ANCA-negative granulomatosis with polyangiitis with diffuse pulmonary nodules presenting as intracardiac mass lesion

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Abstract:

Background: Granulomatosis with polyangiitis (GPA) is a subtype of anti-neutrophil cytoplasmic antibody-associated vasculitis (AAV), involving small and medium sized vessels, often affecting the kidneys and lungs[1]. Cardiac involvement has been reported in 6% to 44% of GPA cases, primarily as pericarditis and coronary arteritis[2]. Literature review reveals a small number of case reports of GPA cases presenting with conduction abnormalities in association with an intracardiac mass[3-4]. Here, we report the presentation of ANCA-negative limited GPA in a young man with an intracardiac mass lesion resulting in a complete atrioventricular heart block.

Case report: A 35-year-old previously healthy Hispanic man was admitted for evaluation of dizziness. Electrocardiogram (EKG) revealed a complete atrioventricular heart block. His chest X-ray showed no acute abnormalities. Transthoracic echocardiography (TTE) demonstrated normal biventricular structure and dimensions and preserved ejection fraction, and cardiac magnetic resonance imaging (CMR) showed a large, intramural infiltrating enhancing mass expanding the interatrial septum and involving the fibrous skeleton of the heart with no late gadolinium enhancement of the left ventricle. The initial differential diagnosis ranged from infectious to autoimmune to malignant etiology. A whole body fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) demonstrated increased FDG uptake within the interatrial region, corresponding to the intracardiac mass seen on CMR, as well as multiple FDG avid nodules in bilateral lungs, precarinal, prevascular and left hilar nodes, and the mid and lower sternum. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) biopsy of corresponding right middle lung nodule, hilar/mediastinal lymph nodes from peribronchial lesions, and transesophageal echocardiogram (TEE) guided percutaneous endomyocardial biopsy were inconclusive. A video-assisted thoracoscopic surgery (VATS) lung biopsy revealed ill-defined necrotizing granulomatous inflammation with hyperchromatic multinucleated giant cells, associated neutrophilic microabscesses, and medium-sized vessel necrotizing vasculitis. Laboratory evaluation revealed elevations of ESR and CRP with negative ANCA antibodies. Angiotensin converting enzyme (ACE) level was within normal range. Extensive testing revealed no infectious etiology for his illness. COVID-19 PCR/Swab and IgG Antibody were negative. The constellation of histologic findings established an underlying diagnosis of vasculitis and high dose corticosteroids were initiated for treatment of limited GPA. The patient underwent successful and uncomplicated dual-chamber pacemaker

implantation and was subsequently discharged. At the 2-week follow up, methotrexate was initiated and he was started on a tapering regimen of corticosteroid therapy.

Conclusion: Cardiac involvement of vasculitis that leads to mass formation and conduction system abnormalities is extremely rare. Further, 10-20% of patients with GPA are ANCA negative, allowing for the diagnosis to be overlooked, particularly in those patients with atypical presentations[5]. Because available treatments such as glucocorticoids and other immunosuppressive therapies can improve the outcome of vasculitis, we believe all physicians should include systemic vasculitis in the differential diagnosis in treating patients who present with a similar intracardiac mass lesion. Early detection and treatment can lead to more favorable outcomes.

References:

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