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 hiliar vascular. 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” chromatia and have inconspicuous or small nucleoli. Mitotic figures are common.

Note. – FDG = fluorodeoxyglucose; PET/CT = positron emission tomography/CT
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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