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CASE REPORT

Primary Malignant Fibrous Histiocytoma in Mediastinum: Imaging with ^{18}F-FDG PET/CT

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Abstract Malignant fibrous histiocytoma (MFH) is the most common soft tissue tumor which often occurs in the extremities and the retroperitoneum. Primary mediastinal MFH is rare; thus, findings on ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) of mediastinal MFH have not been reported yet. We report herein the case of a 64-year-old man who was presented with a superior mediastinal mass. The mass showed intense ^{18}F-FDG uptake with central metabolic defect on PET/CT. The maximum standardized uptake value was 17.4. After tumor removal via median sternotomy, an MFH of the storiform-pleomorphic type was diagnosed on histopathologic examination. We present the first report of ^{18}F-FDG PET/CT imaging of MFH in the superior mediastinum.

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Introduction

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma in adults. This tumor occurs most commonly in the deep fascia and skeletal muscles of the extremities (about 70%), followed by the retroperitoneum (16%) [1]. Although there have been few reports describing MFH in the extremities using ^{18}F-FDG PET, soft tissue sarcoma including MFH generally presented as a hypermetabolic mass [2, 3]. The mediastinum is an uncommon site for MFH. The first reported case of mediastinal MFH detected by ^{67}Ga scan was described in 1976 [4]. To the best of our knowledge, ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) imaging of MFH in the mediastinum has not been reported, thus we present the ^{18}F-FDG PET/CT finding of a primary mediastinal MFH.

Case Report

A 64-year-old man complaining of chest pain and hoarseness for 2 months was referred to our hospital. He had no significant medical history except for diabetes mellitus. Contrast-enhanced chest CT showed a large lobulated mass with a longitudinal dimension of approximately 6.7 cm in the superior mediastinum, extending into the anterior, middle mediastinum, and the right lower neck. The mass encased the right carotid artery, trachea, and the brachiocephalic vein. The mass showed central necrotic portion and heterogeneous enhancement of the boundary area (Fig. 1). A large hypermetabolic mass with a central metabolic defect

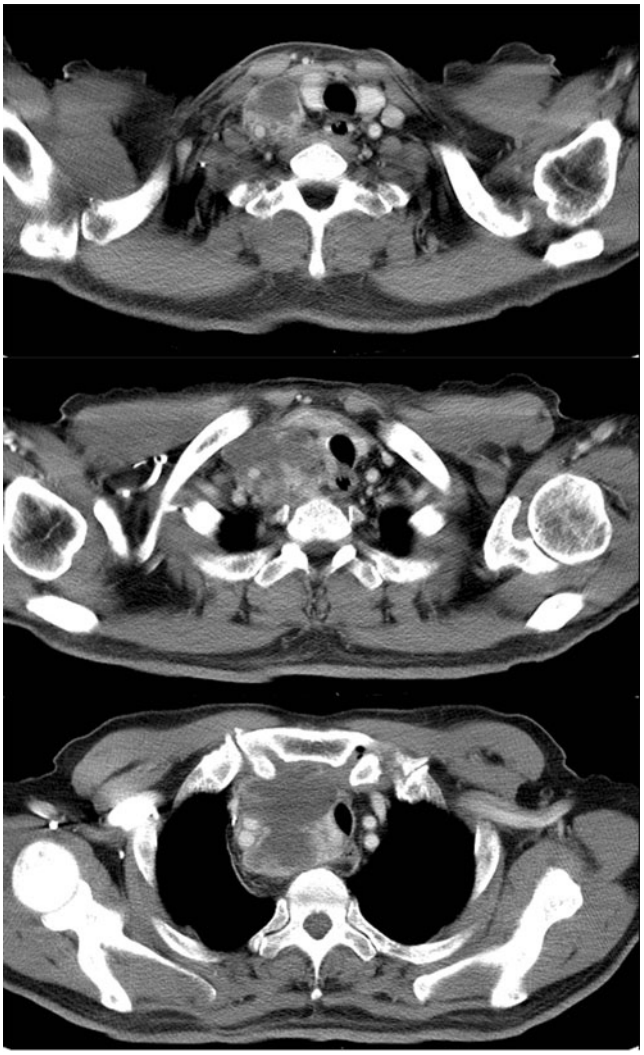

Fig. 1 Contrast-enhanced chest CT images show a large mass involving the superior mediastinum, extending to the right lower neck. The mass contains a central necrotic portion and enhancing lesions in the mass periphery

was demonstrated on ^{18}F-FDG PET/CT (Fig. 2). Maximum standardized uptake value (SUV_{max}) of the mass was 17.4. However, no significant FDG uptake to suggest lymph node or distant metastasis was observed. The excisional biopsy for diagnosis was performed in the right lower anterior neck mass. Histopathologic examination of the specimen revealed a myofibroblastic spindle cell tumor. Finally, tumor removal through median sternotomy was performed, and the final diagnosis was made as a malignant fibrous histiocytoma of the pleomorphic-storiform subtype. Histopathologically, the tumor was composed of pleomorphic spindle-shaped cells arranged in a storiform pattern, and immunohistochemical stainings for vimentin and CD 68 were found positive (Fig. 3). However, immunohistochemical stainings for epithelial membrane antigen, cytokeratin 5/6, smooth muscle antigen, desmin and S100 protein were found negative. Resection margin was involved by the tumor on microscopic examination. The patient died due to tumor recurrence and postoperative complications 2 months after the operation.

Discussion

MFH generally develops in the extremities and the abdominal cavity [1]. However, many other sites, such as the thorax (thymus, mediastinum, chest wall, lung, diaphragm, heart and great vessel), maxillary sinus, bladder, kidney, ovary, anal canal, and the brain, were reported [5–9]. MFH originating in the mediastinum has rarely been reported. According to the literature review of Murakawa et al. [5], 34 MFH cases of the mediastinum have been reported since 1982. The mean age of the patients was 50 years and 62% were male. Two-thirds of the patients had presenting symptoms such as chest pain, back pain, fever, and general malaise. MFH of the mediastinum occurred mostly in the anterior and posterior mediastinum (23 of 34 cases). In the present case, MFH was mainly located in the superior mediastinum, extending to the anterior, middle mediastinum and the lower neck. Most MFHs developed de novo; however, several cases were associated with radiation therapy [10] or previous operation procedures [11]. Our patient had no previous operation and radiation therapy.

CT findings of mediastinal MFH were nonspecific, with heterogeneously enhanced soft tissue mass and a necrotic portion with rarely containing calcifications [5]. Consistent with this finding, a large lobulated mediastinal mass, including a central necrotic portion with enhancement in the peripheral portion of the mass, was observed on contrast-enhanced CT in the present case. ^{18}F-FDG PET/CT demonstrated enhanced glucose metabolism in the peripheral portion of the mass with a central metabolic defect. However, ^{18}F-FDG PET/CT can show a hypermetabolic mediastinal mass-like lesion in other malignancies such as thymic carcinoma, lymphoma, and some benign lesions such as sarcoidosis; thus, it may be challenging to discriminate MFH from other mediastinal neoplasms by the pattern of ^{18}F-FDG uptake [12–14]. According to several reports of MFH developing in the kidney, maxillary sinus, brain and the lung, all MFHs showed high ^{18}F-FDG uptake with a central metabolic defect, which was similar with the present case [6–9]. ^{18}F-FDG-avid lesions in the renal and pulmonary MFH were detected on PET/CT, and their $SUVs_{max}$ were 6.8 and 12.1, respectively [6, 7]. In other cases with MFH in the maxillary sinus and the brain, high ^{18}F-FDG uptake was demonstrated (SUV_{max}=16.9 and 4.0, respectively) [8, 9]. MFH generally manifests

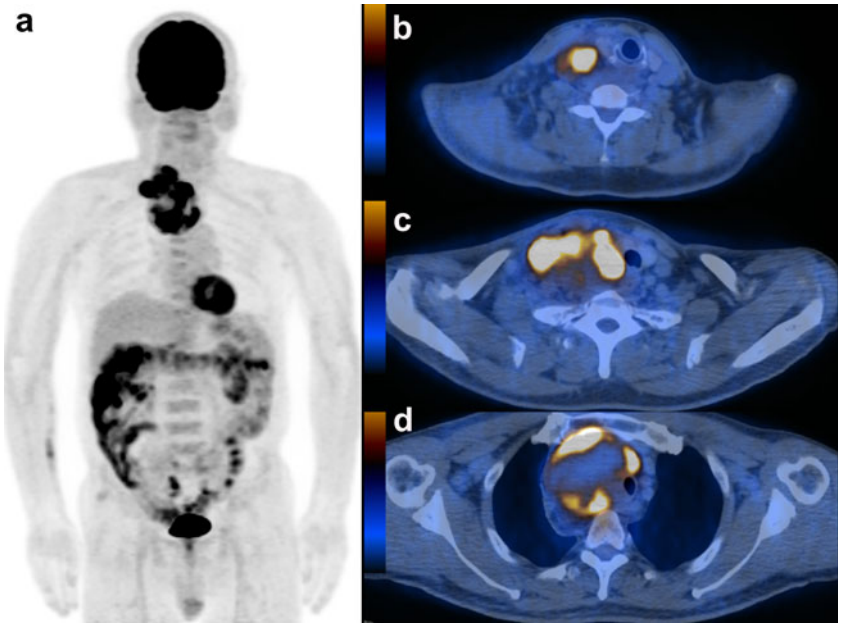

Fig. 2 ^{18}F-FDG PET/CT images reveal a large hypermetabolic mass with a central metabolic defect in the superior mediastinum and the right lower neck (**a** maximum intensity projection image, **b**–**d** transaxial fusion images)

as a bulky, multilobular mass with progressive enlargement over several months. CT scans demonstrate a soft tissue mass that often contains areas of decreased attenuation centrally, which correspond to regions of myxomatous tissue, hemorrhage, or necrosis [15]. In the present case, the central metabolic defect area of the mass on PET/CT corresponded to the hypoattenuated necrotic or hemorrhagic area on CT scan. Histiocytes and fibroblasts in MFH are mainly proliferating cells with increased glucose metabolism, which may contribute to intense ^{18}F-FDG uptake.

Malignant fibrous histiocytoma originates from histiocytes, fibroblasts, or cells with features that are intermediates between fibroblasts and histiocytes [16]. According to various histological features, MFH can be classified into five subtypes: (1) the storiform-pleomorphic type; (2) the myxoid type (myxofibrosarcoma); (3) the inflammatory type; (4) the giant cell type; (5) the angiomatoid type [17]. This case was the storiform-pleomorphic type, the most common subtype of MFH. MFH was shown to frequently express vimentin, CD 68, actin, alpha-1-antitrypsin, and alpha-1-antichymotrypsin. Other antigens such as smooth muscle actin, desmin, cytokeratin, epithelial membrane antigen, and S100 protein were shown to give variable stain in MFH, as reported in previous studies [18, 19]. The immunohistochemical staining of vimentin and CD 68 in the present mediastinal MFH were positively stained. However, other markers for establishing line of differentiation were not stained.

Therapeutic modalities are mainly surgical resection and/or adjuvant radiotherapy [5]. Primary MFH showed a poor response rate according to the report about chemotherapy and radiotherapy [20]. The prognosis of MFH is poor; metastasis to other organs and regional recurrence rate is about 50% and 5-year survival rate is 14% [1]. In our case, mediastinal mass recurred 2 months after the surgical resection and patient died due to the postoperative complications.

In conclusion, large hypermetabolic mass in the superior mediastinum was observed on ^{18}F-FDG PET/CT and primary MFH was confirmed by surgical removal. Although mediastinal MFH occurs rarely, MFH may be included in the differential diagnosis of hypermetabolic mediastinal mass evaluation in elderly patients.

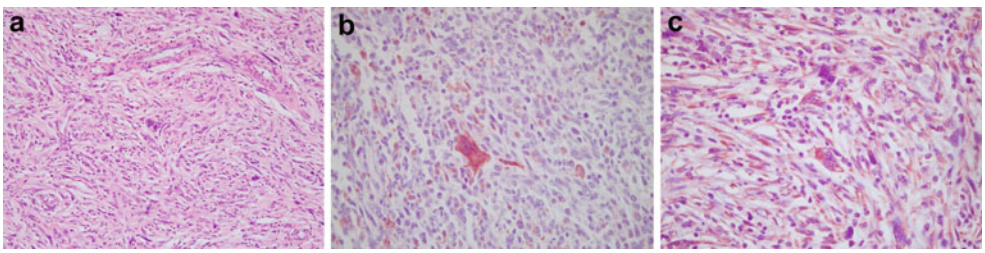

Fig. 3 Microscopic examination shows pleomorphic spindle shaped neoplastic cells forming a storiform pattern (**a**, hematoxylin-eosin stain ×200). Immunohistochemical staining was positive for CD 68 (**b**, ×400) and vimentin (**c**, ×400)

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