^{18}F-FDG PET/CT characterization and response evaluation in a case of lymphomatoid granulomatosis involving the main pulmonary artery

Hell J Nucl Med 2024; 27(1): 71-72 *Epub ahead of print: 18 April 2024* *Published online: 30 April 2024*

Abstract

Main pulmonary artery (MPA) involvement of lymphomatoid granulomatosis (LYG) is extremely rare. We described fluorine-18-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography/computed tomography (PET/CT) findings in a case with LYG originated from the MPA. Fluorine-18-FDG PET/CT demonstrated nodular hypermetabolic foci in the MPA, corresponding well to the intraluminal filling defects on CT pulmonary angiography, and the secondary right heart dysfunction was observed. Final diagnosis was made after transcatheter MPA biopsies and multi-disciplinary consultation. The patient recovered completely following the steroid therapy and MPA stenting, which was illustrated on the second ^{18}F-FDG PET/CT.

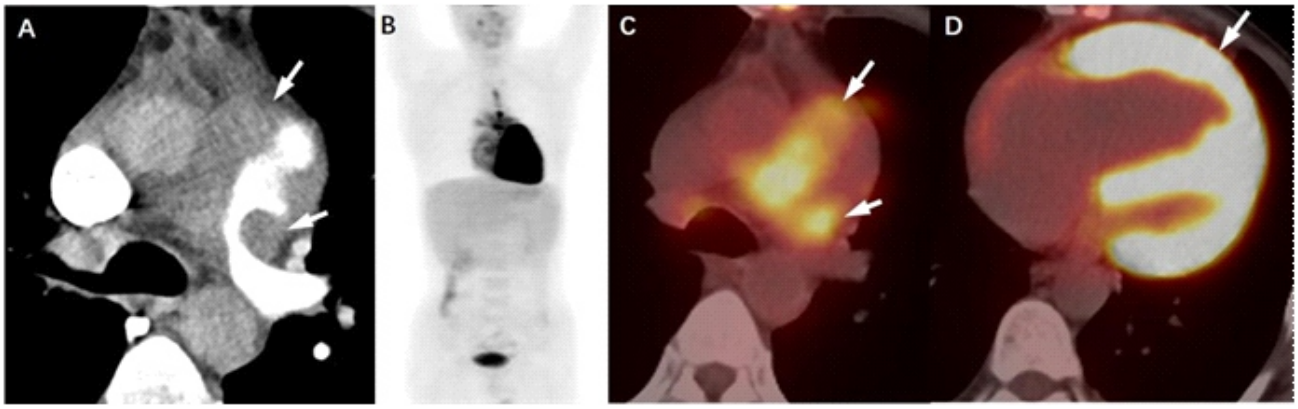

Figure 1. A 46-year-old man presented with a 6-month history of exertional dyspnea. Computed tomography pulmonary angiography (CTPA) manifested multiple filling defects in the main pulmonary artery (MPA), the MPA wall was markedly thickened and the lumen was narrowed (A). The patient underwent a routine whole body ^{18}F-FDG PET/CT to further evaluate and stage the MPA lesions. While the MPA lesions revealed nodular enhanced glucose metabolism on ^{18}F-FDG PET/CT, with maximum standardized uptake value (SUVmax) 5.21. Furthermore, the right cardiac enlargement and the right ventricle with a fierce ^{18}F-FDG activity mimicking the left ventricle were observed, which indicated pulmonary hypertension and right heart dysfunction (B-D). Based on the CTPA and PET findings, neoplastic disease rather than pulmonary embolism (PE) was suspected, such as lymphoma or sarcoma. The diagnosis of low-grade of lymphomatoid granulomatosis (LYG) involving MPA was made after the transcatheter MPA biopsies and multi-disciplinary consultation.

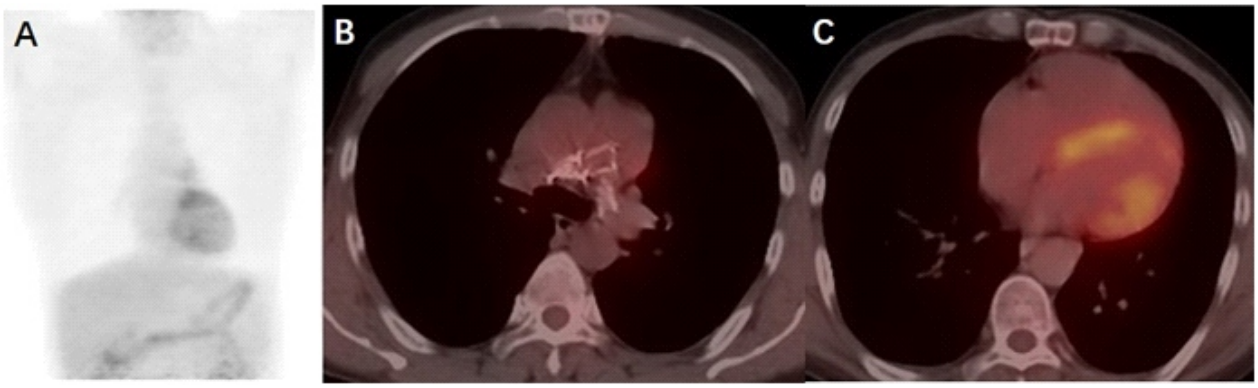

Figure 2. The patient recovered 2 years later after receiving the steroid treatment and a MPA stent implantation. Thoracic ^{18}F-FDG PET/CT showed the original hypermetabolic foci in the MPA disappeared completely and the secondary cardiac changes also restored (A-C).

Lymphomatoid granulomatosis (LYG) is a rare Epstein-Barr virus (EBV)-driven B-cell lymphoproliferative disease that was classified as a subtype of mature B-cell lymphoma in the 2016 revision of WHO Classification of Haematolymphoid neoplasms. It preferentially involves the lungs, skin, central nervous system, liver and kidneys [1-3]. Pulmonary artery involvement is extremely rare [4]. Pathologically, LYG is divided into three grades on the basis of the proportion of large atypical EBV+ B-cells, the angioinvasive/angiodestructive reactive lymphocyte infiltrate and varying degree of necrosis. Treatment varied according to the grade, stage, and patient's immunosuppression status. Low-grade disease requires immune-modulation such as careful follow-up, interferon alpha-2b (IFN-a), steroid alone, or less aggressive chemotherapy, while high-grade disease is treated like diffuse large B-cell lymphoma with immuno-chemotherapy [1, 3, 5]. It is hard to distinguish neoplasm from PE by non-specific radiological findings. Fluorine-18-FDG activity could reflect the degree of inflammatory cell infiltration, thus LYG often shows increasing ^{18}F-FDG uptake [2, 6-8]. Fluorine-18-FDG PET/CT was not only used to rule out PE, but also to guide the biopsy, assist with the treatment by staging and evaluate the therapy response in this case. The differential diagnosis of LYG involving MPA includes septic PE, granulomatosis, tumor embolus, sarcoma, fibrous tumor, angiomatoid fibrous histiocytoma, and so on [9-11].

In conclusion, ^{18}F-FDG PET/CT is more effective in the pre- and post-treatment evaluating of disease activity with MPA involvement of LYG compared to the traditional imaging.

Bibliography

1. Melani C, Jaffe ES, Wilson WH. Pathobiology and treatment of lymphomatoid granulomatosis; a rare EBV-driven disorder. *Blood* 2020; 135(16): 1344-52.
2. Higashihara M, Ishibashi K, Tokumaru AM et al. Brain PET imaging of ^{11}C-Methionine, ^{18}F-FDG, and ^{18}F-THK5351 in a case of lymphomatoid granulomatosis. *Clin Nucl Med* 2022; 47(12): e749-e51.
3. Sanguedolce F, Zanelli M, Zizzo M et al. Primary pulmonary B-cell lymphoma: a review and update. *Cancers* 2021; 13: 415.
4. Su H, Wei R, Duan Y et al. A rare case of severe right heart failure due to pulmonary artery lymphomatoid granulomatosis. *ESC Heart Fail* 2020; 7(6): 4343-7.
5. Aiko N, Sekine A, Umeda S et al. The spontaneous regression of grade 3 methotrexate-related lymphomatoid granulomatosis: a case report and literature review. *Intern Med* 2018; 57(21): 3163-7.
6. Roarke MC, Nguyen BD. PET/CT characterization and monitoring of disease activity in lymphomatoid granulomatosis. *Clin Nucl Med* 2007; 32(3): 258-9.
7. Kesim S, Şen F, Özgüven S et al. ^{18}F-FDG PET/CT imaging of a grade 3 lymphomatoid granulomatosis in an immunocompromised pediatric patient. *Mol Imaging Radionucl Ther* 2023; 32(1): 68-70.
8. Chung JH, Wu CC, Gilman MD et al. Lymphomatoid granulomatosis: CT and ^{18}F-FDG-PET findings. *Korean J Radiol* 2011; 12(6): 671-8.
9. Haug LP, Zarka MA, D'Cunha J et al. Pulmonary artery angiomatoid fibrous histiocytoma mimicking a large pulmonary embolism. *Clin Nucl Med* 2022; 47(12): e738-e9.
10. Ren J, Li H, Zhang Q et al. Clinical utility of ^{18}F-FDG PET/CT imaging in patients with pulmonary artery sarcoma. *EJNMMI Res* 2022; 12(1): 18.
11. Méndez-Echevarria A, Coronado-Poggio M, Baquero-Artigao F et al. Septic pulmonary emboli detected by ^{18}F-FDG PET/CT in children with S. aureus catheter-related bacteremia. *Infection* 2017; 45(5): 691-6.

Financial support

This study was supported by Natural Science Foundation of Gansu Province (21JR7RA633).

Lanlan Cui[1*] MM, Zhimin Wang[1*] MD, Weidong Hu[2] BS, Huichun Wang[1] MM

1. PET/CT Center, Gansu Provincial Hospital, Lanzhou, Gansu, China, 2. Department of Respiratory Medicine, Gansu Provincial Hospital, Lanzhou, Gansu, China.

[#] Lanlan Cui and Zhimin Wang contributed equally to the work.

Corresponding author: Huichun Wang MM, PET/CT Center, Gansu Provincial Hospital, No. 204 Donggang West Road, Lanzhou, 730000, Gansu, China. E-mail: gssypetct@163.com