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Pulmonary-Cardiac Involvement in a Patient with IgA Vasculitis

CASE REPORT

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ABSTRACT

Immunoglobulin A (IgA) vasculitis (IgAV), formerly known as Henoch–Schönlein purpura, is a systemic small-vessel vasculitis caused by IgA immune-complex deposition; cardiac involvement is exceedingly rare and incompletely characterized. We describe a 41-year-old woman with biopsy-proven IgA nephropathy who presented with progressive dyspnea, hemoptysis, and a painful purpuric rash and was found to have severe multisystem IgAV with simultaneous cardiac, pulmonary, renal, and cutaneous involvement. High-sensitivity troponin T peaked at 20,208 ng/L and N-terminal pro-B-type natriuretic peptide at 49,428 pg/mL. Cardiac magnetic resonance imaging showed a left ventricular ejection fraction of 25%, diffuse mid-wall and epicardial late gadolinium enhancement with abnormal T1 and T2 values, and a 25 × 15 mm apical thrombus. Skin biopsy confirmed leukocytoclastic vasculitis and bronchoscopy confirmed diffuse alveolar hemorrhage, while multimodality imaging and a negative serologic work-up excluded cardiac sarcoidosis, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, and anti-glomerular basement membrane disease. The patient was treated with pulse methylprednisolone, plasma exchange, rituximab, and cautious anticoagulation, with resolution of alveolar hemorrhage, a progressive decline in cardiac biomarkers, resolution of the left ventricular thrombus, and partial recovery of left ventricular systolic function. This case expands the recognized spectrum of cardiovascular involvement in adult IgAV and highlights the value of multimodality imaging and multidisciplinary management in establishing the diagnosis and guiding therapy.

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INTRODUCTION

IgA vasculitis (IgAV), also known as Henoch–Schönlein purpura, is a systemic vasculitis caused by the deposition of IgA-containing immune complexes within the vessels of affected organs.[1,2] It predominates in children and classically presents with palpable purpura, arthralgia or arthritis, abdominal pain, and renal involvement, most commonly IgA nephropathy. Although IgAV is more common in children, it also affects adults, in whom it tends to behave differently, with a greater tendency toward renal involvement and a generally more severe course.[1,3] In adults, the most frequent manifestation is renal disease; less commonly affected systems include the peripheral and central nervous systems, the pulmonary system, and the skin.[1] Cardiac involvement is exceedingly rare in both pediatric and adult populations and is typically attributed to vasculitic injury of the coronary arteries and small intramyocardial vessels.[1,4] We report a case of acute perimyocarditis occurring in the setting of IgA vasculitis with concurrent pulmonary hemorrhage and cutaneous involvement.

CASE PRESENTATION

A 41-year-old woman from Saudi Arabia with biopsy-proven IgA nephropathy (Oxford MEST-C score M1 E0 S1 T1 C1), diagnosed in January 2025, presented to the emergency department on August 17, 2025, with a 4-day history of progressive shortness of breath, dry cough, and skin rash. The Oxford MEST-C score is a standardized grading of kidney biopsies in IgA nephropathy; her score indicated mesangial hypercellularity (M1), no endocapillary hypercellularity (E0), segmental sclerosis (S1), moderate (26-50%) tubular atrophy/interstitial fibrosis (T1), and crescents in fewer than 25% of glomeruli (C1).[5] She had completed a steroid taper in March 2025 and was maintained on valsartan 80 mg daily for proteinuria. The dyspnea occurred at rest and with exertion; the cough produced minimal hemoptysis (pink-tinged sputum), and the painful rash began with the respiratory symptoms, predominantly over the extremities. She denied chest pain, ischemic symptoms, and syncope, with no history of cardiac disease, and no viral prodrome, gastrointestinal symptoms, diabetes, smoking, immobilization, or prior thromboembolism.

On presentation she was tachycardic (134 beats/min), tachypneic (30 breaths/min), and normotensive, with oxygen saturation of 100% on 2 L/min nasal-cannula oxygen. Cardiovascular examination showed a regular tachycardic rhythm without murmurs or rubs; respiratory examination showed mild distress with a right-sided wheeze. Cutaneous examination revealed a painful petechial rash with a tender purpuric plaque over the dorsum of the left foot and scattered purpuric papules and macules over both feet and hands (Figure 1).

Initial laboratory evaluation revealed profound myocardial injury. High-sensitivity troponin T was 365 ng/L on admission (reference < 14 ng/L) and increased dynamically, peaking at 20,208 ng/L. N-terminal pro-B-type natriuretic peptide was markedly elevated at 12,552 pg/mL and increased to 49,428 pg/mL within 24 hours. Inflammatory markers were elevated, with

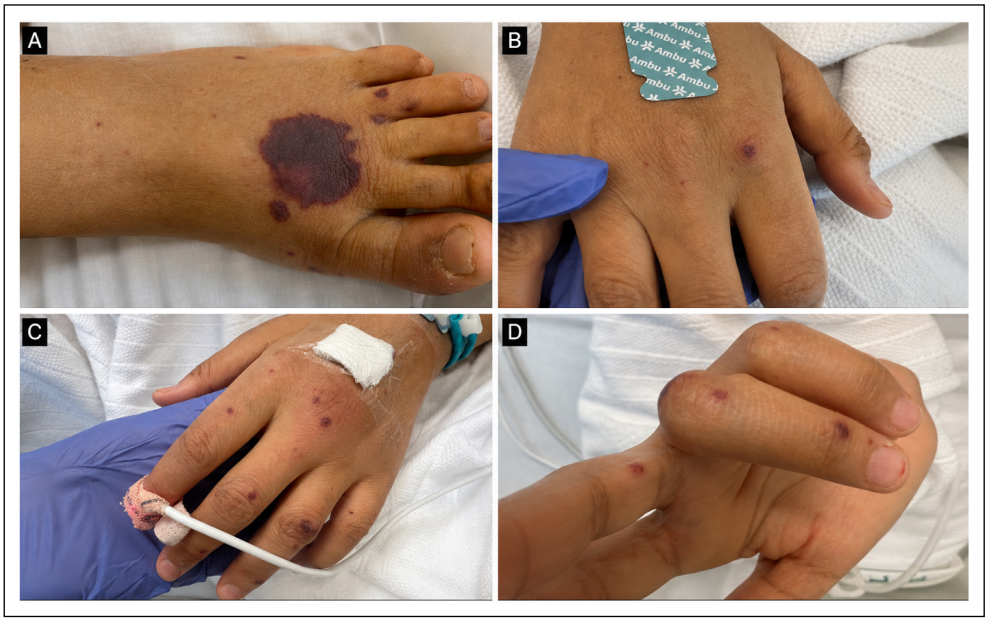


Figure 1 Cutaneous findings at presentation. **(A)** Dorsum of the left foot showing a large, tender, focally necrotic purpuric plaque with smaller surrounding lesions. **(B, C)** Dorsum of the hand showing scattered purpuric papules and macules. **(D)** Purpuric lesions on the digits.

mild anemia, leukocytosis, and marked thrombocytosis. Creatinine worsened from a baseline of 208 µmol/L to 255 µmol/L on admission. Autoimmune testing was negative for antineutrophil cytoplasmic antibodies (ANCA) and anti-glomerular basement membrane antibodies, with normal complement and a low-titer antinuclear antibody; antiphospholipid antibodies were subsequently negative.

Electrocardiography demonstrated sinus tachycardia with diffuse ST-segment elevation in the inferior, lateral, and precordial leads with PR-segment depression, and reciprocal changes in lead augmented vector right—a pattern consistent with acute perimyocarditis rather than territorial ST-elevation myocardial infarction. Transthoracic echocardiography showed moderately reduced left ventricular systolic function (ejection fraction ~35%) with global hypokinesis and no regional wall-motion abnormality, a markedly reduced global longitudinal strain (GLS) of -8% (GLS measures how much the left ventricular myocardium shortens along its long axis during systole; in healthy adults it is approximately -20%, with values closer to zero indicating worse function),[6] mild-to-moderate mitral and mild tricuspid regurgitation, and a small pericardial effusion with pericardial thickening but no tamponade. Cardiac magnetic resonance imaging revealed severely reduced left ventricular systolic function (ejection fraction 25%) with global hypokinesis and superimposed regional akinesis of the apical segments and basal-to-mid inferoseptum. There was diffuse patchy mid-wall and epicardial late gadolinium enhancement, particularly involving the basal inferolateral wall and basal-to-mid inferoseptum, with corresponding abnormal T1 and T2 values consistent with active myocardial inflammation (Figure 2C). Transmural (>90%) late gadolinium enhancement was present in the apical segments in a distal left anterior descending artery distribution, associated with a large left ventricular apical thrombus measuring approximately 25 × 15 mm (Figure 2A,B).

Because of acute kidney injury and the attendant risk of contrast-induced nephropathy, ischemia was evaluated noninvasively with technetium-99m stress myocardial perfusion imaging, which showed moderate-to-large fixed perfusion defects in the apical, anterolateral, and inferolateral walls without reversible ischemia, suggesting established scar rather than active obstructive coronary disease. Whole-body fluorine-18 fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) performed on September 10, 2025 (hospital day 24) demonstrated multifocal left ventricular myocardial FDG uptake, particularly involving the basal interventricular septum, without FDG-avid lymphadenopathy or evidence of extracardiac sarcoidosis (Figure 3). Although reported as metabolically suggestive of cardiac sarcoidosis, FDG uptake is not specific in active systemic inflammation and may also reflect active myocarditis.

Given persistent hemoptysis, high-resolution CT of the chest demonstrated diffuse bilateral ground-glass opacities with patchy consolidation, raising concern for diffuse alveolar hemorrhage. Flexible bronchoscopy on August 19, 2025, showed progressively hemorrhagic sequential bronchoalveolar lavage with active bleeding from the right upper lobe and the basal segments of both lower lobes; lavage microbiology and cytology were unremarkable. Punch biopsy of the cutaneous lesions demonstrated leukocytoclastic vasculitis with superficial and deep perivascular neutrophilic infiltration, leukocytoclasia, hemorrhage, and fibrin deposition within vessel walls extending into the subcutis. Direct immunofluorescence was negative for IgA, IgM, C3, C1q, and fibrinogen, with weak nonspecific perivascular IgG. Despite the absence of IgA on immunofluorescence, the prior biopsy-proven IgA nephropathy and concurrent multisystem involvement remained most consistent with IgA vasculitis; she also fulfilled the EULAR/PRINTO/PRES classification criteria for

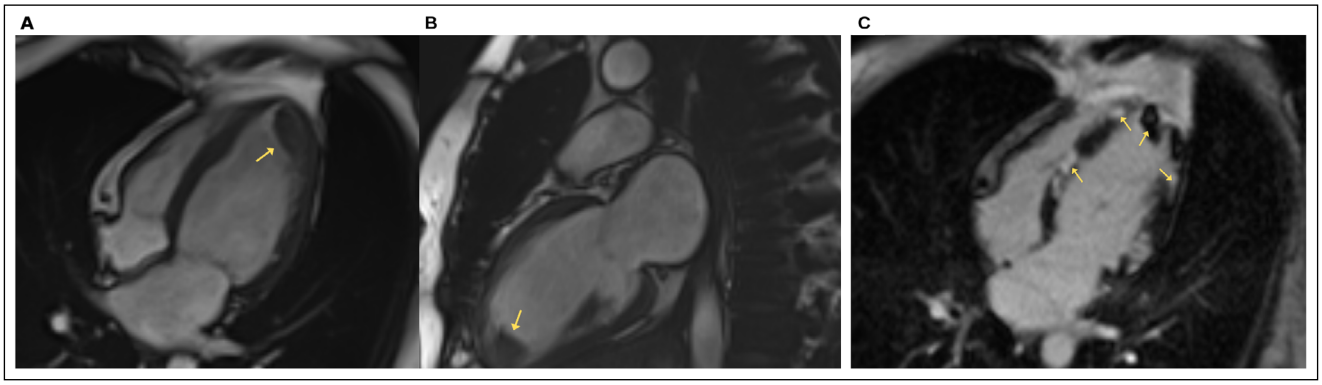


Figure 2 Cardiac magnetic resonance imaging. **(A)** Four-chamber and **(B)** two-chamber cine images showing a large left ventricular apical thrombus measuring approximately 25 × 15 mm (arrows). **(C)** Late gadolinium enhancement image showing diffuse patchy mid-wall and epicardial enhancement involving most myocardial segments, particularly the basal inferolateral wall and basal-to-mid inferoseptum (arrows), with transmural (> 90%) enhancement of the apical segments adjacent to the thrombus.

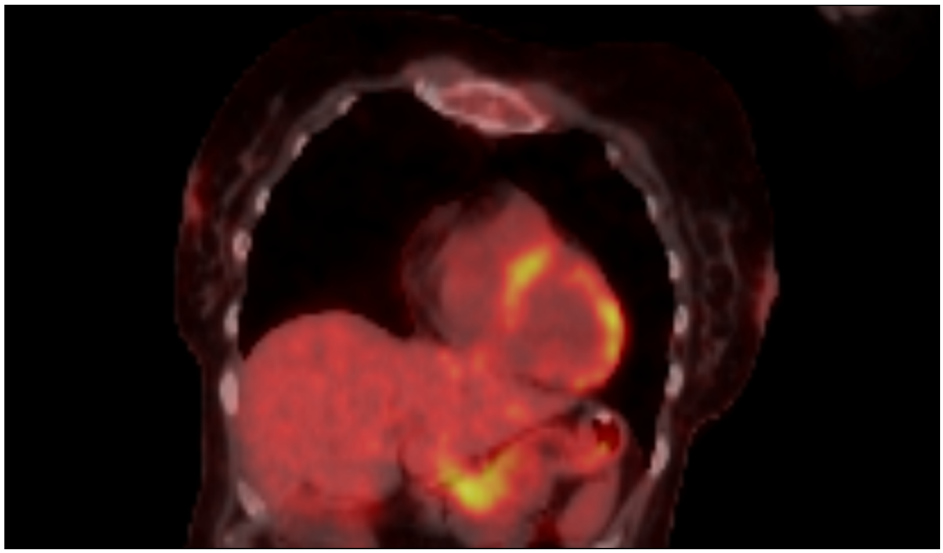


Figure 3 Fused fluorine-18 fluorodeoxyglucose (FDG) positron emission tomography-computed tomography image showing multifocal significant left ventricular myocardial FDG uptake, particularly involving the basal interventricular septum. The whole-body study showed no FDG-avid lymphadenopathy or other evidence of extracardiac sarcoidosis.

IgAV (purpura with renal involvement and IgA-dominant proliferative glomerulonephritis on kidney biopsy).[7]

Collectively, these findings established severe inflammatory perimyocarditis. The absence of extracardiac sarcoidosis, normal complement, negative ANCA and anti-glomerular basement membrane antibodies, and biopsy-proven IgA nephropathy with concurrent leukocytoclastic vasculitis and diffuse alveolar hemorrhage favored a systemic IgA vasculitis flare with multiorgan involvement as the unifying diagnosis.

The treatment course is summarized in Figure 4. Following bronchoscopy-confirmed alveolar hemorrhage, pulse methylprednisolone (1 g/day, days 2-4) was given, followed by intravenous methylprednisolone 60 mg/day (days 5-26) and then an oral prednisolone taper. Given the aggressive course and the left ventricular thrombus, catastrophic antiphospholipid syndrome was considered, and daily plasma exchange was performed on days 8 through 12 pending antibody results. Rituximab 1 g was administered in two doses, the first during day 18 and the second after discharge (day 37); the first was delayed by hypogammaglobulinemia and preceded on the same day by intravenous immunoglobulin. After the apical thrombus was identified, the competing risks of systemic embolization and recurrent pulmonary hemorrhage were balanced through multidisciplinary discussion; low-dose unfractionated heparin was cautiously initiated on day 8, held on days 12 through 14 for a hemoglobin drop without active bleeding, and escalated on day 15 to anti-factor Xa-guided therapeutic heparin with bridging to warfarin (target international normalized ratio 2-3).[8] Heart-failure therapy included diuresis and bisoprolol; renin-angiotensin system inhibitors, mineralocorticoid receptor antagonists, and sodium-glucose co-transporter 2 inhibitors were deferred given severely reduced renal function, and empiric antibiotics were discontinued once the infectious work-up remained negative.

The patient improved progressively. Hemoptysis resolved, and hemoglobin stabilized despite therapeutic anticoagulation. High-sensitivity troponin T fell from its peak of 20,208 ng/L to 48 ng/L at outpatient follow-up. Creatinine peaked at ~350 µmol/L and improved to 329 µmol/L at discharge; nephrology favored nonoliguric acute kidney injury superimposed on chronic kidney disease stage G4 A3. She was discharged on hospital day 30 on oral prednisolone with a planned taper, warfarin, bisoprolol, and diuretics, with outpatient cardiology and rheumatology follow-up. Follow-up echocardiography on December 15, 2025, 3 months after discharge, showed improved but persistent left ventricular dysfunction (ejection fraction 30-35%) with no residual thrombus and no pericardial effusion. Follow-up CT of the chest after 3 months of treatment showed significant improvement in the consolidative and ground-glass opacities.

DISCUSSION

Our patient's presentation posed several diagnostic challenges. Markedly elevated cardiac biomarkers, diffuse electrocardiographic abnormalities, and severe left ventricular dysfunction initially raised concern for acute coronary syndrome. Contemporary American College of Cardiology recommendations emphasize that myocarditis

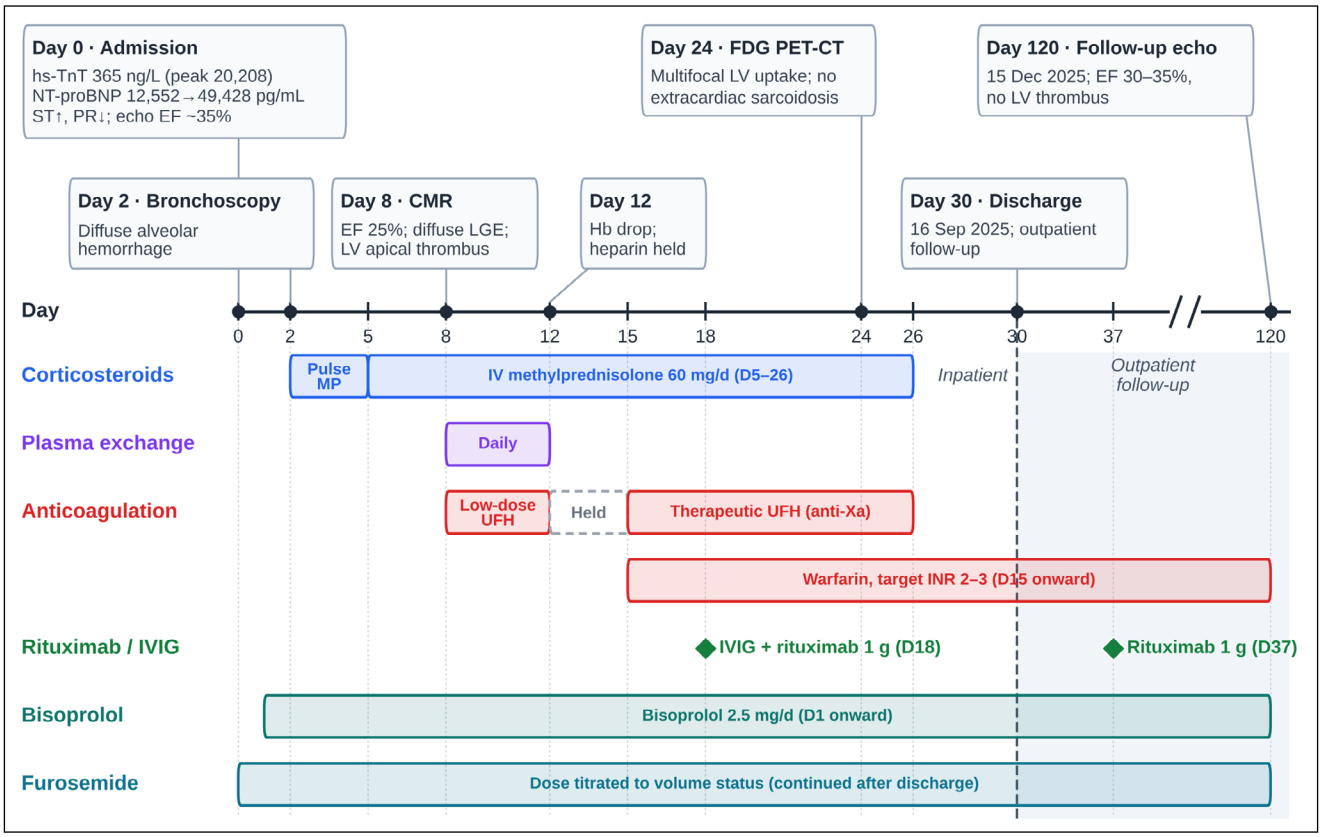


Figure 4 Timeline of clinical events and treatments during the index hospitalization (August 17 to September 16, 2025; admission = day 0) and outpatient follow-up (shaded). Bars show treatment periods and diamonds show single doses. Anti-Xa: anti–factor Xa activity; CMR: cardiac magnetic resonance; EF: ejection fraction; FDG PET-CT: fluorine-18 fluorodeoxyglucose positron emission tomography–computed tomography; Hb: hemoglobin; hs-TnT: high-sensitivity troponin T; INR: international normalized ratio; IVIG: intravenous immunoglobulin; LGE: late gadolinium enhancement; LV: left ventricular; MP: methylprednisolone; NT-proBNP: N-terminal pro–B-type natriuretic peptide; PR↓: PR-segment depression; ST↑: ST-segment elevation; UFH: unfractionated heparin

should be considered when such findings are accompanied by elevated biomarkers and supportive imaging.[9] Cardiac magnetic resonance (CMR) has become a pivotal noninvasive modality through tissue characterization based on T1 and T2 mapping. The updated 2018 Lake Louise criteria support a CMR diagnosis of active myocardial inflammation in the presence of at least one T2-based marker of myocardial edema (increased T2 relaxation time or T2-weighted signal) and at least one T1-based marker of myocardial injury (increased native T1, increased extracellular volume, or nonischemic late gadolinium enhancement)[10]; our patient fulfilled both. PET imaging may provide complementary information on active myocardial inflammation in selected patients.[9]

Although the transmural apical enhancement and left ventricular thrombus may suggest ischemic injury, inflammatory myocardial disease can demonstrate infarct-like patterns, particularly with microvascular injury and necrosis; because coronary angiography was deferred owing to acute kidney injury, concomitant ischemic injury could not be entirely excluded. Multifocal FDG uptake raised the possibility of cardiac sarcoidosis, but the absence of extracardiac involvement, biopsy-confirmed IgA-mediated disease, and concurrent systemic manifestations favored multisystem IgAV. The mechanisms underlying cardiac involvement remain incompletely understood but are thought to involve galactose-deficient IgA1 immune complexes with complement activation, endothelial injury, and inflammatory cell recruitment affecting the coronary microvasculature and myocardium.[1]

The coexisting diffuse alveolar hemorrhage further increased diagnostic complexity. Pulmonary involvement is an uncommon but potentially life-threatening manifestation of IgAV and should prompt careful exclusion of anti–neutrophil cytoplasmic antibody-associated vasculitis and antiglomerular basement membrane disease, particularly with concurrent pulmonary and renal involvement; negative serology and compatible histopathology supported IgAV in our

patient.[1] Management required balancing competing thrombotic and hemorrhagic risks, and the coexistence of recent alveolar hemorrhage with a large left ventricular thrombus created a therapeutic dilemma addressed through multidisciplinary collaboration with cautious anticoagulation under close monitoring.

Immunosuppressive therapy was guided by disease severity rather than established protocols. Current strategies for severe IgAV are largely based on observational data and expert opinion, with high-dose corticosteroids remaining the cornerstone; pulse methylprednisolone, cyclophosphamide, rituximab, and plasma exchange have all been reported in life-threatening disease despite the absence of randomized evidence.[3,11] In our patient, aggressive immunosuppression was associated with resolution of pulmonary hemorrhage, a progressive decline in cardiac biomarkers, stabilization of renal function, disappearance of the left ventricular thrombus, and partial recovery of systolic function. Clinically overt cardiac involvement in IgAV is exceptionally rare and likely underrecognized; in one large pediatric cohort, cardiac abnormalities, mostly asymptomatic biomarker elevation, were found in 12.3% of 1,451 children.[12] Published reports describing heterogeneous manifestations—myocarditis, pericarditis, hemopericardium, heart failure, myocardial infarction, and arrhythmias—are limited to isolated case reports and small series, and a review of reported cases suggested that cardiac involvement is more often fatal in adults than in children.[4,13] Our case expands this spectrum and underscores the need for a high index of suspicion for myocardial injury in systemic vasculitis.

CONCLUSION

This case illustrates severe multisystem adult IgA vasculitis with simultaneous cardiac, pulmonary, renal, and cutaneous involvement, an exceptionally rare and potentially fatal combination. Early recognition of cardiac involvement, integration of multimodality imaging—including cardiac magnetic resonance and PET—with the broader clinical context, and individualized, multidisciplinary management were central to establishing the diagnosis and guiding therapy and may contribute to improved outcomes in similarly complex inflammatory cardiovascular presentations.

COMPETING INTERESTS

The authors have no competing interests to declare.

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