Case Report
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CT and Positron Emission Tomography/CT Findings of Mediastinal Extraskeletal Ewing's Sarcoma with Extensive Distant Metastasis: A Case Report

광범위한 원격 전이를 보인 종격동 골격외 유잉씨 육종의 전산화단층촬영과 양전자방출전산화단층촬영소견: 증례 보고

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Extraskeletal Ewing's sarcoma (EES) is a rare aggressive malignant tumor, usually occurring in the chest wall of children and young adults. Although EES shows aggressive growth pattern, it seldom demonstrates distant metastasis at diagnosis. Herein, we present computed tomography (CT) and positron emission tomography computed tomography (PET/CT) findings of EES in anterior mediastinum in a 68-year-old man, showing multi-organ distant metastasis at diagnosis. It is another atypical case with unusual presentation in point of old age, mediastinal location and distant metastasis at diagnosis, showing PET/CT findings.

Index terms
Extraskeletal Ewing's Sarcoma
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Positron Emission Tomography/CT

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INTRODUCTION

Extraskeletal Ewing's sarcoma (EES) is a rare malignant small round cell tumor of neural crest origin, which is histologically similar to the more common osseous Ewing sarcoma. Different from the osseous counterpart, it shows a wider age presentation, preferentially occurring in children and young adults, younger than 30 years (1, 2). The most frequent sites of occurrence are the chest wall, lower extremities and paravertebral region, but it rarely occurs in the mediastinum. The recently reported CT finding of mediastinal EES is a large, non-calcified mass with heterogeneous enhancement, frequently infiltrating to the adjacent tissues. In spite of its aggressive behavior, distant metastasis is rare (1, 3). Although positron emission tomography (PET)/CT findings of EES have been rarely reported, it has been reported to show relatively weak fluorodeoxyglucose (FDG) uptake, considering the growth pattern (4).

We report a case of a 68-year-old man diagnosed as mediastinal EES with extensive hematogenous and lymph node metastasis by a CT and PET/CT.

CASE REPORT

Institutional Review Board exemption was obtained to perform this case report.

A 68-year-old man was presented with a 3-week history of a painless palpable mass in the left supraclavicular region. Plain chest radiograph showed an elongated multilobulated mass in the left parahilar region, not obscuring the overlying hilar vasculatures. An ovoid homogeneous opacity was seen in the left supraclavicular region.

Contrast-enhanced CT scans of neck and chest were per-

formed. Chest CT revealed a multilobulated mass, about 7 cm in size, in the anterior mediastinum, which was heterogeneously enhanced with internal non-enhancing hypodense area. Fat planes between the mass, aorta and main pulmonary artery were obscured (Fig. 1A). Multiple enlarged nodes with heterogeneous enhancement pattern were also visible in mediastinum, both supraclavicular regions and celiac axis (Fig. 1B). Multiple small hypodense lesions were detected in the liver (Fig. 1C). In PET/CT performed subsequently, the anterior mediastinal mass showed strong FDG uptake [peak standard uptake value (pSUV), 8.0], and lymphadenopathy that was detected by CT showed a strong uptake (range of pSUV 3.6-7.5) (Fig. 1D). In addition, T3, T9, T10, T11, L1 and L5 vertebral bodies and in the pelvic bone, multiple, variable sized and round osteoblastic lesions that were associated with FDG uptake could be observed (Fig. 1E).

The patient subsequently underwent surgical resection of the left supraclavicular mass lesion for pathologic examination. Histopathological examination showed a poorly differentiated malignant tumor, with neuroendocrine differentiation (Fig. 1F). The mass lesion was negative for leukocyte common antigen, cytokeratin, CK20, CK7, CK5/6 and S100. Immunohistochemical evidence of CD99 in Ewing sarcoma, were weakly positive, confirming ESS.

Starting 1 month after neck mass resection, the patient received chemotherapy, which composed of vincristine, doxorubicin, cyclophosphamide and actinomycin D. In follow-up CT, performed after 5 months, the overall tumor size was increased, new lung metastasis was detected, and malignant pericardial effusion and pleural effusion were developed; thus, pericardio-

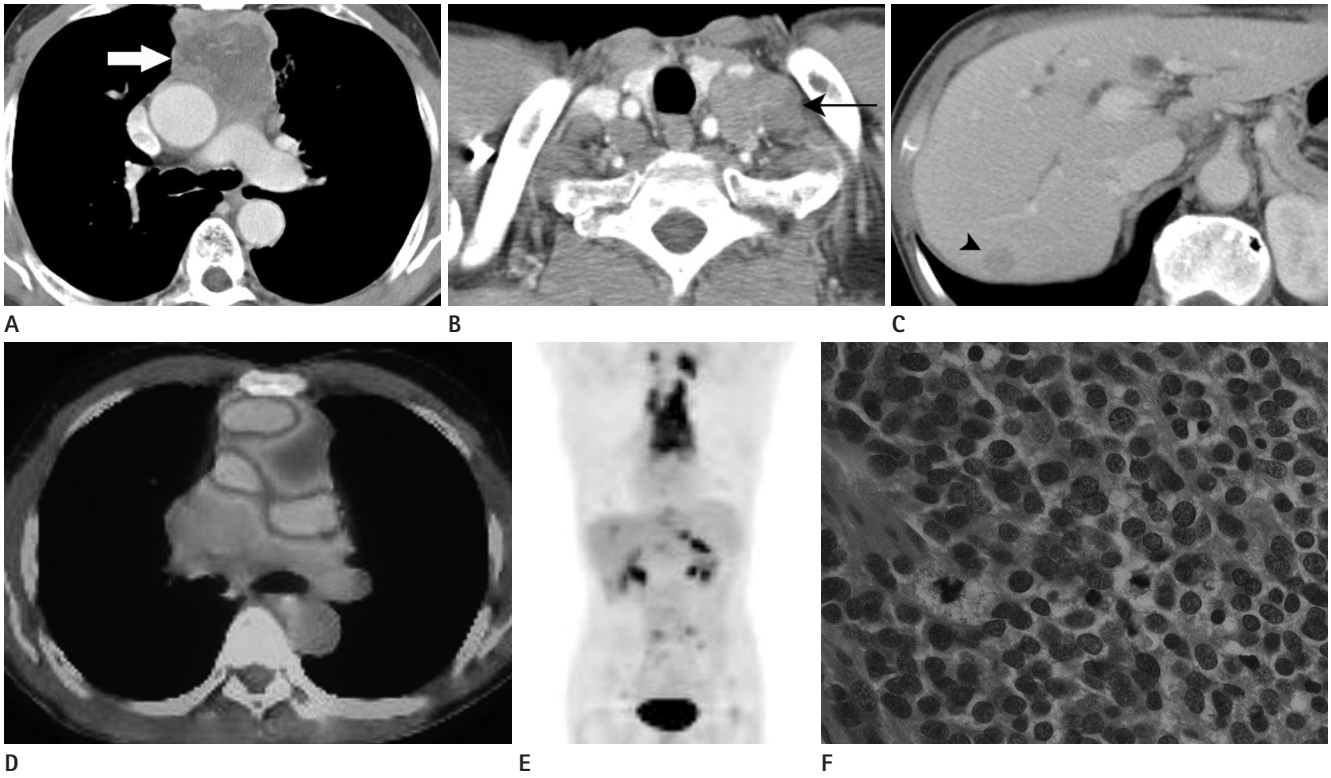

Fig. 1. A 68-year-old man with extraskeletal Ewing's sarcoma in anterior mediastinum.
A. Axial CT scan shows a multilobulated, heterogeneously enhancing mass with extensive necrosis in the anterior mediastinum (arrow). Fat planes between the mass and adjacent vasculatures, aorta and main pulmonary artery, are obliterated.
B. Axial CT scan shows a left supraclavicular lymphadenopathy with homogenous enhancement (thin arrow).
C. Axial CT scan shows a small hypodense nodular lesion (arrowhead) in the liver.
D. Axial PET/CT scan shows an anterior mediastinal mass with peripheral intense FDG uptake and central metabolic defect.
E. Maximum-intensity-projection FDG PET image shows multifocal FDG uptakes in the anterior mediastinum, both supraclavicular regions, liver, retroperitoneal nodes, spines and pelvic bones.
F. Photomicrograph (hematoxylin-eosin, original × 400) shows densely packed sheets of small round neoplastic cells. The nuclei are round with "salt and pepper" chromatin and have inconspicious or small nucleoli. Mitotic figures are common.
Note.—FDG = fluorodeoxyglucose, PET/CT = positron emission tomography/CT

centesis was performed. After 1 month, the patient presented with diplopia and thus brain MRI and spinal tap were performed. It was diagnosed as leptomeningeal metastasis. Afterward, the patient is under conservative care.

DISCUSSION

The classical histopathologic description of EES is small with round blue cells that are uniform in appearance and organized in solid sheets that are divided by fibrous strands. Its cytoplasm is scanty, pale staining and vacuolated because of the presence of glycogen, and the nuclei are round with "salt and pepper" chromatin and small nucleoli. A sensitive and relatively specific antigen, CD99/MIC2 and a characteristic chromosomal translocation, t(11;22), have been identified in skeletal Ewing's sarcoma and EES, which shares histopathologic and immunohistochemical findings with Ewing's sarcoma (5). However, different from Ewing sarcoma, which is developed in the childhood and the early adolescence primarily, EES occurs in young people who are between the ages of 15-30 years with wider age distribution (6, 7). In addition, EES occurs frequently in the chest wall, lower extremities and paravertebral region, and it may occur in the mediastinum, although rare (1, 3).

The CT findings of mediastinal EES have been reported recently. It is shown as a soft tissue mass with unclear boundary and changes of hemorrhage, cystic change and necrosis in many cases, and calcification within tumors has not been reported, until now (1). This is similar to ESS developed in the chest wall that is frequently referred to as Askin tumor. In addition, just like EES of other areas, it shows aggressive growth and infiltrates to the adjacent pleura or the anterior chest wall; thus, pleural effusion or neighboring bone destruction may be associated (1, 2). In our case, it was a large heterogenous mass in the anterior mediastinum on CT, associated with internal necrotic or cystic changes, and there was no tumoral calcification. However, the adjacent sternum was not destructed by the mass.

Ewing family is a high-grade malignancy, so it is expected to show high pSUV in most cases. However, PET/CT findings were hardly reported, and thus, it is difficult to conclude. Nonetheless, Kara Gedik et al. (4) have reported that the average pSUV was 4.54 and it was not as high as anticipated. In our case, the pSUV of EES was 8.0, and it was relatively strong.

Mediastinal EES occasionally shows a local relapse or distant metastasis during treatments or after treatments. However, cases showing distant metastasis at the time of diagnosis are very rare. Differently from Ewing's sarcoma with common metastatic sites in the lung and bone, mediastinal EES metastasizes in the skeleton and liver most frequently (1). At the time of diagnosis, our case had metastatic lymphadenopathy and liver metastasis. Even after treatments, additional metastasis had developed in the bone, leptomeninges, pericardium and pleura.

In regard to treatments, as for EES with distant metastasis, it is better to perform early aggressive combination chemotherapy rather than single agent monotherapy. Further, these tumors are also radiosensitive; tumors are not appropriate to surgical resection or have positive surgical margins, and are treated with radiation (8). In our case, combined chemotherapy consisting of vincristine, doxorubicin, cyclophosphamide and actinomycin D was performed; nonetheless, the outcome was not good.

Although it is generally known that EES shows poor prognosis, a 5-year survival rate is over 60%, if surgeries and appropriate chemotherapies are applied (1, 6). Prognostic factors are age, tumor location, tumor size, with or without metastasis, genetic mutation type and treatment programs (6). Our patient showed disease progression even after the treatments; thus, the prognosis was speculated to be poor. Patient's advanced age, distant metastasis at the time of diagnosis and atypically high pSUV might be associated with such poor prognosis.

In conclusion, mediastinal EES is a tumor that may show diverse imaging findings, clinical manifestation and PET-CT findings. It should be considered in the differential diagnosis of any patient, of any age, with a non-calcified mediastinal mass with malignant feature.

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광범위한 원격 전이를 보인 종격동 골격외 유잉씨 육종의 전산화단층촬영과 양전자방출전산화단층촬영소견: 증례 보고

김봄이 · 고정민 · 박현진

골격외 유잉씨 육종은 드문 악성종양으로, 대개는 소아나 청소년에서 발생하며 흉벽이 가장 흔한 원발장소이다. 비록 이 종양이 공격적으로 자라는 경향을 보이나, 진단 당시부터 원격전이를 보이는 경우는 거의 없다. 이에 우리는 전종격동에서 발생한 골격외 유잉씨 육종이 다장기 원격전이를 보인 68세 남자 환자를 경험하였기에 이를 보고하고자 한다. 이 증례는 환자가 고령인 점, 원발장소가 종격동인 점 그리고 진단당시 광범위한 원격전이를 보인 점에서 비전형적이라 할 수 있으며, 또한 비교적 잘 알려진 전산화단층촬영소견 외에도 거의 알려지지 않은 골격외 유잉씨 육종의 양전자단층촬영소견을 보여주고 있다.

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