadjuvant chemotherapy, Matsuno et al[10]. reported that the prognosis for patient with telangiectatic osteosarcoma is dismal compared to those with conventional osteosarcoma. On the other hand, Huvos et al[11]. reported that there was no difference in the prognosis between patients with conventional and telangiectatic osteosarcoma. The reason for the argument may be due to the differences in the diagnostic criteria used by these researchers. Lately, with the advent of chemotherapy, the prognosis for patients with telangiectatic osteosarcoma is as good as or even better than those with conventional osteosarcoma[12].

Duffaud et al[13]. studied clinical features and treatment outcomes of osteosarcomas involving the flat bones including the jaw bone, pelvic bone and rib. Osteosarcomas of the flat bones apparently affect an older age group compared to conventional osteosarcomas of the extremity with more male patients being affected. Patient's age, gender, site of the primary tumor and type of resection did not affect survival. But, the extent of the disease at diagnosis and the adequacy of local control had a significant impact on survival. In contrast to osteosarcomas of the extremities, local treatment failure is very common in this type of tumor involving the flat bones because of functional and cosmetic problems. Common failure

sites were the craniofacial region and the pelvis. Our case is a telangiectatic osteosarcoma arising from the rib of a 36 year-old male, fitting the characteristic patient profile previously described. And in our case, an en bloc resection and adjuvant chemotherapy was given. The complete removal of the tumor with adjuvant therapies and no evidence of metastasis at diagnosis suggest a favorable prognosis for the patient. No recurrence is evident till now.

Primary osteosarcoma presenting in the rib is a significant diagnostic challenge, especially with its telangiectatic variant. As we mentioned previously, to rule out a destructive bony lesion like osteosarcoma, though rare in incidence, a CT is recommended for evaluation of a chest wall mass.

In summary, we report a case of telangiectatic osteosarcoma of the rib. The important pathologic and radiologic differential diagnosis of aneurysmal bone cyst and osteosarcoma with secondary aneurysmal bone cyst were discussed. The literature on osteosarcomas arising from flat bones were reviewed.

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Abstract

Telangiectatic Osteosarcoma Arising in a Rib

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Telangiectatic osteosarcoma is a rare variant of osteosarcoma. The rib is an uncommon primary site for conventional osteosarcoma, and even more for telangiectatic osteosarcoma. Because this tumor consists of single to multiple cystic cavities that contain blood or necrotic tumor with no appreciable areas of sclerosis, careful observation is needed to differentiate this malignancy from other benign cystic lesion. Therefore primary differential diagnosis including telangiectatic osteosarcoma is important, although rib is not the predilection site of telangiectatic osteosarcoma. We present a case of telangiectatic osteosarcoma arising in a rib. We reviewed the literature concerning telangiectatic osteosarcomas and primary osteosarcomas arising from the rib. The differential diagnosis of telangiectatic osteosarcoma and clinical features of osteosarcomas arising from unusual locations were discussed.

Key Words: Telangiectatic osteosarcoma, Rib

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