

The Characteristics of FDG PET/CT Imaging in Pulmonary Langerhans Cell Sarcoma

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Abstract: A 53-year-old man, who was a heavy smoker, presented with recent severe cough. Radiography demonstrated a large pulmonary mass in the right upper lung. FDG PET/CT demonstrated heterogeneous high-grade activity in the pulmonary mass located in the right upper lung (standardized uptake value of 20), with central necrosis, bilateral upper mediastinal lymphadenopathy, right supraclavicular lymphadenopathy, direct left sternal manubrium invasion, and distal bilateral peripheral lung metastasis. Histology revealed significant malignant cytologic features and CD1a- and S-100-positive cells by immunohistochemistry staining, typical for Langerhans cell sarcoma.

Key Words: pulmonary, langerhans cell sarcoma, FDG, PET, CT

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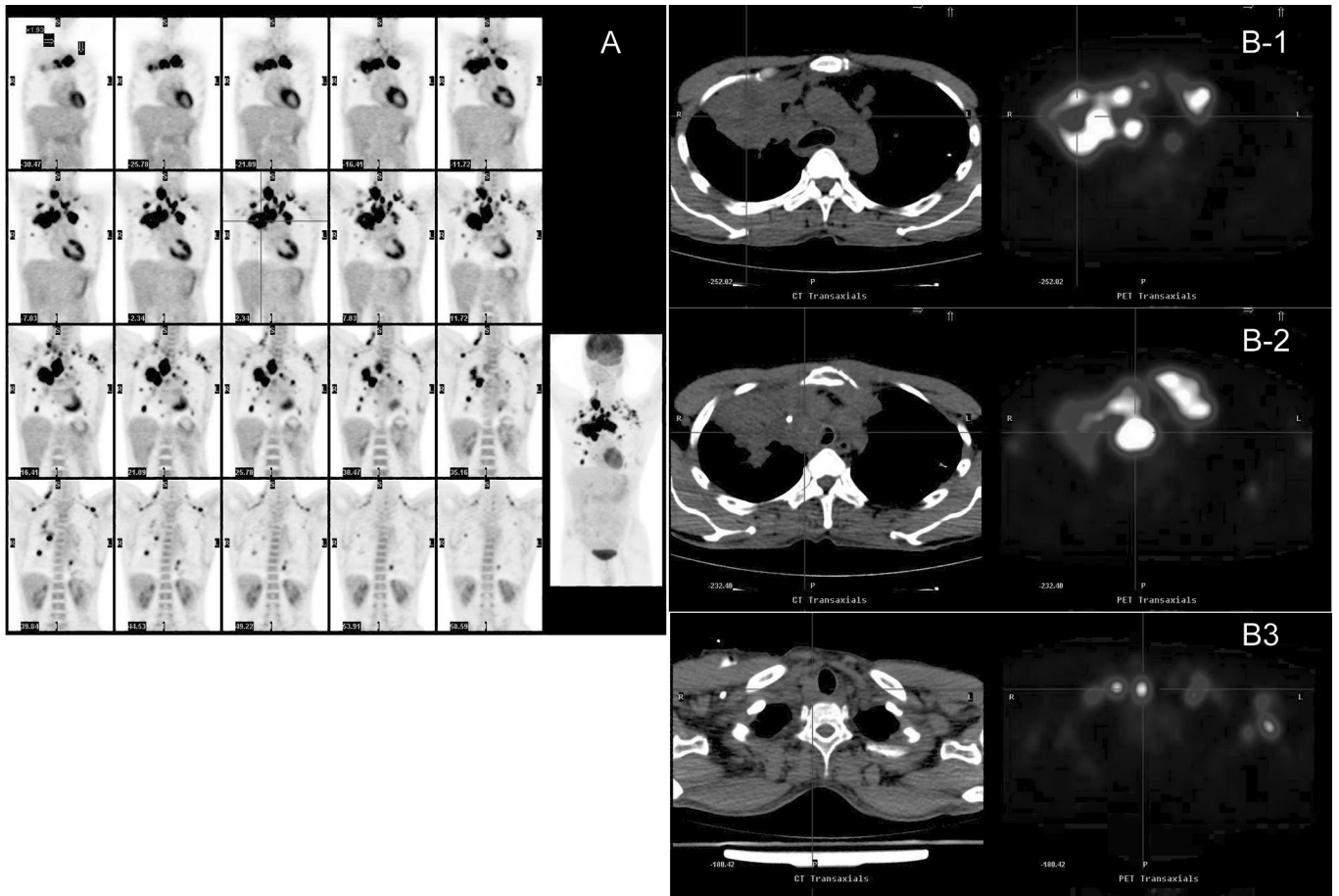


FIGURE 1. A, Coronal views of FDG PET imaging: Based on sequential imaging, it is shown that multiple high-grade FDG-avid lesions distribute above the diaphragm. A large extremely high-grade FDG-avid pulmonary mass (standardized uptake value of 20) with central necrosis is found in the anterior right upper lobe (as marker). The upper mediastinum fills with conglomerated high-grade FDG-avid masses (lymphadenopathy). Focally high grade of right supraclavicle and several discrete axillary lymph nodal activities are simultaneously seen beyond thorax. Multiple roundish peripheral pulmonary nodules with low-grade FDG-avidity scatter in bilateral lung fields. B, Transverse section of FDG PET/CT imaging: Depending on CT localization, 3 major sections of fusion imaging of FDG PET/CT also demonstrate aggressive behavior of Langerhans cell sarcoma.

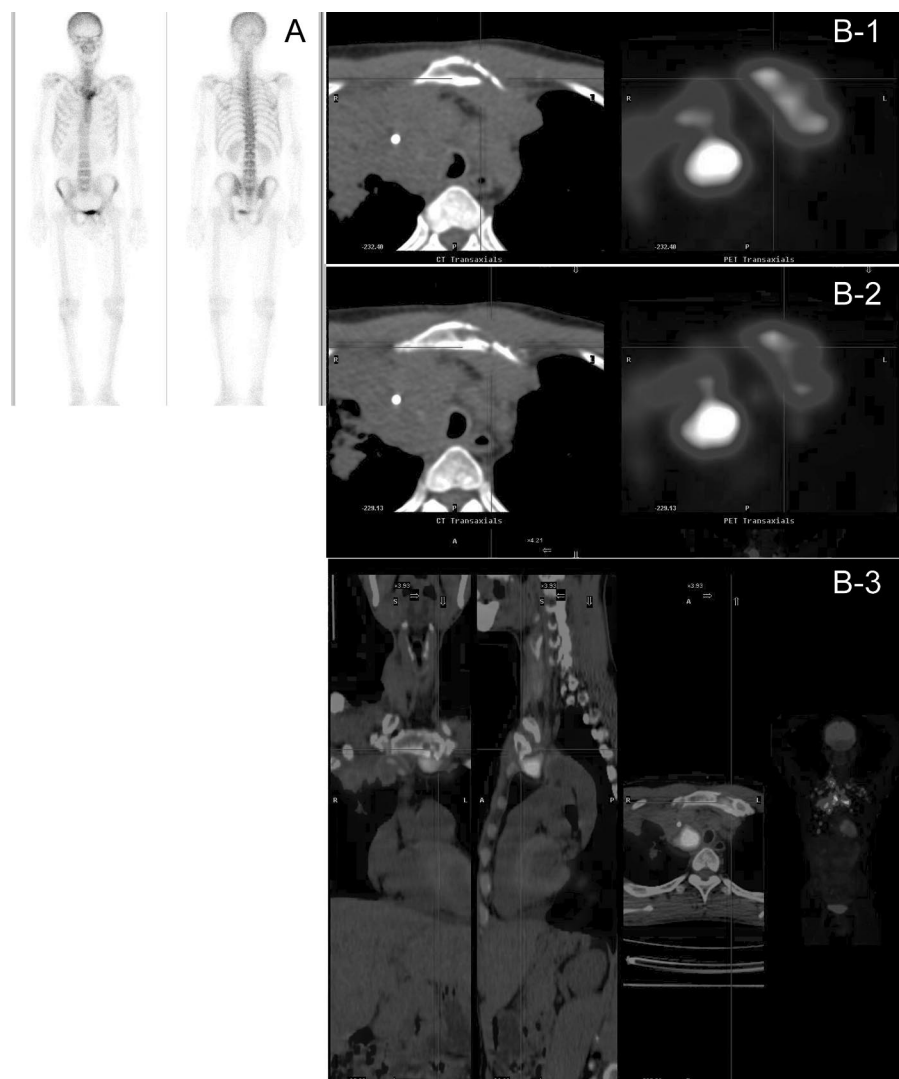


FIGURE 2. A, ^{99m}Tc MDP whole-body bone scan: Regionally increased activity in left sternal manubrium corner is only seen on whole-body imaging. **B,** Transverse section of FDG PET/CT imaging: We can find a tiny linear cortical defect of inner side of left sternum on CT and high FDG-avid sarcoma cellular activity from left anterior mediastinal lymphadenopathy infiltrating into adjacent marrow cavity of left sternum on sequential FDG PET/CT imaging. Therefore, the destruction of sternum is not traditionally the result of hematologic spreading. This fusion imaging of FDG PET/CT demonstrates and localizes unusually detailed characteristics of Langerhans cell sarcoma locally spreading into adjacent left sternum via periosteum. Neoplasms of dendritic or histiocytic cells are extremely rare.¹ Based on literature review, the first case of Langerhans cell sarcoma was published by Wood et al in 1984.² Only 14 cases of Langerhans cell sarcoma have been reported.^{2–10} Langerhans cell sarcoma usually shows multiple organ involvement, including lymph nodes, skin, pulmonary, liver, spleen, and bone. Similar to the last case report published in 2006,⁹ we present another case of pulmonary Langerhans cell sarcoma based on FDG PET/CT findings. The World Health Organization classification of histiocytic and dendritic cell neoplasms differentiates between histiocytic sarcoma, Langerhans cell histiocytosis, Langerhans cell sarcoma, interdigitating dendritic cell sarcoma/tumor, follicular dendritic cell sarcoma/tumor, and dendritic cell sarcoma, not otherwise specified.¹¹ To distinguish the different types, a combination of morphologic and immunophenotypic characterization is necessary. Langerhans cells are positive for CD1a and S-100 protein; negative for CD21, CD35, and CD68; and show Birbeck granules ultrastructurally. In our patient, immunohistochemistry stain is consistent with positive for CD1a and S-100 protein. The first imaging of FDG PET/CT on Langerhans cell sarcoma demonstrates the high FDG-avid aggressive behavior of this sarcoma.¹² In the imaging, distal hematogeneous spreading into bilateral pulmonary fields is demonstrated. However, there is 1 lesion in left hemisternum seen on ^{99m}Tc MDP bone scan. Based on FDG PET/CT, focally increased uptake is because of direct invasion of sarcoma via periosteum effect, and not because of hematologic spreading. In recent literature review, periosteum owns mesenchymal stem cell.^{13–15} We propose a certain mechanism of sarcoma direct invasion of bony structure via periosteum.^{16,17}