

Unusual cardiac involvement in granulomatosis with polyangiitis manifesting as acute congestive heart failure

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Introduction

Granulomatosis with polyangiitis (GPA), known as Wegener's granulomatosis, is characterized by systemic necrotizing vasculitis in small and medium sized vessels affecting the upper and lower respiratory tracts, paranasal sinuses, and kidneys (1). We here describe a patient who presented with acute congestive heart failure (CHF) and progressive renal failure requiring hemodialysis and was later diagnosed with GPA based on positive renal biopsy.

Case Report

A 69-year-old man presented with 2–3 weeks' duration of progressive dyspnea. His exercise capacity was unlimited until 2 months ago, but lately, he was able to walk only 1-2 blocks before experiencing shortness of breath along with leg edema. System review was overall negative, except for dry cough for 3–4 weeks and chronic sinusitis for which he was taking antihistamines.

On examination, he was in mild respiratory distress with inspiratory crackles, elevated JVP of 7-8 cm (S3 gallop sound), and palpable liver. Electrocardiogram (ECG) showed sinus rhythm with a nonspecific intraventricular conduction delay and congested pulmonary vessels on chest X-ray. Laboratory tests revealed a WBC count of 13.6 (normal range, 4.00–11.0 $\times 10^9/L$), serum creatinine level of 0.9 (normal range, 0.6–1.2 mg/dL), BNP of 2844 (normal range, 100–300 pg/mL), and troponin I level of 0.08 at admission with a peak level of 0.1 (normal range, ≤ 0.04 ng/mL). As the clinical presentation was consistent with acute CHF, he was promptly treated with intravenous diuretic and oxygen via nasal cannula. Transthoracic echocardiogram (TTE) showed severely reduced left ventricular ejection fraction (LVEF, 15%) and dilated cardiomyopathy (DCM) with left ventricular end-diastolic diameter of 6.6 cm (normal range, 4.2–5.9), interventricular septum diastolic thickness of 1 cm, left atrium systolic diameter of 5.2 cm (normal range, 3–4

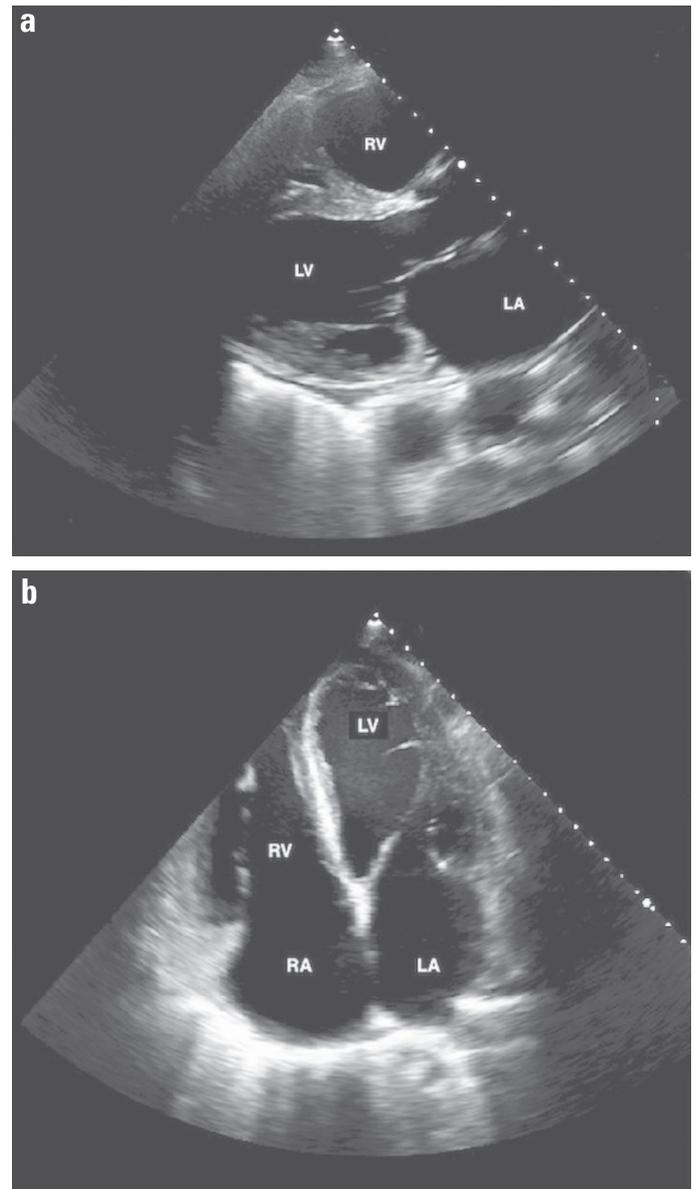


Figure 1. Transthoracic echocardiogram: left parasternal (a) and apical 4-chamber (b) views demonstrating dilated cardiomyopathy and severely reduced global left ventricular systolic function with an ejection fraction of 15%

LA - left atrium; LV - left ventricle; RA - right atrium; RV - right ventricle

cm), and pulmonary artery systolic pressure of 53 mm Hg (Fig. 1). Coronary artery catheterization was performed, and it revealed normal coronary vasculature (Fig. 2).

Heart failure symptoms had significantly improved with IV diuretic, but on hospitalization day 5, the serum creatinine level elevated to 1.7 mg/dL and WBC count to $15 \times 10^9/L$ with absolute eosinophil of 0.1 $K/\mu L$ (normal range, 0–0.9). Therefore, diuretic was immediately stopped, but the serum creatinine level elevated to 3.2 mg/dL on hospitalization day 10. Urinalysis revealed an RBC count of 132 (normal range, 0–3/HPF) and WBC count of 18 (normal range, 0–5/HPF) without symptoms or bacterial growth in

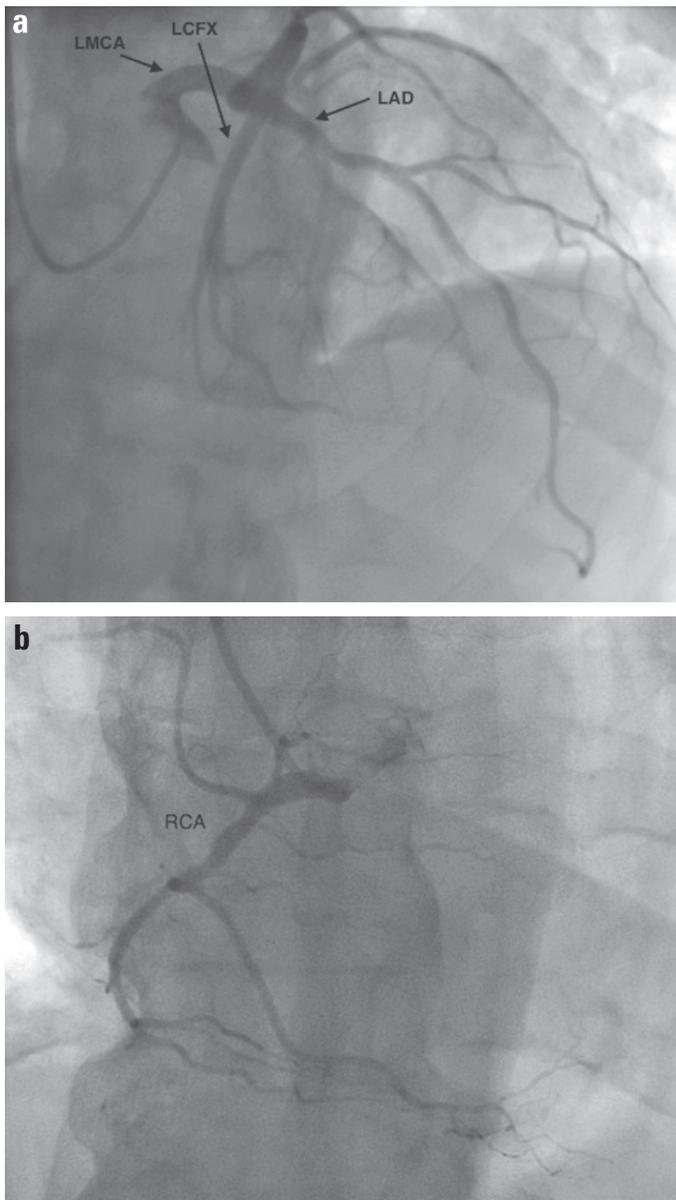


Figure 2. Coronary angiographic views reveal no significant stenosis in (a) left and (b) right coronary arteries

LAD - left anterior descending artery; LCFX - left circumflex artery; LMCA - left main coronary artery; RCA - right coronary artery

urine culture. Renal ultrasound showed normal kidney structure without stone. Cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) were 315 AU/mL (normal range, 0–19), ESR was 95 mm in 1 h, and CRP was 30 (normal range, <8 mg/L). Therefore, cANCA-associated vasculitis including GPA, microscopic polyangiitis, and eosinophilic granulomatosis with polyangiitis (EGPA) were considered as differential diagnoses, but renal biopsy exhibited pauci-immune glomerulonephritis (Fig. 3). Diagnosis of GPA was made, and plasmapheresis, oral cyclophosphamide, and steroid were initiated. However, renal function was getting worse, which required hemodialysis on hospitalization day 25. He was able to perform physical activities, but still experienced dyspnea with some activity (NYHA class 3). Then, the patient

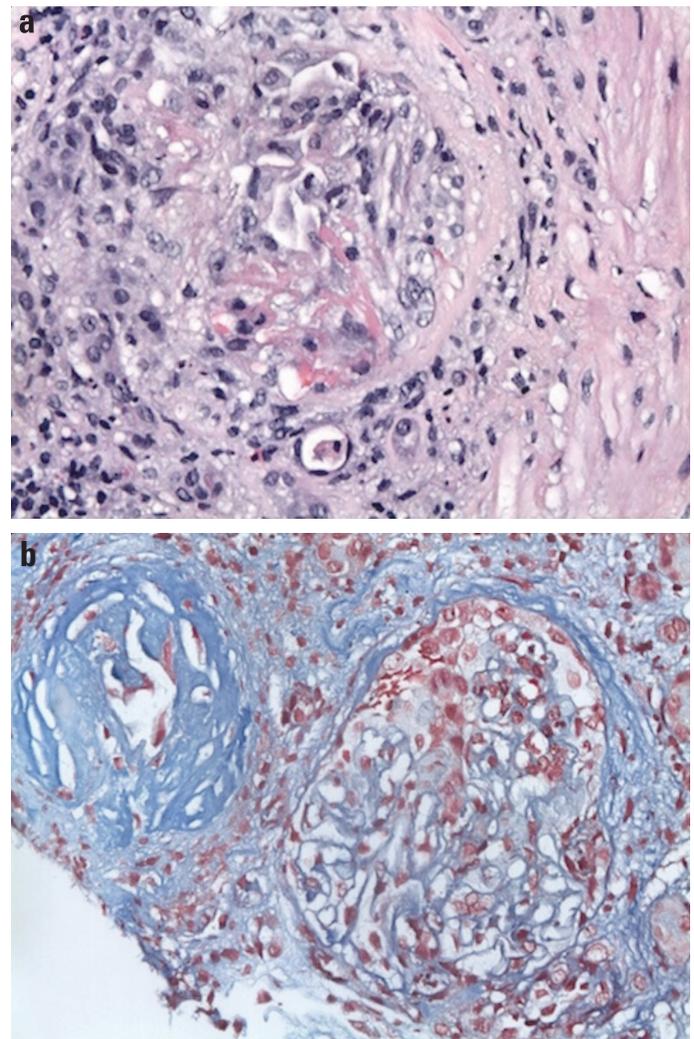


Figure 3. Renal biopsy demonstrates (a) sclerotic glomeruli with segmental cellular crescent formation in a background of mixed interstitial inflammation composed of mature lymphocytes, plasma cells, and eosinophils (400× hematoxylin and eosin stain) and (b) shrunken globally sclerotic glomeruli and fibrocellular crescent glomerulus with fibrin deposition-red on trichrome stain (400× trichrome)

was discharged to a long-term care facility on cyclophosphamide, valsartan 40 mg 12 hourly, and metoprolol ER 25 mg daily.

Discussion

GPA can manifest multiple organ systems and present with a variety of symptoms. Cardiac involvement in GPA is infrequent with an estimated incidence of 3.3% in which pericarditis, myocarditis, and conduction system defects are the most frequent one (2). Furthermore, DCM with CHF related to GPA is exceedingly exceptional, and our extensive literature search revealed only eight cases reported in the past since GPA was described in 1936 by Wegener (3–9). Of the eight cases, only three presented with acute heart failure (4, 8, 9). The presence of small vessels necrotizing vasculitis could be an attributing factor of cardiac involvement in GPA, but the exact mechanism is uncertain.

With an advancement of imaging modalities, MRI has been useful to evaluate the end organs involvement along with cANCA, ESR, and CRP, although tissue biopsy remains the gold standard (10). In this case, the presence of cANCA along with normal eosinophil count and positive renal biopsy confirmed GPA. Cyclophosphamide therapy in GPA could lead to a DCM, but in our case, congestive cardiomyopathy was seemingly due to GPA as he was not taking any medicines (7). Endomyocardial biopsy or cardiac MRI with contrast was not performed given biopsy-positive GPA, positive inflammatory markers, and impaired renal function, but nonetheless, it could be a limitation of this case. To the extent of our knowledge, this is the fourth case with acute CHF as the initial presentation of GPA.

Conclusion

This case reminds clinicians that acute CHF with worsening renal function could be an initial manifestation of GPA, which should be included in the differential diagnosis.

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Radiofrequency ablation of accessory pathways in a toddler with Ebstein's anomaly and functional single ventricle physiology

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Introduction

The incidence of multiple accessory pathways (AP) in patients undergoing electrophysiology study (EPS) for tachycardias is higher in structural heart disease such as Ebstein's anomaly. Ablation of APs is necessary at a younger age, because tachycardia is poorly tolerated in these patients owing to compromised cardiac reserve.

Case Report

A 3-year-old girl, weighing 11 kg, with a diagnosis of Ebstein's anomaly was referred to our center due to recurrent supraventricular tachycardia (SVT) attacks resistance to multidrug medical therapy. She had a modified Blalock-Taussig shunt operation in the neonatal period, and thereafter suffered from recurrent SVT attacks compromising hemodynamics, requiring cardioversion. A surface electrocardiogram showed preexcitation consistent with Wolf-Parkinson-White Syndrome. An electrophysiology study with RF ablation of AP followed by hemodynamic study before bidirectional Glenn operation was planned.

The electrophysiology study was conducted under general anesthesia. A three-dimensional mapping with the ESI system (EnSite System, St. Jude Medical, Minneapolis, MN, USA) was utilized during the procedure.

Recurrent SVT attacks induced during diagnostic catheter placement and causing hypotension and desaturation were stopped with adenosine administration. Baseline measurements were performed (AH: 82 ms, HV: 0 ms, BCL: 700 ms, PR: 115 ms, QRS: 132 ms, and QT: 450 ms).

Standard atrial stimulation protocol was carried out and orthodromic SVT with narrow QRS and tachycardia cycle length of 324 ms was induced. Because of hemodynamic compromise during SVT ESI, system mapping was done only for a short duration and the earliest VA conduction was found in right posteroseptal region of the tricuspid annulus with 63 ms (PERP: 320 ms, shortest preexcited R-R interval in AFIB: 380 ms). This region was marked via ESI system (Fig. 1) and a 5F RF ablation catheter was advanced into the right atrium positioned directly to this site. With a 50 W-50 C0 application for 2 s, AP was lost and most of the preexcitation on the 12-lead electrocardiogram was also lost