

Wegener's granulomatosis with very unusual presentation

Mansur Somaily, Abdurhman S Al Arfaj

ABSTRACT

Introduction: Wegener's granulomatosis (WG) involves mainly upper and lower respiratory tract and the kidneys. Cardiac involvement is not common. **Case Report:** This is a report of a 34-year-old, non-smoker male with very unusual presentation of (WG) namely a combination of large pericardial effusion, cardiac tamponade (without uremia) and conduction defects, all of which responded very well to immunosuppressive therapy without the need for surgical intervention. **Conclusion:** Cardiac complication of WG may start early and early diagnosis and management improves the outcome and decrease, overall mortality.

Keywords: Wegener's granulomatosis, Cardiac involvement, Pericardial effusion, Cardiac tamponade

Somaily M, Arfaj ASA. Wegener's granulomatosis with very unusual presentation. International Journal of Case Reports and Images 2012;3(12):47–51.

doi:10.5348/ijcri-2012-12-237-CR-11

INTRODUCTION

Wegener's granulomatosis is one of the most common forms of systemic vasculitis, with a reported annual incidence of 10 cases per million. The disease involves small and medium-sized blood vessels, and mainly affects the upper and lower respiratory tract and kidneys [1].

Cardiac involvement is reported to occur in 6–44% cases of Wegener's granulomatosis [2–5]. Pericarditis is the most common cardiac manifestation accounting for about 50% of cardiac diseases in Wegener's granulomatosis, which is asymptomatic in most of the cases, or may be manifested by chest pain and dyspnea [2–5].

Pericarditis and coronary vasculitis are the most frequent findings but myocarditis, endocarditis, valvulitis, and conduction system defects are also described [2–7]. Pericardial effusion reported in Wegener's granulomatosis can be due to the disease itself or due to uremia in cases of renal failure [7, 8]. We are reporting here a very unusual presentation of Wegener's granulomatosis which is a large pericardial effusion (without evidence of uremia), and cardiac tamponade combined with a conduction defect. This combined presentation has not been reported previously.

CASE REPORT

Our patient is a 34-year-old non-smoker male, who was admitted to the emergency room with history of chest pain and cough for five days, and fever for one month. His cough was mostly productive of whitish sputum but he had five attacks of hemoptysis of fresh blood of moderate amount (about 10 mL per day). He

Mansur Somaily¹, Abdurhman S Al Arfaj²

Affiliations: ¹(SB-Med), Department of Medicine, College of Medicine and King Khalid University Hospital, King Saud University, Riyadh; ²(FRCP(C)), Department of Medicine, College of Medicine and King Khalid University Hospital, King Saud University, Riyadh.

Corresponding Author: Prof. Abdurhman Saud Al Arfaj, Professor and Consultant, Rheumatology division, Department of Medicine, College of Medicine, King Saud University, Riyadh, KSA; Ph: 00966 1 4671510; Email: asarfaj@ksu.edu.sa

Received: 08 May 2012

Accepted: 22 July 2012

Published: 01 December 2012

also had one month history of arthralgia and daily epistaxis. He had no history of weight loss, nor contact with any patient of tuberculosis.

When he presented, he had a temperature of 37.8°C, but was hemodynamically stable. Examination of the nose revealed a nasal ulcer and tenderness over the left maxillary sinus. Skin, chest, cardiovascular and musculoskeletal examination were unremarkable. The initial differential diagnosis included: pulmonary tuberculosis, pneumonia, acute bronchitis and WG. His initial laboratory investigations showed WBC count $11 \times 10^3/L$, Hb 13.6 g/dL, platelets $560 \times 10^3/\mu L$, urea 5.3 mmol/L and creatinine 61 $\mu\text{mol/L}$. His erythrocyte sedimentation rate (ESR) was 120 mm/1st hr, aspartate aminotransferase (AGOT) 50 U/L, alanine aminotransferase (SGPT) 101 U/L, alkaline phosphatase 198 U/L and gamma glutamyl transferase (GGT) 232 U/L. His bilirubin, PT and APTT were normal. Cytoplasmic antineutrophil cytoplasmic antibody (cANCA) was 1:160 (normal range <1:40), and anti-proteinase 3 (anti-PR3) was 97 U/mL. Anti-myeloperoxidase (anti-MPO) and ANA were negative. Urine analysis showed hematuria with RBCs of 80/hpf, no WBCs and dipstick examination revealed 1+ positive protein. The 24-hour urine collection for protein was 0.5 g/day. Tuberculin test (5 units) was negative and sputum culture for acid fast bacilli was also negative. Chest X-ray was reported to be normal (Figure 1), but computed tomography (CT) scan of the lungs showed multiple nodules in both upper lobes, one of which was cavitating (Figure 2). CT scan of sinuses showed thickening of lining of left maxillary sinus. A nasal biopsy was obtained and showed necrotizing granulomatosis, consistent with diagnosis of WG. After two weeks of work-up the patient was diagnosed as WG based on clinical picture, cANCA positivity and tissue biopsy result. The patient was started on oral prednisolone 1 mg/kg/day and given one gram of intravenous cyclophosphamide. On the next day, the patient developed severe chest pain, dyspnea and hemoptysis. On examination, he was found to have high jugular venous pulse (JVP) with positive Kussmaul's sign, pulsus paradoxus and distant heart sounds. The chest X-ray at that time showed flask shaped heart (Figure 3). Echocardiography and CT scan of chest showed large pericardial effusion tamponade was thought to be most likely due to cardiac involvement by WG and as such, he was switched from oral prednisolone to one gram of intravenous pulse methylprednisolone. The next day patient developed light-headedness and dizziness, and was found to have bradycardia, with heart rate dropping down to 32 beats per minute (Figure 5). He was assessed by the cardiology team who advised continuous chronotropic monitoring and bed rest. He was continued on intravenous methylprednisolone one gram daily for three days which was reduced to 30 mg three times daily for two weeks and then switched to oral prednisolone which was tapered down to zero over the following six months. The heart rate returned to normal after 12 days.

Two weeks after the first dose of cyclophosphamide, the patient was asymptomatic, with normal heart rate and complete resolution of pericardial effusion on echocardiography. He was discharged home on prednisolone, to be admitted for further five pulses of intravenous cyclophosphamide. He remained free of symptoms at follow-up.

DISCUSSION

Echocardiographic abnormalities were found in 80% patients with Wegener's granulomatosis but lesions were considered as related to Wegener's granulomatosis



Figure 1: Chest X-ray was reported to be normal.

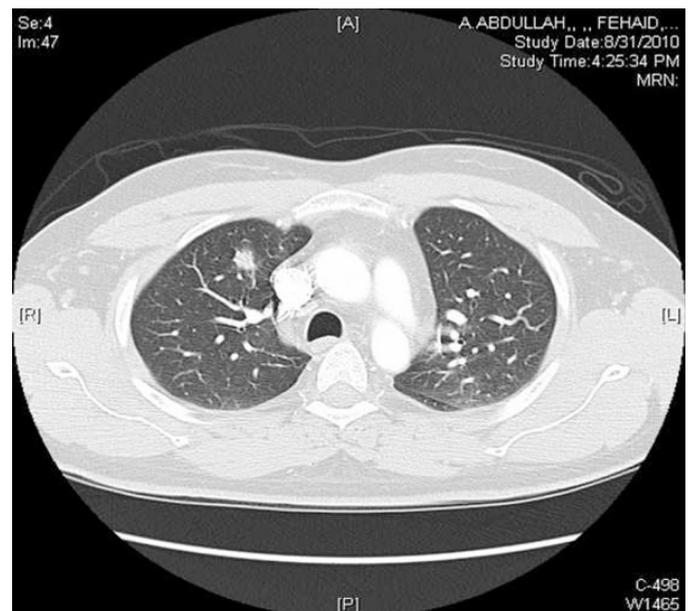


Figure 2: Computed tomography (CT) scan of the lungs showing multiple nodules in both upper lobes, one of which was cavitating.



Figure 3: Chest X ray showing flask shape heart.



Figure 4: CT scan chest, showing large pericardial effusion.

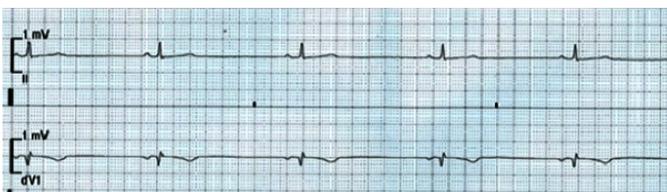


Figure 5: ECG demonstrating bradycardia, with heart rate of 32 beats per minute.

only in one-third of the patients [9]. Pericardial involvement represents almost 50% of cardiac involvement of Wegener's granulomatosis [5]. These are usually asymptomatic and carry a good prognosis.

However some cases may lead to considerable morbidity and mortality. The overall mortality rate of Wegener's granulomatosis with cardiac involvement has been reported to be between 15–45% [9].

Most of Wegener's granulomatosis patients reported previously with pericardial tamponade had hemodialysis dependant renal failure resulting in constrictive pericarditis [2–5, 7, 8]. However, massive pericardial effusion without constrictive pericarditis or uremia were reported in two cases [3, 4].

The peculiar occurrence in our patient of large pericardial effusion and tamponade combined with conduction defect in the absence of any renal affection, has not been reported previously in any Wegener's granulomatosis patients. Cardiac tamponade in patient with Wegener's granulomatosis and renal failure are described before [2, 7]. Pericardial effusion per se is not so rare as a cardiac manifestation of Wegener's patients [10]. In a series of eleven Wegener's granulomatosis patients, who had transthoracic echocardiography (TTE) and cardiac magnetic resonance (CMR), pericardial effusion was observed in five patients, while localized pericardial thickening was seen in six patients [11]. Similarly, mild pericardial effusions were described in five out of nine Wegener's patients [5]. However, the degree of inflammation did not reach the stage of excessive effusion leading to tamponade. Conduction defects are also described in cases of Wegener's granulomatosis [12, 13]. However, the combination of both cardiac tamponade and conduction defects in the absence of uremia have not been described previously. We believe that the extreme intensity of the inflammatory process of Wegener's granulomatosis in our patient resulted in a such an unusual combination of rare manifestations. This inflammatory process may be severe and lead to permanent damage. This was reported by Ohkawa et al., in a 61-year-old female with Wegener's granulomatosis who developed by recurrent episodes of ventricular tachycardia developing bradycardia. Her autopsy revealed generalized necrotizing angitis and severe granulomatous inflammatory foci affecting the common bundle of His and right bundle branch in addition to the myocardium [14].

We do not believe that the occurrence of such combined presentation was due to a delay in the diagnosis or instituting treatment, as it took only six weeks to do so. On the contrary, it could be considered early diagnosis and treatment, as one of the largest series on Wegener's granulomatosis reported that the median and mean period from disease onset to diagnosis of Wegener's granulomatosis were 4.7 and 15 months, respectively [15]. That report found only ten cases of pericarditis among 158 cases of Wegener's granulomatosis [6%] and only one patient had large effusion requiring surgical intervention [15]. Some authors recommend echocardiographic screening examination for all patients with active Wegener's granulomatosis [9].

It is also worth noting that, the cardiac involvement in our patient responded very well to

immunosuppressive therapy alone with complete resolution of pericardial effusion and return of heart rate to normal and without the need for surgical or invasive procedures.

CONCLUSION

Our case serves as an example that cardiac complications of Wegener's granulomatosis may start early and emphasizes the need for early diagnosis of cardiac involvement and their management, so that patient outcome can be improved and overall mortality decreased.

Author Contributions

Mansur Somaily – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Abdurhman S Al Arfaj – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© Mansur Somaily et al. 2012; This article is distributed under the terms of Creative Commons Attribution 3.0 License which permits unrestricted use, distribution and reproduction in any means provided the original authors and original publisher are properly credited. (Please see www.ijcasereportsandimages.com/copyright-policy.php for more information.)

REFERENCES

1. Leonard H Calabrese. Antineutrophil Cytoplasmic Antibody–Associated Vasculitis :In Kelley's textbook of rheumatology/[edited by] Gary S Firestein et al. 8th ed; 2008 W.B. Saunders Company.
2. Grant SC, Levy RD, Venning MC, Ward C, Brooks NH. Wegener's granulomatosis and the heart. *Br Heart J* 1994;71(1):82–6.
3. Koyalakonda SP, Krishnan U, Hobbs WJ. A rare instance of multiple valvular lesions in a patient with Wegener's granulomatosis. *Cardiology* 2010;117(1):28–30.
4. Goodfield NE, Bhandari S, Plant WD, Morley-Davies A, Sutherland GR. Cardiac involvement in

- Wegener's granulomatosis. *Br Heart J* 1995;73(2):110–5.
5. Morelli S, Gurgo di Castelmenardo AM, Conti F, et al. Cardiac involvement in patients with Wegener's granulomatosis. *Rheumatol Int* 2000;19(6):209–12.
 6. Attaran S, Desmond M, Ratnasingham J, Scawn N, Pullan DM. Mitral valve involvement in Wegener's granulomatosis. *Ann Thorac Surg* 2010;90(3):996–7.
 7. Yildizer K, Paydas S, Serin E, Sagliker Y. Wegener's granulomatosis complicated by pericardial tamponade and renal failure. *Nephron* 1996;72(2):339–40.
 8. Meryhew NL, Bache RJ, Messner RP. Wegener's granulomatosis with acute pericardial tamponade. *Arthritis Rheum* 1988;31(2):300–2.
 9. Oliveira GH, Seward JB, Tsang TS, Specks U. Echocardiographic findings in patients with Wegener Granulomatosis. *Mayo Clin Proc* 2005 Nov;80(11):1435–40.
 10. Korantzopoulos P, Papaioannides D, Siogas K. The Heart in Wegener's Granulomatosis. *Cardiology* 2004;102(1):7–10.
 11. Miszalski-Jamka T, Szczeklik W, Sokolowska B, et al. Cardiac involvement in Wegener's granulomatosis resistant to induction therapy. *Eur Radiol* 2011 Nov;21(11):2297–304.
 12. Forstot JZ, Overlie PA, Neufeld GK, Harmon CE, Forstot SL. Cardiac complications of Wegener granulomatosis: a case report of complete heart block and review of the literature. *Semin Arthritis Rheum* 1980 Nov;10(2):148–54.
 13. Ghaussy NO, Du Clos TW, Ashley PA. Limited Wegener's granulomatosis presenting with complete heart block. *Scand J Rheumatol* 2004;33(2):115–8.
 14. Ohkawa S, Miyao M, Chida K, et al. Extensive involvement of the myocardium and the cardiac conduction system in a case of Wegener's granulomatosis. *Jpn Heart J* 1999 Jul;40(4):509–15.
 15. Hoffman GS, Kerr GS, Leavitt RY, et al. Wegener granulomatosis: an analysis of 158 patients. *Ann Intern Med* 1992;116(6):488–98.

Access full text article on
other devices



Access PDF of article on
other devices

