Unusual unilateral renal mass with pulmonary multiple nodules as the initial presentation of granulomatosis with polyangiitis

Abstract

We report a case of a 32-year-old man with recurrent fever, cough and left lumbago for more than one month. Computed tomography (CT) and magnetic resonance imaging (MRI) found bilateral multiple pulmonary nodules and a tumor-like mass in the left kidney. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) revealed increased uptake in the right pharyngeal recess along with pulmonary and renal hypermetabolic lesions. The pathologic findings of pulmonary and renal specimens were suggestive of granulomatous inflammatory changes. Further laboratory examinations showed an elevated level of serum cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA) and serum proteinase 3-ANCA (PR3-ANCA). Clinical symptoms were significantly improved, and the size of pulmonary and renal lesions reduced following the use of steroids and cyclophosphamide together. Therefore, a final diagnosis of granulomatosis with polyangiitis (GPA) was made.

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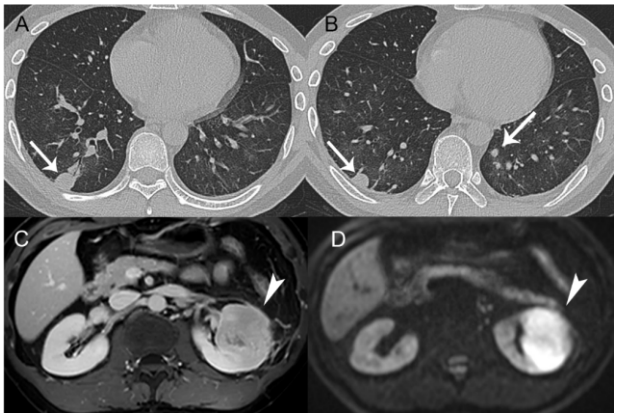

Figure 1. A 32-year-old man presented with a 1-month history of recurrent fever, cough and left lumbago. He also complained of hearing impairment, asthenia, anorexia and arthralgia. The patient was a smoker with a fifteen-year history of smoking about 10 cigarettes a day. Routine laboratory studies were unremarkable, with normal renal function and urinalysis. Clinical symptoms persisted during antibiotic therapy. The enhanced thoracic CT revealed bilateral pulmonary multiple solid nodules without calcification or cavitation (A, B: arrow); and the abdominal MRI exhibited an irregular mass measuring 53x48 mm at the enlarged left kidney. The lesion showed mild to moderate enhancement on postcontrast T1-weighted image and hyperintense signal on diffusion weighted image (C, D: arrowhead). Primary renal carcinoma with pulmonary metastasis was suspected.

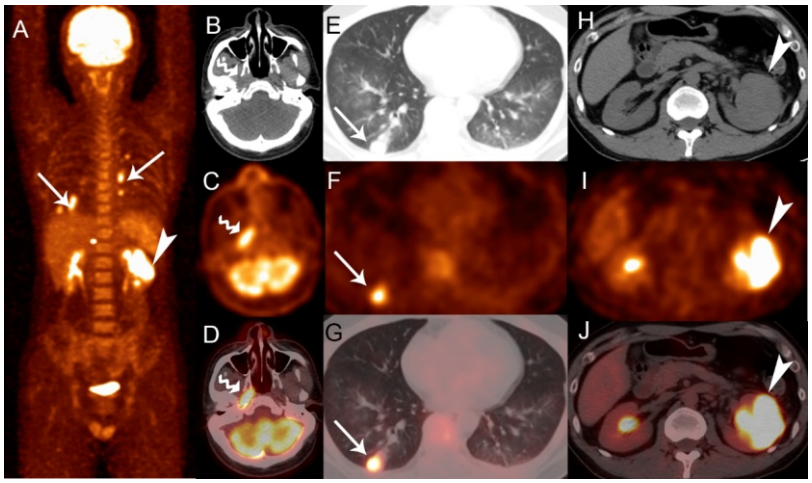

Figure 2. Fluorine-18-FDG PET/CT was subsequently performed for further evaluation. Multiple hypermetabolic lesions were found on maximum intensity projection (MIP) image (A). The axial images of PET, CT, and PET/CT fusion revealed hypermetabolic lesions in the bilateral pulmonary nodules (E, F and G: arrow) and left renal mass (H, I and J: arrowhead) with SUVmax of 7.64 and 13.29 respectively, which were compatible with findings in CT and MRI. Beyond these, there was increased tracer uptake in the right slightly narrowed pharyngeal recess with SUVmax of 6.71 (B, C and D: crooked arrow). In view of imaging features, malignancy especially lymphoma or systemic disease was suspected.

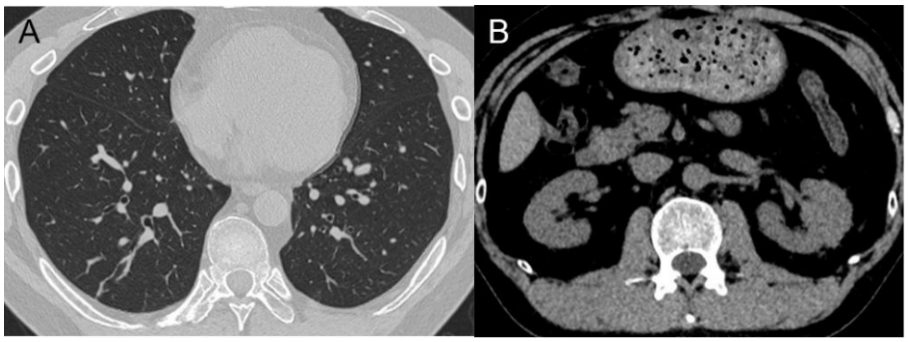

Figure 3. Computed tomography-guided percutaneous lung and kidney biopsy was then performed. The results indicated infiltration of acute and chronic inflammatory cells, hyperplasia of fiber and granulation tissue, which were rather nonspecific except for granulomatous inflammatory features. Otolaryngoscopy examination proved to be negative. Further laboratory examinations showed serum c-ANCA and serum PR3-ANCA tests were positive with c-ANCA titer equal to 1:10 (normal reference range: negative) and PR3-ANCA 8.6U/mL (normal reference range: less than 1.0 U/mL), respectively. Granulomatosis with polyangiitis (GPA) was highly suspected, subsequently, the patient was treated with methylprednisolone and cyclophosphamide. After eight cycles, the patient had significant clinical improvement and radiographic remission with reduction of pulmonary nodules and renal mass (A, B), which finally led to a diagnosis of GPA.

Granulomatosis with polyangiitis, previously known as Wegener granulomatosis, is one of ANCA-associated necrotizing vasculitis characterized by necrosis, vasculitis and granulomatous inflammation [1]. As a multiorgan systemic disease, it mainly involves the upper and lower respiratory tracts and kidneys [2]. Besides, articular manifestations are found in 50-70% and may be evaluated by MRI. The pulmonary nodules in GPA are commonly cavitary, and renal involvement is usually bilateral with pauci-immune necrotizing crescentic glomerulonephritis [3]. However, the diagnosis of GPA can be challenging for its various presentations [4-8]. In addition, GPA can emerge as a paraneoplastic syndrome, most frequently in renal cell carcinoma [9]. In our case, GPA manifested as a unilateral tumorous kidney mass with multiple bilateral solid pulmonary nodules and increased tracer uptake in the right pharyngeal recess upon ^{18}F-FDG PET/CT imaging. There was no abnormal metabolic radioactivity within joints, although the patient complained of arthralgia. Through ^{18}F-FDG PET/CT study, the extent of disease was better assessed, which was helpful for the diagnosis and management alteration [10, 11]. Fluorine-18-FDG PET/CT is a valuable modality for GPA.

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