中文題目:肉芽腫併多發性血管炎以多處皮膚結節表現案例報告

英文題目: Granulomatosis with polyangiitis presenting with multiple subcutaneous nodules

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Introduction

Granulomatosis with polyangiitis(GPA) is an autoimmune systemic disease characterized by necrotizing granulomatous inflammation and anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis. Cutaneous manifestations are varied and could be presented before, during or after the diagnosis of systemic disease. Here, we presented a case of GPA with initial manifestation of subcutaneous nodules, and further positron emission tomography (PET) showed diffuse skin lesions and multiple organs involvement.

Case presentation

A 75-year-old female presented with a 3-month history of multiple subcutaneous tender nodules over the trunk and extremities. She was admitted to our hospital due to fever and malaise. The lab data showed leukocytosis and elevated C-reactive protein level. Bilateral pulmonary cavitary nodules were noted under chest computed tomography(CT) scan. Amoxicillin/Clavulanate and Azithromycin were used for pneumonia and suspected subcutaneous abscess . Three days later, histology of skin specimens showed granulomatous inflammation with negative results of bacteria, fungus, mycobacterium tuberculosis and atypical mycobacteria infection. Echocardiogram revealed no obvious vegetation. The antibiotics were changed to Ceftriaxone plus Teicoplanin but fever persisted with worsening skin lesions noted (Figure 1). In addition, bilateral ear fullness and right facial swelling were mentioned. PET/CT revealed multiple FDG(2-deoxy-2-[18F] fluoroglucose)-avid lesions in bilateral lungs, right maxillary sinus and diffuse skin/subcutaneous in the whole body(Figure 2). Under the suspicion of granulomatosis with polyangiitis(GPA), we checked the autoimmune parameters and added intravenous methylprednisolone for her. Dramatic improvement was noted and the serum PR3(proteinase 3)- ANCA showed positive (51 IU/mL) eventually.

The pathology report of right upper lung mass via bronchoscopy was compatible with GPA. Pulse therapy with methylprednisolone 500mg once daily was given and she was discharged with oral prednisolone.

Discussion

Granulomatosis with polyangiitis(GPA), previously known as Wegener's granulomatosis, is an autoimmune systemic necrotizing vasculitis associated with the presence of anti-neutrophil cytoplasmic antibodies(ANCA) directed against proteinase 3(PR3). GPA can involve any organ, but it predominantly affects the upper and lower respiratory tracts (90%), renal (60%) and eyes (1). Cutaneous manifestations, presenting in 35-50% of the patients, are polymorphic including palpable purpura, subcutaneous nodules, pyoderma gangrenosum-like ulcers and livedo reticularis. Palpable purpura is the most common skin lesion and mostly seen in the lower limbs(2). Histopathologic features of cutaneous GPA contain not only leukocytoclastic vasculitis but also extravascular palisading granuloma and granulomatous inflammation(3). In reported cases, skin lesions may occur as the initial manifestation of GPA. Previous report also concluded that cutaneous manifestations were potentially associated with a higher frequency of articular and renal involvement (2,4). Regarding otological manifestations, the most common one described is serous otitis media followed by chronic otitis media. Besides, sensorineural hearing loss is also mentioned due to the cochlear vessel vasculitis (5).

Back to our patient, fever and multiple subcutaneous nodules were the initial problems, combined with bilateral pulmonary cavitary nodules. Septic emboli or malignancies with distant metastasis were our initial impression. However, her general condition deteriorated under antibiotic treatment and the skin lesions progressed to widespread pyoderma gangrenosum-like ulcerations. PET/CT provided the strong suspicion of GPA due to FDG-avid lesions involving the right maxillary sinus, bilateral lungs and diffuse subcutaneous areas simultaneously. Histology of skin biopsy showed granulomatous inflammation. The PR3-ANCA also showed positive results. Thus, GPA was confirmed according to ACR/EULAR guidelines. Pulse therapy was given and there was no obvious renal involvement during this admission. However, she was admitted to our hospital 3 weeks later due to rapidly progressive renal function (creatinine 0.54 to 5.78 mg/dl). GPA with flare up and renal involvement was highly suspected; therefore, she received intravenous methylprednisolone treatment again.

Our case outlines the importance of the cutaneous manifestations of GPA that may facilitate the early diagnosis of the disease. Meanwhile, skin lesions may be considered a predictor of further kidney damage. Furthermore, her PET/CT revealed a precious image for GPA with multiple organ involvement, including maxillary sinus, lung and skin.

Conclusion

Granulomatosis with polyangiitis(GPA) can involve any organ. Its cutaneous manifestations vary and could be the initial presentation of the disease.

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Figure 1. Multiple subcutaneous nodules were noted over the trunk and progressed to pyoderma gangrenosum-like ulcerations.

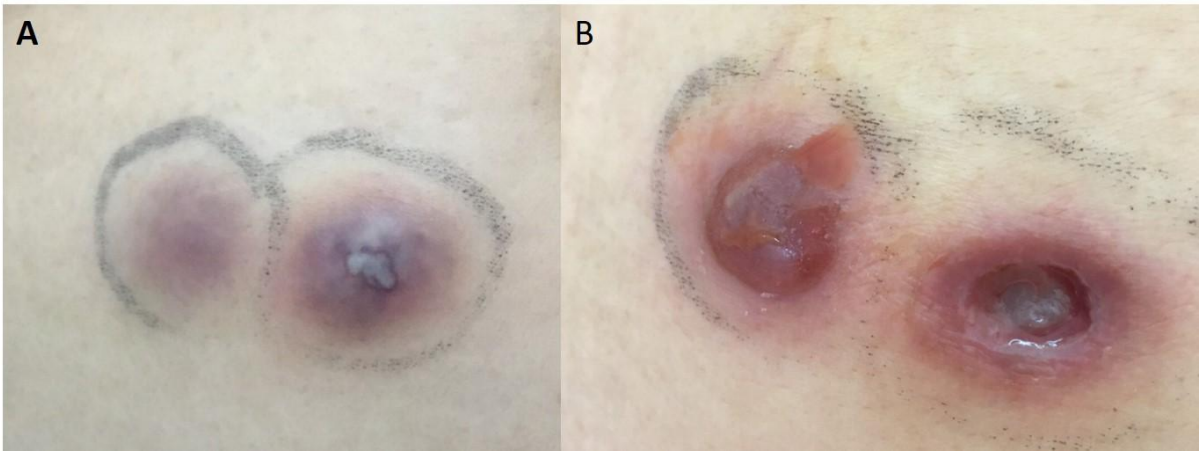

Figure 2. PET/CT: FDG-avid lesions involving the right maxillary sinus, bilateral lungs and diffuse subcutaneous areas (as arrows).