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Letter to the Editor

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Letter to the Editor (Case report)

Pericardial tamponade: a rare life-threatening manifestation of eosinophilic granulomatosis with polyangiitis

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Key message

- Eosinophilic granulomatosis with polyangiitis can rarely present with life-threatening cardiac tamponade and responds to CSs and rituximab.

Dear Editor, We describe a case of a 27-year-old male who presented to the hospital with a 3-week history of epistaxis, cough, chest pain, haemoptysis and bilateral lower extremity swelling. His past medical history was significant for post-streptococcal glomerulonephritis diagnosed at age 7 years, for which he received glucocorticoids for 3 years. Despite treatment, he progressed to chronic kidney disease stage 5. Other past medical history was notable for non-crystal-proven gout and three or four episodes of shortness of breath, wheezing, dry cough and sinus congestion over the past 2 years, all glucocorticoid responsive, with a presumed diagnosis of asthma. The patient resided in Ohio, without recent travel or tuberculosis exposure. He had never smoked or used recreational drugs.

Physical examination at presentation revealed stable vital signs. The patient appeared pale and had friction rubs and bilateral crackles on cardiac and lung auscultation, respectively. He was noted to have diffuse sinus tenderness and nasal crusting, without nasal deformity or septal perforation. There was no evidence of a purpuric rash.

Initial laboratory evaluations were notable for normocytic anaemia, with a haemoglobin of 7.6 g/dl (baseline haemoglobin ~12 g/dl), leucocytosis of 16.39×10^3/µl (79% neutrophils, 10% eosinophils, with absolute eosinophil count 1740/µl), elevated creatinine at 22.44 mg/dl, blood urea nitrogen of 223 mg/dl, high-sensitivity troponin at 264 ng/l and B-type natriuretic peptide at >70 000 pg/ml. ECG showed sinus tachycardia, without ST segment elevation or T wave inversion.

Plain chest radiography showed diffuse airspace opacities in both lungs. CT of the chest without i.v. contrast revealed multifocal bilateral ground glass and consolidative opacities involving all lobes, a cavitary lesion in the left lower lobe, and trace pericardial and bilateral pleural effusions. Initial echocardiogram showed normal left ventricular systolic function, with an ejection fraction of 61%, and a small pericardial effusion, without tamponade physiology.

The patient was started on haemodialysis immediately for treatment of severe fluid overload and uraemic pericarditis. Uraemic pericarditis was considered owing to the presence of chest pain and prior dyspnoea in the setting of known kidney disease. However, owing to other accompanying clinical features, inconsistent ECG findings and the presence of eosinophils, alternative aetiologies were explored. An extensive respiratory infectious work-up, including a viral panel, *Legionella* and *Streptococcus pneumoniae* urine antigens and a tuberculosis test, was negative. He underwent bronchoscopy, which showed no evidence of alveolar haemorrhage or underlying infectious process. However, cell differential on bronchoalveolar lavage fluid showed 6% eosinophils. Autoimmune work-up was notable for an elevated IgE level at 567 kU/l, negative ANCA and anti-GBM antibodies, and normal complement levels (C3 and C4). A CT of the sinuses showed severe diffuse mucosal thickening in most sinuses. The combination of presumed diagnosis of adult-onset asthma, sinonasal symptoms, peripheral and tissue eosinophilia, and pulmonary opacities on CT chest raised the suspicion for eosinophilic granulomatosis with polyangiitis (EGPA). While awaiting cardiac PET to evaluate potential myocardial involvement, the patient became haemodynamically unstable. Repeated chest imaging showed stable diffuse airspace opacities but an enlarged cardiac silhouette (Fig. 1A); an echocardiogram showed worsening pericardial effusion, with tamponade physiology (Fig. 1B–D). Therefore, the patient underwent emergency pericardiocentesis, during which 700 ml of bloody fluid was drained, with analysis showing 11% eosinophils. The patient was administered pulse-dose CSs for 3 days and given rituximab infusions, which led to significant improvement in clinical status. Eventually, cardiac

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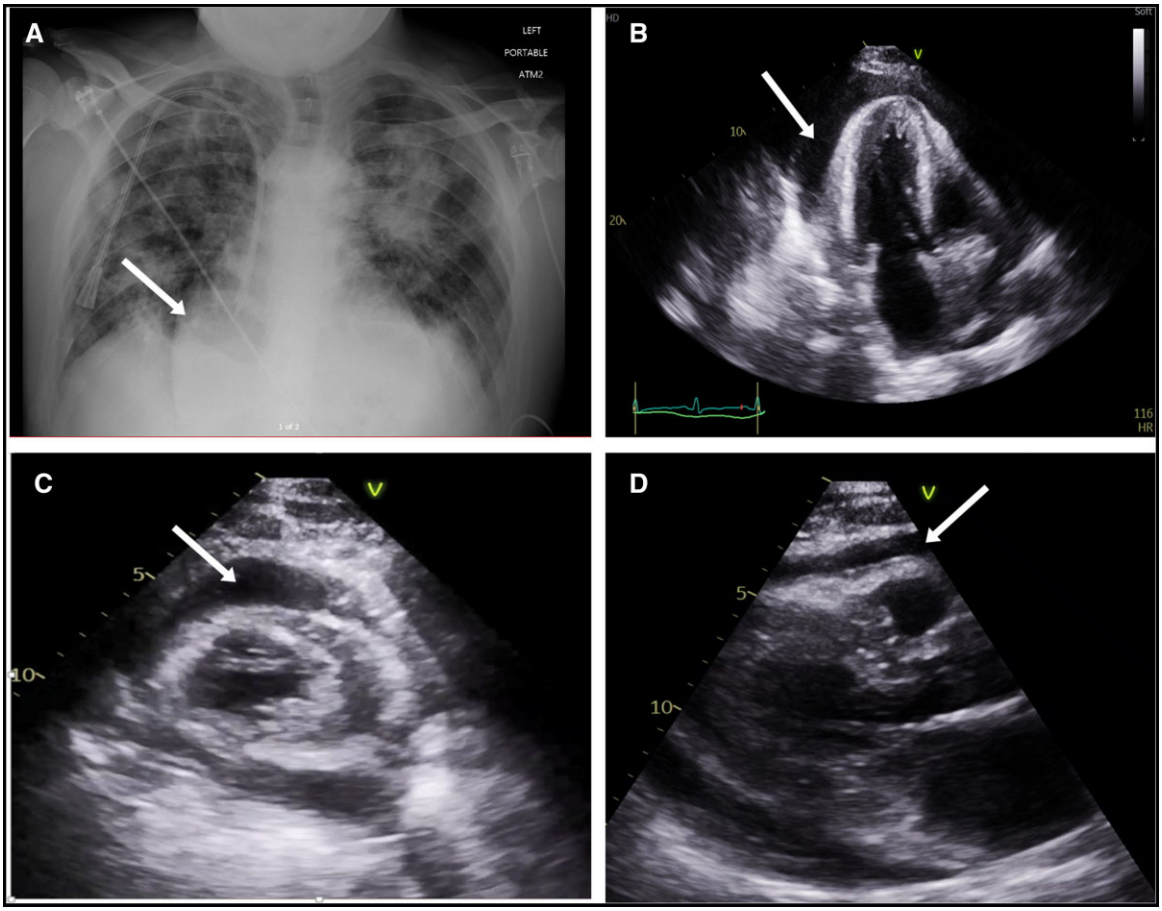

Figure 1. Plain chest radiograph and echocardiogram images. (**A**) Plain chest radiograph showing diffuse airspace opacities with enlarged cardiac silhouette. (**B**) Echocardiogram four-chamber view showing large anterior pericardial effusion. (**C**) Echocardiogram short-axis view at the level of the apex showing large circumferential pericardial effusion. (**D**) Echocardiogram parasternal long view showing right atrial collapse during early systole suggestive of tamponade physiology

PET revealed no myocardial enhancement. Open lung biopsy was not pursued owing to iatrogenic tension pneumothorax during the emergency pericardiocentesis.

Eosinophilic granulomatosis with polyangiitis (previously called Churg–Strauss syndrome) is a rare systemic necrotizing vasculitis that primarily affects small and medium vessels [1]. The disease is classified as ANCA-associated vasculitis; however, only one- to two-thirds of patients have positive ANCA, which often makes the diagnosis challenging [2]. EGPA is characterized by adult-onset asthma, chronic sinusitis, nasal polyposis, peripheral or tissue eosinophilia, pulmonary opacities or nodules, peripheral neuropathy and cardiac and renal involvement [1, 2]. Cardiac involvement, which is noted in 27–43% of cases, is more prevalent in ANCA-negative patients with high eosinophilic counts and is associated with a high mortality rate [2–5]. Although it may be subclinical in 17% of cases, most cases are symptomatic. Manifestations include cardiomyopathy (51.6%), coronary artery vasculitis (34%), intracardiac thrombus (22.6%) and pericardial effusion (37.1%), as reported in a large meta-analysis that included 62 EGPA patients with cardiac involvement [6]. Although pericarditis and a variable degree of pericardial effusion presenting as mild chest pain and dyspnoea are described in many studies [2, 4], pericardial tamponade with life-threatening presentation is rarely reported (only 6.5% of cases) [6, 7]. In such cases, urgent pericardiocentesis must be performed, which may reveal tissue eosinophilia and help in confirming the diagnosis of EGPA. Recent guidelines by the ACR and Vasculitis Foundation recommended high doses of glucocorticoids in addition to either CYC or rituximab for the treatment of severe EGPA [8]. This case highlights that cardiac involvement in EGPA is considered a severe manifestation with potential life-threatening presentation and consequences, especially when severe cardiomyopathy or pericardial tamponade is present.

Data availability

The data underlying this article cannot be shared, in order to protect the privacy/anonymity of the patient.

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References

1. Jennette JC, Falk RJ, Bacon PA *et al.* 2012 Revised International Chapel Hill Consensus Conference nomenclature of vasculitides. Arthritis Rheum 2013;65:1–11.
2. Comarmond C, Pagnoux C, Khellaf M *et al.*; French Vasculitis Study Group. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss): clinical characteristics and long-term followup of the 383 patients enrolled in the French Vasculitis Study Group cohort. Arthritis Rheum 2013;65:270–81.
3. Szczeklik W, Miszalski-Jamka T. Cardiac involvement in eosinophilic granulomatosis with polyangitis (Churg Strauss). J Rare Cardiovasc Dis 2013;1:91–5.
4. Hazebroek MR, van Paassen P, Dennert R, Heymans S. ANCA-associated vasculitis: microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis (Churg–Strauss Syndrome) and granulomatosis with polyangiitis (Wegener's Granulomatosis). In: The heart in rheumatic, autoimmune and inflammatory diseases. Academic Press, 2017: 459–502.
5. Sartorelli S, Cohen P, Dunogue P *et al.* Cardiac involvement of eosinophilic granulomatosis with polyangiitis (Churg–Strauss): initial manifestations and outcomes based on data from a monocenter patient cohort [abstract]. Arthritis Rheumatol 2019;71:71.
6. Pakbaz M, Pakbaz M. Cardiac involvement in eosinophilic granulomatosis with polyangiitis: a meta-analysis of 62 case reports. J Tehran Heart Cent 2020;15:18–26.
7. Yano T, Ishimura S, Furukawa T *et al.* Cardiac tamponade leading to the diagnosis of eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome): a case report and review of the literature. Heart Vessels 2015;30:841–4.
8. Chung SA, Langford CA, Maz M *et al.* 2021 American College of Rheumatology/Vasculitis Foundation guideline for the management of antineutrophil cytoplasmic antibody-associated vasculitis. Arthritis Care Res 2021;73:1088–105.