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Fatal Cardiac Involvement Revealing Granulomatosis with Polyangiitis

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Abstract Granulomatosis with polyangiitis (GPA), formerly known as Wegener's disease, is a rare necrotizing and granulomatous systemic vasculitis of small vessels. Cardiac involvement in GPA is unusual and the heart is part of the so-called "atypical" locations of this vasculitis. This complication is often little-known and overlooked by clinicians despite its severity and high mortality. Symptomatic acute myocardial infarction is an exceptional presentation of cardiac involvement associated to GPA. We report an original observation of fatal cardiac damage revealing GPA in a 50-year-old Tunisian man with no pathological medical history. The diagnosis was made on post-mortem examination.

Keywords: *granulomatosis with polyangiitis, Wegener's disease, cardiac involvement, myocardial infarction, sudden death, vasculitis*

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1. Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's disease, is a necrotizing and granulomatous systemic vasculitis of small vessels [1]. It is characterized by predominant Ear/Nose/Throat (ENT), pulmonary, and renal involvement; but can spread to all organs and tissues with a large clinical polymorphism [2,3]. Antineutrophil cytoplasmic antibodies with cytoplasmic fluorescence pattern (ANCA-c) and anti-proteinases 3 specificity (anti-PR3) represent the specific immunologic signature of this autoimmune vasculitis (ANCA-associated vasculitis); they are noted in 90% of the generalized forms of this vasculitis [4]. GPA is potentially serious with high mortality and frequent fatal forms [4].

Cardiac involvement is rare and often little-known and overlooked by clinicians [5,6]. It is characterized by its severity and its particularly high mortality and therefore conditions the prognosis of this disease [6,7,8].

We report an original observation of fatal cardiac damage revealing GPA.

2. Case Report

A 50-year-old Tunisian man with no pathological medical history suddenly died in the morning upon his arrival at work. There was no evidence of trauma, stress, consumption of any toxic substance or any particular medication.

The somatic examination was without apparent abnormalities. Autopsy examination noted a right pleural fluid effusion of moderate abundance, congestive lungs (right lung=990 g and left lung-980 g), a 450g globular heart with left ventricular hypertrophy, right atrial dilatation, and interventricular septal congestion (Figure 1) with a mottled appearance alternating areas of hyperemia and whitish scar areas (Figure 2).

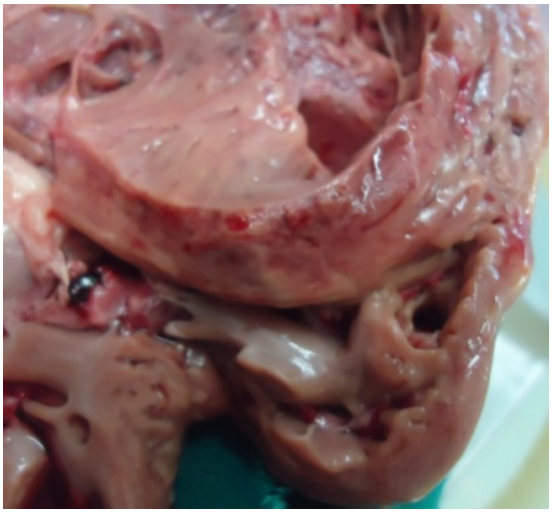

Figure 1. Hyperemia of the interventricular septum

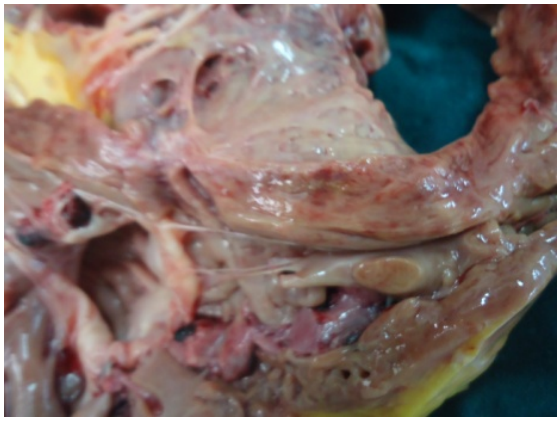

Figure 2. Mottled appearance of the interventricular septum with areas of hyperemia and whitish scar areas

The search for drugs and toxins in the blood and gastric fluid was negative (alcohol, salicylates, tricyclic antidepressants, carbamates, benzodiazepines, phenothiazines, pesticides, organophosphorus, organochlorines).

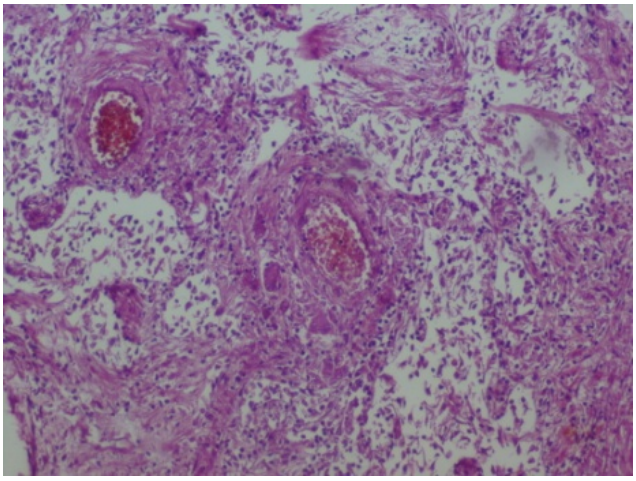

Figure 3. Histological examination (H&E×200): Granulomatous inflammation of lung tissue

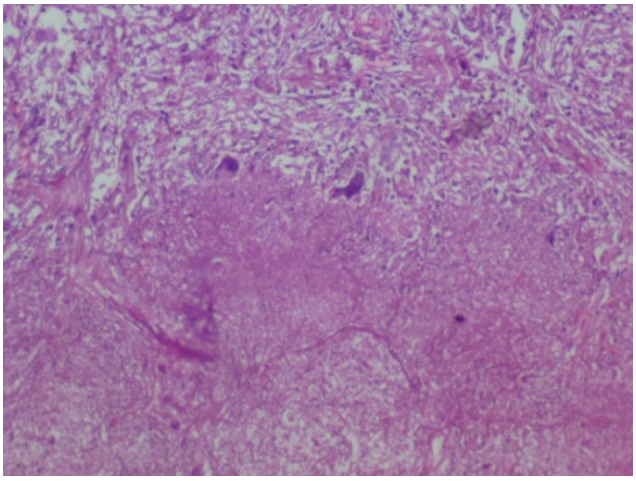

Figure 4. Histological examination (H&E×200): Fibrinoid necrosis zones of lung tissue

The histological exam revealed diffuse granulomatous inflammation of the lungs with fibrinoid necrosis and necrotizing vasculitis of the small and medium-sized arteries (Figure 3, Figure 4, Figure 5 and Figure 6), coronary vasculitis, fibrous scar of an old basal myocardial infarction (Figure 7); also an area of acute and extensive anterior myocardial necrosis (Figure 8). The post-mortem ANCA test was strongly positive.

These lesions were consistent with the diagnosis of generalized GPA with pulmonary, pleural, and cardiac involvement. The direct cause of death was anterior acute myocardial infarction secondary to GPA-specific coronary artery vasculitis.

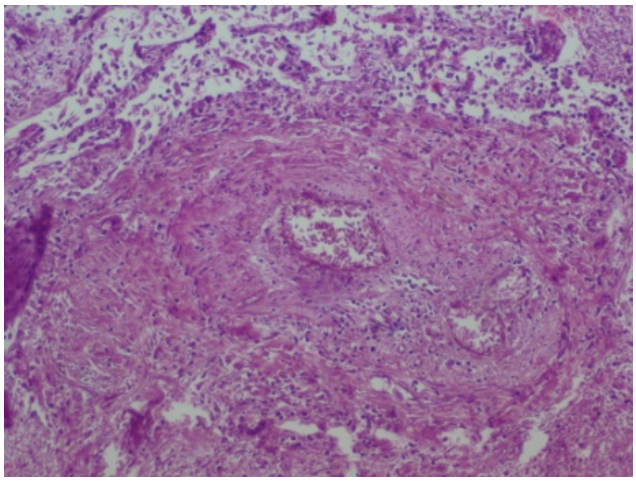

Figure 5. Histological examination (H&E×200): Pulmonary vasculitis

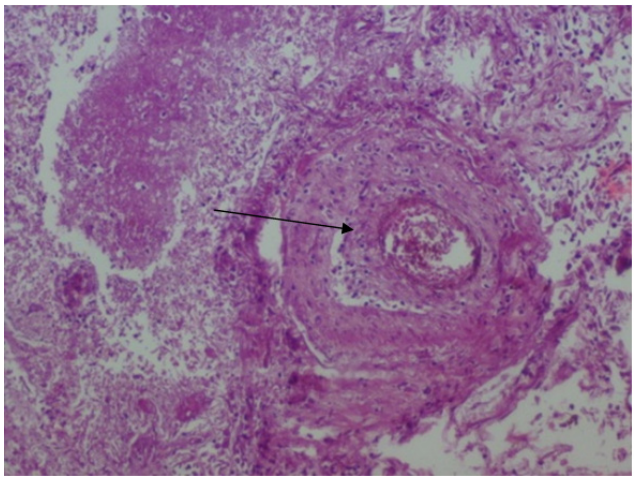

Figure 6. Histological examination (H&E×200): Necrosis zone with pulmonary granulomatous vasculitis (arrow)

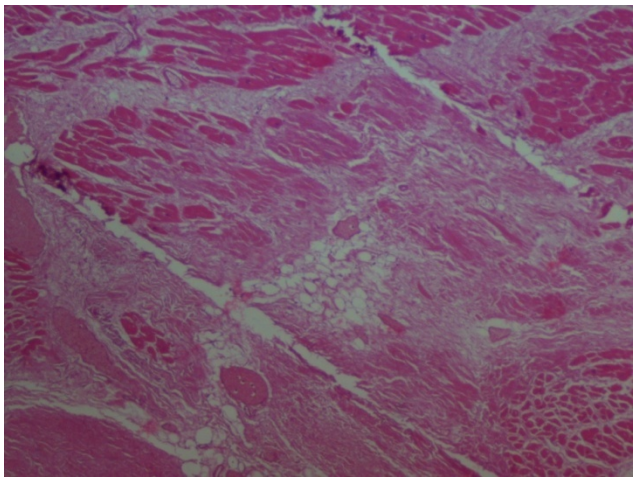

Figure 7. Histological examination (H&E×200): Fibrous scar tissue of myocardial infarction

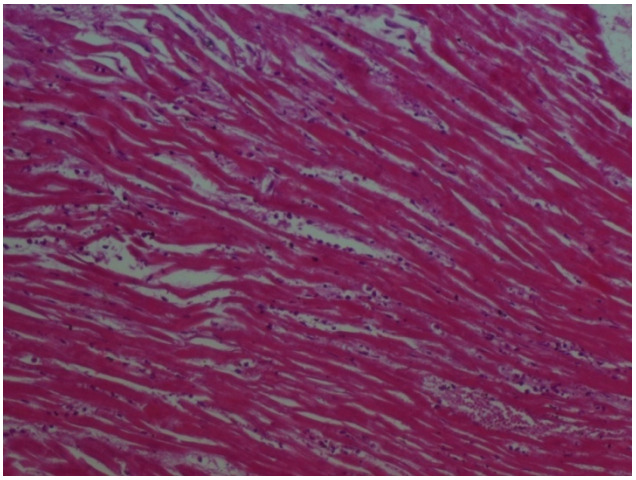

Figure 8. Histological examination (H&E×200): Recent myocardial necrosis: necrotic myocardial cells with presence of inflammatory neutrophils infiltrate

3. Discussion

Cardiac involvement during GPA remains rare and unusual [5,9] and the heart is part of the so-called "atypical" locations of this vasculitis [3,10]. Indeed, it has been clinically reported in 4 cases in the series of Di Comite G et al of 50 patients followed for GPA (8%) [11], in 5.71% of cases in the Indian series of Sharma A et al of 105 patients with GPA [12], and only in 17 cases (3.3%) in the large American series of 517 patients with GPA (Vasculitis Clinical Research Consortium Longitudinal Study of GPA) [13].

This involvement seems however to be very underestimated since a large discrepancy has been reported between the frequencies of reported symptomatic cases of this location and the post-mortem examination data [6,14]. Indeed, when it's systematically investigated by cardiac magnetic resonance imaging, cardiac involvement was objectified in 61% of patients treated for GPA in the Pugnet G et al series [15].

Cardiac involvement of GPA may be clinically manifested by: myocarditis, acute pericarditis, constrictive pericarditis, fibroblastic endocarditis, coronaritis/coronary vasculitis, valvulitis, arrhythmia, congestive heart failure, dilated cardiomyopathy, and complete atrioventricular block [5,6,9,14,16,17,18].

Acute myocardial infarction with clinical expression is an exceptional complication of GPA [9]. It is most often silent [14] and fatal [14,19,20] in both adults and children [21]. This often silent character of cardiac disease associated with GPA (25% of cases for Sarlon G et al [6]) explains the significant delay in diagnosis and the fact that in the majority of cases, the diagnosis is made at autopsy; indeed post-mortem examination could objectify coronary arteritis in up to 50% of cases of GPA [22].

Despite its histologically proven high frequency and potential severity, cardiac involvement remains largely unknown and often overlooked by medical practitioners making undiagnosed GPA a cause of natural sudden death [23].

Cardiac localization of GPA requires early diagnosis and adapted rapid management [5] since the evolution is usually favorable with rapidly systemic corticosteroids and immunosuppressive drugs (particularly cyclophosphamide) [9]. More recently, biotherapy, particularly rituximab, has been shown to be very effective in treating this localization [10,24].

Thus, and given the large clinical polymorphism, the potential severity, and the often asymptomatic nature of this location; some authors recommend systematic and regular cardiac monitoring in any patient with GPA [6]. It is also recommended that GPA be considered among the differential diagnoses of any non-specific illness with cardiac involvement [17].

4. Conclusion

Cardiac involvement during GPA is histologically frequent but is rarely symptomatic. It remains little-known and often neglected in current medical practice. It is characterized by its gravity and its often fatal evolution.

The heart disease of GPA deserves to be well-known by clinicians in order to diagnose it in time and avoid therefore the high risk of associated sudden death.

Conflicts of Interest

None.

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