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Fulminant Granulomatosis With Polyangiitis (GPA) Associated With Intracardiac Mass: A Case Report

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Abstract

Granulomatosis with Polyangiitis (GPA) affects nearly every organ system in the body. Cardiac involvement with GPA, particularly intracardiac masses, is a very unusual occurrence. The present case report demonstrates the diagnosis and successful treatment of an intracardiac mass in GPA with multiorgan involvement.

Keywords: Granulomatosis with Polyangiitis, rapidly progressing glomerulonephritis, cardiac mass, anti-neutrophil cytoplasmic antibodies, vascular thrombus

History of presentation

A 23-year-old male presented to a family physician's clinic with complaints of recurrent upper respiratory tract infections, bilateral eye congestion, gangrenous lesions on the toes of his right leg, andnodular skin lesions over both the elbows **(Figure 1).** On examination, the patient was anemic, anicteric, and had bilateral scleroconjunctival congestion. No regional lymphadenopathy or facial deformities were noted. He was hospitalized for further evaluation and management.

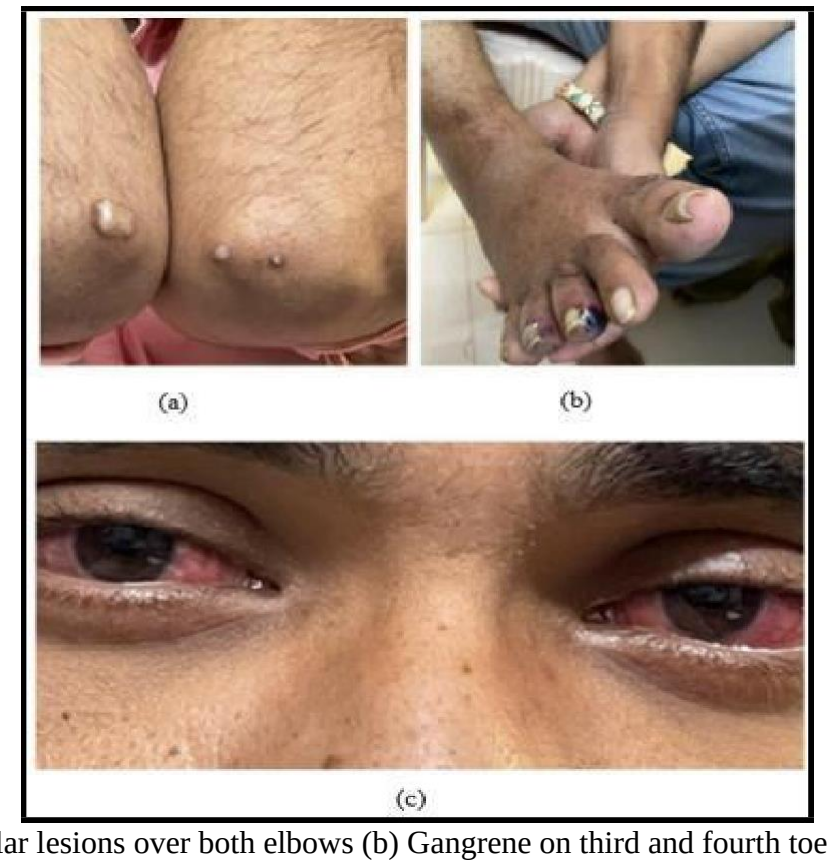

Figure 1: Clinical presentation (a) Nodular lesions over both elbows (b) Gangrene on third and fourth toe of right leg (c) Bilateral scleroconjunctival congestion.

Past Medical History

The patient had a history of sinusitis coupled with hemoptysis, spontaneous nasal bleeding, and mastoiditis unresolved with antibiotics. No records of drug exposure, drug addiction, or family history of similar diseases were noted.

Investigations

Preliminary laboratory analysis showed normocytic anemia, leukocytosis, and elevation in inflammatory markers. He was positive for anti-neutrophil cytoplasmic antibodies (ANCA) by indirect immunofluorescence and for anti-proteinase 3 antibodies by enzyme-linked immunosorbent assay (ELISA). However, anti-nuclear antibodies (ANA) and anti-DNA were negatives (**Table 1**).

Table 1: Summary of clinical investigations

Hematological parameter	**Result**	**Biological reference interval**
Hemoglobin (g/dL)	12.6	13-17
Hematocrit-packed cell volume (%)	40.1	40-50
Mean corpuscular volume (fL)	78.0	83-101
Mean corpuscular hemoglobin (pg)	24.7	27-32
White blood cell count (cells/cumm)	15100	4000-10000
ESR (mm/hr)	97	< 10
Creatinine (mg/dL)	1.2	Male: 0.9-1.3
C-reactive protein (mg/L)	**168**	**< 5.0**
Lupus anticoagulant aPTT (sec)	**Negative**	**NA**
Anti PR3 IgG	**218.71**	**Negative: < 20; weak positive: 21-30; moderate positive to strong positive: > 30**
Anti MPO IgG	3.13	Negative: < 20; weak positive: 21-30; moderate positive to strong positive: > 30
Anti-cardiolipin IgG (GPL U/mL)	Negative (0.30)	Negative: < 8.0; equivocal: 8.0-12.0; positive: >12.0
Anti-cardiolipin IgM (MPL U/mL)	Negative (0.14)	Negative: < 8.0; equivocal: 8.0-12.0; positive: >12.0
cANCA (1 in 10 dilution)	Negative	NA
cANCA (1 in 20 dilution)	**Positive**	**NA**
ANA	Negative	-
Anti-native DNA	Negative	-
Tuberculosis QuantiFERON	Negative	-

Abbreviations: ANCA, anti-neutrophil cytoplasmic antibody; ANA, anti-nuclear antibody; aPTT, activated partial thromboplastin time; cumm, cubic millimeter; dL, deciliter; DNA, deoxyribonucleic acid; fL, femtoliter; g, gram; GPL U, a microgram of IgG antibody; hr, hour; IgG, immunoglobulin G; IgM, immunoglobulin M; ESR, erythrocyte sedimentation rate; L, liter; mg, milligram; mL, milliliter; mm, millimeter; MPL U, a microgram of IgM antibody; pg,

Excision biopsy of the skin lesion on the elbows depicted necrobiotic interstitial granulomatous dermatitis with sinus tract formation and suppuration. Biopsies of skin lesions indicated that they were a part of a systemic connective tissue disease and not infection. Computed tomography (CT) of the paranasal sinuses (**Figure 2**) showed left maxillary sinusitis, minimal mucosal thickening in the right maxillary sinus, and deviated nasal septum to the left. Concha bullosa of the left middle turbinate opacified osteomeatal complex on the left side, soft tissue density in bilateral mastoid air cells, and middle ear otomastoiditis were also noted.

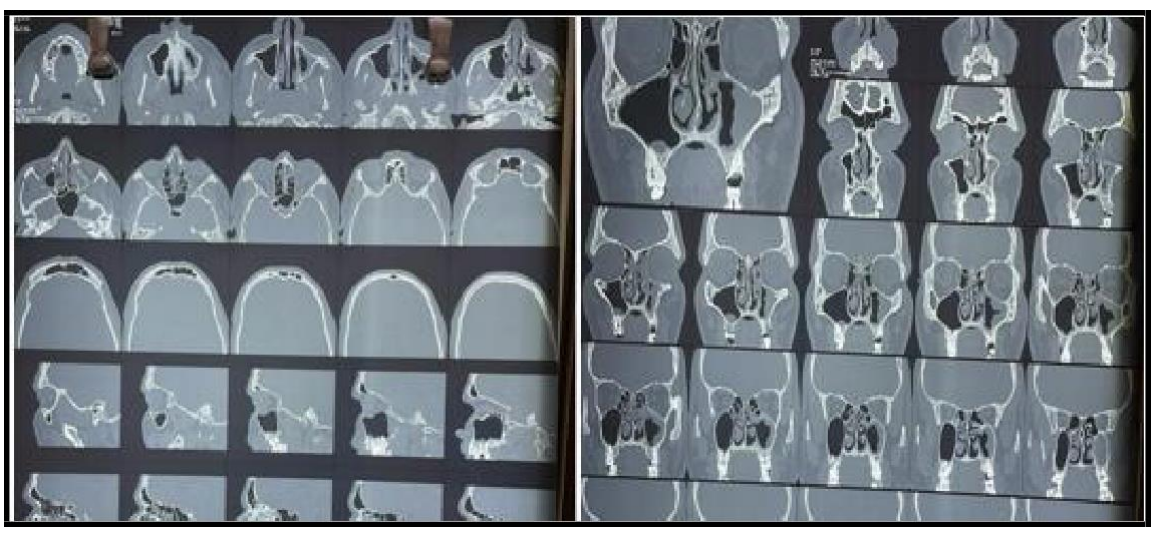

Figure 2: Computed tomography (CT) of the paranasal sinuses

Thoracic and trans oesophageal echocardiography (ECHO) revealed a large mobile intracardiac right ventricular (RV) mass, measuring 0.5 x 2.4 cm. The mass appeared soft, non-vascular, and non-calcified in RV inflow extending up to outflow. Further, cardiac magnetic resonance imaging (MRI) detected 3 small, highly mobile masses, each measuring 1 x 6-7 cm, attached to tricuspid valve chordae tendinae **(Figure 3)**.

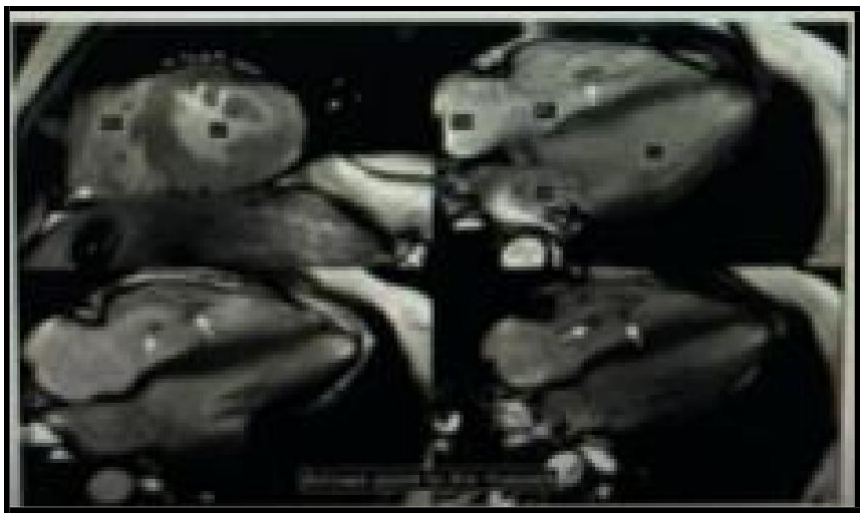

Figure 3: Cardiac MRI

Rising creatinine levels over the next couple of weeks indicated rapid deterioration of patient's renal function, which led to renal biopsies. Biopsy findings showed necrotizing and pauci-immune crescentic glomerulonephritis, consistent with ANCA associated vasculitis **(Figure 4)**.

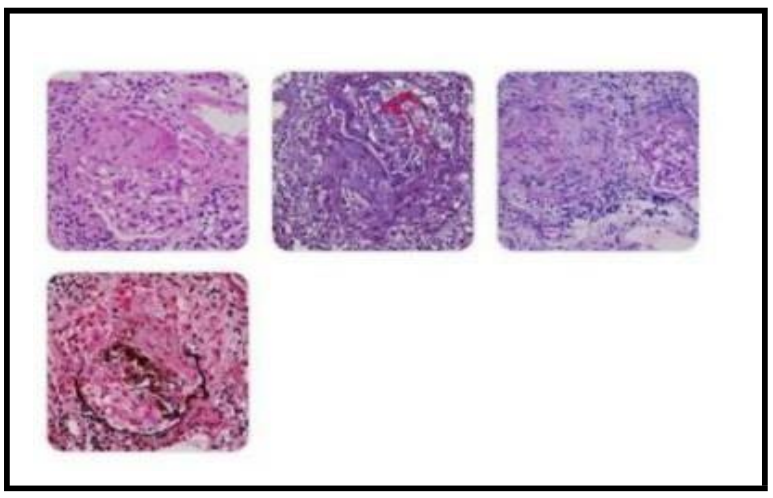

Figure 4: Renal biopsy, necrotizing and crescentic glomerulonephritis, pauci-immune, consistent with ANCA associated vasculitis

The patient developed swelling in right foot with cellulitis and progressive worsening of gangrene. Complete thrombus of right tibial artery was seen, along with cellulitis on the right foot, with an acute thrombus causing complete vascular lumen obliteration.

Subsequently, the patient was readmitted following complaints of chest pain and respiratory distress. ECHO was repeated, Doppler revealed deep vein thrombosis, and CT pulmonary angiography confirmed pulmonary embolism in the patient **(Figure 5)**.

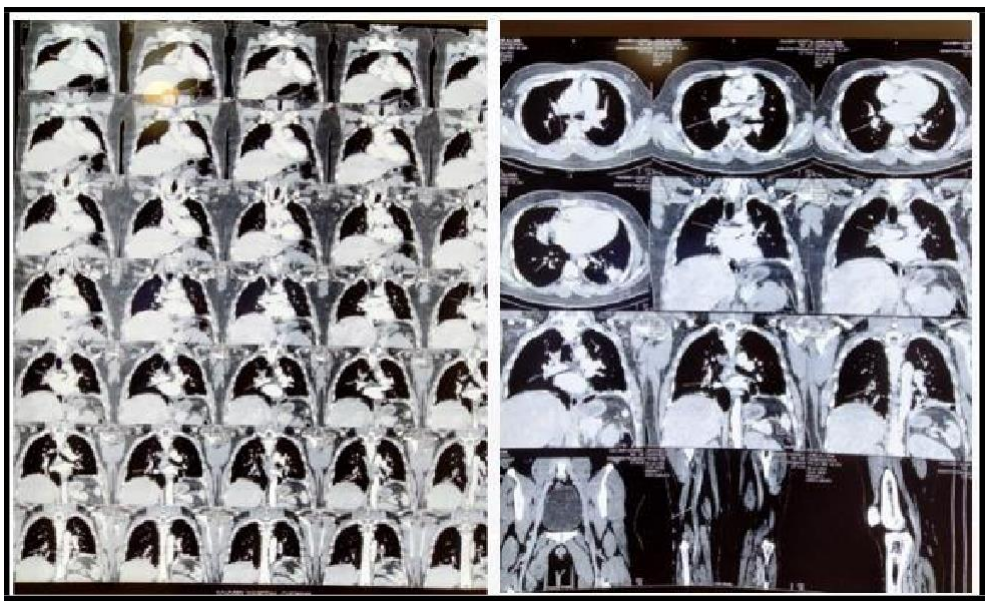

Figure 5: CT scan showing pulmonary embolism

Differential diagnosis

Cardiac MRI characterized the mass as a pedunculated, narrow base with free movement, multiple, and attached to the chordae tendinae of the tricuspid valve without any edema. These features did not align with adipose tissue myxoma, malignancy, or granuloma. Therefore, the intracardiac mass was most likely a thrombus or fibroelastoma.

Blood cultures taken during the patient's admission were reported negative, excluding infective endocarditis. The following conditions were ruled out based on patient history and clinical presentation: antiphospholipid syndrome (no history of blackish discoloration of the limbs, pain, or swelling); tuberculosis (absence of fever, night

sweats, loss of weight, and negative TB quantiferron); congestive cardiac failure (no history of breathlessness on exertion, lying down, no fatigue, or nocturnal cough) and conduction disorders (no history of syncope, palpitations, and chest pain). Despite the ocular and dermal changes, sarcoidosis was ruled out due to the absence of lymphadenopathy.

Management

The patient was treated with immunosuppressive medications comprising cyclophosphamides and corticosteroids. Plasmapheresis was initiated, followed by treatment with anticoagulants for deep vein thrombosis (DVT) and pulmonary embolism (PE), along with supportive care. With treatment, he recovered his renal function, and venous thrombosis was resolved with the complete disappearance of the intracardiac mass without any surgical intervention **(Table 2)**.

Table 2: Clinical investigations and interventions

Investigation	Intervention
GPA and scleritis	Corticosteroid and cyclophosphamide
Intracardiac mass	Dalteparin 5000 IU
Rise in creatinine levels	Plasmapheresis, with 5 cycles done every 3 days
Anterior tibial artery obstruction	Dalteparin 5000 IU

Abbreviations: g, gram; GPA, granulomatosis with polyangiitis; IU, international units; mg, milligram

Discussion

GPA affects nearly every organ system in the body, with the respiratory and renal systems being the most affected. Symptoms can range from mild conjunctivitis and episcleritis to more severe inflammation such as keratitis, scleritis, uveitis, and retinal vasculitis. Ocular involvement was recorded in about 50-60 % of cases, with 8-16 % of patients presenting at the time of diagnosis **[1]**. ENT manifestations are frequently the initial symptoms, with sinonasal symptoms (52 %) being the most common, followed by otological indications (32 %) **[2]**. Skin problems like petechiae or purpura (16 %), painful skin lesions (9.4 %), and maculopapular rash (6.7 %) were most prevalent in GPA **[3]**. According to Bünyamin Kisacik et al. study, unusual symmetrical polyarthritis of the small joints was observed in patients with GPA, showing the impact of GPA on joints **[4]**.

Cardiac involvement with GPA has been reported, and mostly includes pericarditis (50 %), myocarditis (25 %), endocarditis (21 %), and conduction system anomalies (17 %) as the most common abnormalities. However, the occurrence of cardiac masses in GPA is rare. The case discussed is unique, as it involves multiple systems and includes an intracardiac mass. Based on the patient's presentation, history of repeated upper respiratory tract infections, and chronic sinusitis, vasculitis, positive ANCA **[5]**, renal and cardiac involvement, a diagnosis of GPA was made.

The patient had a rare cardiac manifestation of GPA, which presented itself as three extremely mobile masses linked to the tricuspid valve. Following a cardiac MRI, aberrant tissue was identified as three tiny masses that were cleared following dalteparin treatment. This demonstrates that all intracardiac masses are not atrial myxomas and can be treated without surgery. The intracardiac mass of the current case was a thrombus, which was eliminated with an anticoagulant. As thrombus is present, GPA should be considered in the differential diagnosis of any intracardiac masses.

Follow-up

No further complications related to GPA or reappearance of intracardiac mass were noted on the follow-up consultation 3 months post-discharge. Presently, the patient is on oral corticosteroids and monthly cyclophosphamide and is doing well.

Conclusions

This case demonstrates a unique medical approach taken for diagnosisand successful treatment of an intracardiac mass in GPA with multiorgan involvement. The presence of an intracardiac mass in GPA is a rare entity and this condition is often linked with poor prognosis, but timely intervention and accurate diagnosis can result in better clinical outcomes.

Abbreviations:

ANA = anti-nuclear antibodies

ANCA = anti-neutrophil cytoplasmic antibodies

CT= computed tomography

DVT = deep vein thrombosis

ECHO = echocardiography

GPA = Granulomatosis with Polyangiitis

MRI= magnetic resonance imaging

PE = pulmonary embolism

RPGN = rapidly progressive glomerulonephritis

SLE = systemic lupus erythematosus

Learning Objectives

- To identify a case of GPA, a rare systemic necrotizing vasculitis, in a fulminant life-threatening course.
- To recognize that intracardiac masses other than myxoma can be a presentation in GPA, which can be resolved by non-surgical means.
- To demonstrate that in patients with intracardiac masses, the possibility of GPA cannot be excluded from the diagnosis.

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