Clinical Case Report

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18F-FDG PET/CT imaging of relapsing polychondritis

A case report

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Abstract

Background: Relapsing polychondritis (RP) is an uncommon autoimmune inflammatory disease that may affect cartilage throughout the body.

Case report: We report on a case of fever of unknown origin in which 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) was performed to make a diagnosis of RP.

Conclusion: Our case demonstrates that the use of 18F-FDG PET/CT is a useful diagnostic tool to accurately determine the extent of inflammation throughout the body which can be identified by an increased 18F-FDG uptake.

Abbreviations: 18F-FDG PET/CT = 18F-fluorodeoxyglucose positron emission tomography/computed tomography, ESR = erythrocyte sedimentation, MDP = Tc-99m methylene diphosphonate, MIP = maximum intensity projection, RP = relapsing polychondritis.

Keywords: 18F-FDG, PET/CT, Relapsing polychondritis

1. Introduction

Relapsing polychondritis (RP) is an uncommon autoimmune disorder that is characterized by recurrent inflammation and destruction of the cartilaginous tissue in various sites of the body with high risk of misdiagnosis.[1] Auricular and nasal cartilage is usually the first to be affected at the onset of the disease. Airway involvement is present in up to 50% of patients with RP and is a major cause of morbidity and mortality.[2,3] The disease has an intermittent characteristic. The clinical course of RP is irregular. No specific laboratory methods or specific histologic findings are considered pathognomonic for RP. We describe a case of RP imaged on 18F-FDG PET/CT.

2. Case report

A 67-year-old man was admitted to hospital for presented with a 2-month history of persistent cough with sputum and 10-day history of low-grade fever. Additionally, the patient's auricles swelled rapidly before admission, and the left one is markedly enlarged. Physical examination revealed coarse crackles in both lungs and swollen auricles. A chest CT scan showed bilateral pneumonia and pulmonary emphysema. The white-cell count was 4.97×109/L with 75.7% neutrophils. The erythrocyte sedimentation (ESR) was 58 mm per hour. Serum tumor markers were unremarkable. Diagnostic antibacterial and antituberculous therapy did not cause any significant improvement in the symptoms and he had a persistent fever of 39 °C or higher.

The patient then underwent an 18F-FDG PET/CT examination as a systemic search to explain fever of unknown origin. PET images and PET/CT fusion images demonstrated intense symmetric FDG uptake in auricle, larynx, tracheobronchial tree and all intercostal cartilages (Fig. 1A–C, E). FDG was also accumulated in hilar, mediastinal, and axillary lymph nodes (Fig. 1C, D). Maximum intensity projection (MIP) also showed intense FDG uptake in these tissue and nasal cartilages (Fig. 2). The patient underwent a tragus cartilage biopsy, and pathological results showed cartilaginous lymphocytic infiltration and degeneration, in line with the performance of osteochondritis. In addition, the computed tomographic (CT) scan showed the corresponding parachondral soft tissue around intercostal cartilages was swollen and the wall of tracheobronchial tree was thicken. The lymph nodes mentioned above were enlarged (please confirm it on CT) (Fig. 1). The patient was treated with glucocorticosteroids that induced rapid improvement of the symptoms. Two months after starting therapy, the laboratory data, such as whole blood count and ESR, had returned to normal levels.

3. Discussion

RP is an uncommon autoimmune disease without clear process, and often occur with connective tissue disease simultaneously. It can affect multiple organs including nose, ears, peripheral joints, tracheobronchial cartilage and eyes, heart and skin.[4,5] Its diagnosis is mostly based on clinical manifestations and

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The subjects have signed a written informed consent.

The authors report no conflicts of interest.

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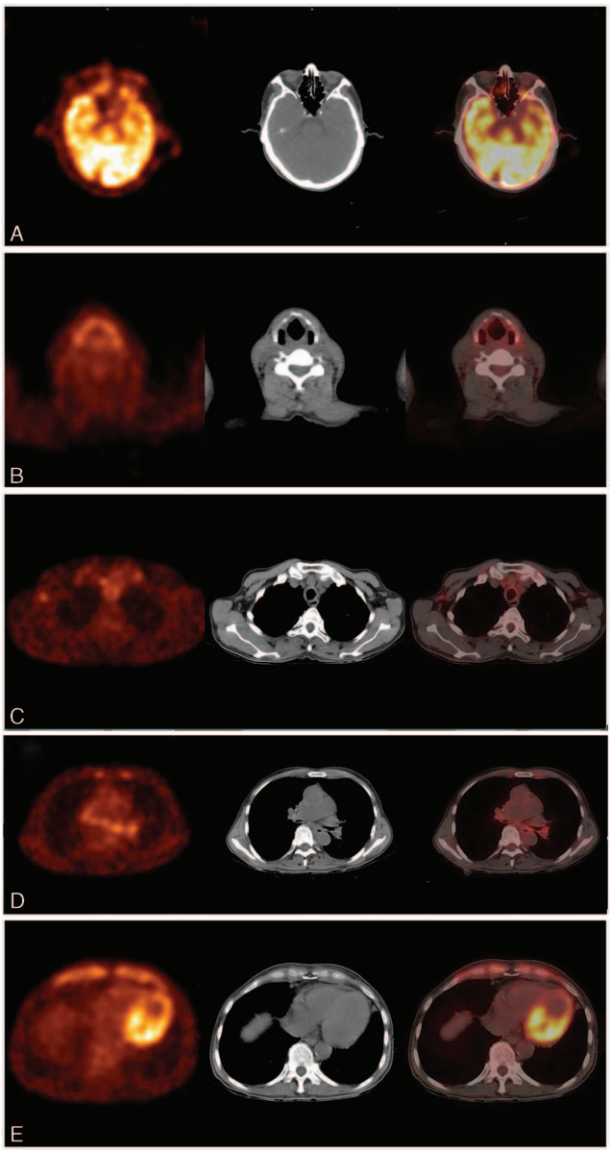

Figure 1. PET images (left), unenhanced CT images (middle), and the PET/CT fusion imagings (right) of auricle, larynx, trachea, and all costal cartilages and the axillary, hilum, and mediastinal lymph nodes, respectively. CT = computed tomography, PET = positron emission tomography.

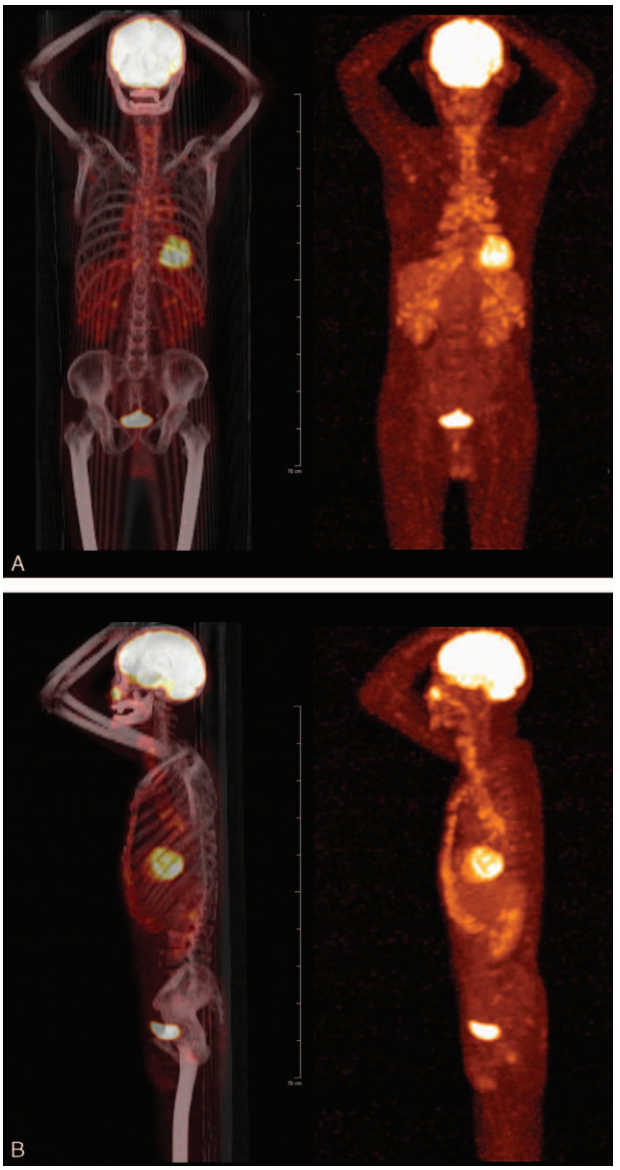

Figure 2. Coronal (A), sagital (B), and MIP PET images showed moderate FDG accumulation in the nasal cartilages, laryngeal cartilages, all costal cartilages, tracheobronchial tree and the axillary, hilum and mediastinal lymph nodes. FDG = fluorodeoxyglucose, MIP PET = maximum intensity projection positron emission tomography.

symptom-driven diagnostic testing multidetector. The corresponding clinical manifestations are ear cartilage inflammation, nasal cartilage inflammation, peripheral non-erosive polyarthritis, episcleritis, keratitis, and other multisystem diseases which involve respiratory system, cardiovascular system, and nervous system. As the disease progresses, respiratory tract involvement usually perform affect larynx, trachea, bronchus stenosis, bronchiectasis, pneumonia, atelectasis, etc,[6] in which the main airway and the left and/or right bronchus stenosis are most common. Davis et al[7] reported that not just the main airway and the left and right bronchial stenosis but the surrounding small bronchi may also be involved.

The diagnosis bases on clinical manifestations, and no specific laboratory methods or specific histologic findings are considered pathognomonic for RP. Awareness should be raised about systemic multiple cartilage damage associated with unexplained chronic cough, sputum, hoarseness, wheezing, and even dyspnea.

Multidetector CT can clearly define the location and extent of the fixed airway narrowing and wall thickening. Bronchoscopy can visually observe the edema, thickening of bronchial wall, and disappeared cartilage ring as an invasive operation. However, tracheobronchial lumina narrowing is not specific to RP. Other causes that should be considered include infection, amyloidosis, tuberculosis etc. Tc-99m methylene diphosphonate (MDP) bone scintigraphy has also been used to assess RP. Some case reports demonstrated that scintigraphic findings were improved after prednisolone therapy,[8,9] therefore, MDP scanning may be a valuable method in the follow-up of RP.

Our case demonstrates that although the diagnosis is mainly established clinically, the use of 18F-FDG-PET/CT has been proven to be a useful diagnostic tool to accurately determine the extent of inflammation throughout the body. Several studies have reported clinical value of FDG PET/CT imaging for the diagnosis

of RP.[10–12] Most of them well depicted tracheobronchial tree and intercostal cartilages involvement, but the nasal cartilages, larynx, and reactive lymphadenopathy were rarely mentioned entirely, as was noted in our case.

Currently, there is no ideal treatment of this disease. Primary therapy includes corticosteroids, dapsone, and other immunosuppressants, which are partly useful for acute episode. For the tracheobronchial stenosis and/or softening, intratracheal stent implants can significantly improve respiratory symptoms, which is an effective treatment.

In conclusion, FDG PET was found to have a growing role in the diagnosis and follow-up of relapsing polychondritis.

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