

# Complete Heart Block due to Limited Wegener's Granulomatosis: A Case Report

SA Jahed<sup>1</sup>, M Zabihyeganeh<sup>2</sup>

<sup>1</sup>Department of Internal medicine, Booali General Hospital, Islamic Azad University, Tehran Medical Branch, <sup>2</sup>Department of Rheumatology, Firouzgar General Hospital, Tehran University of Medical Sciences, Tehran, Iran

Complete heart block is a rare manifestation of various cardiac involvements seen in Wegener's granulomatosis. To our knowledge there have only been two previous cases of complete heart block reported in patients with limited form of Wegener's granulomatosis in English literature. We describe a 62-year-old Iranian woman presenting with complete heart block who was candidate for pacemaker insertion. Limited Wegener's granulomatosis was suspected because of bloody nasal discharge, fatigue, fever, arthralgia, dacryocystitis, and history of recurrent subglutic stenosis, but no renal involvement. The diagnosis was subsequently confirmed by presence of granuloma in nasal mucosal biopsy and also high level of serum anti-proteinase 3. Prednisolone and cyclophosphamide therapy was started and her electrocardiography returned to normal sinus rhythm. Awareness of unusual presentation of Wegener's granulomatosis is important for proper diagnosis and management of the potentially fatal cardiac involvement of the disease. Complete heart block may be reversed by appropriate medical therapy of Wegener's granulomatosis.

**Keywords:** Complete heart block, Wegener's granulomatosis, subglutic stenosis

*Manuscript received: January 10, 2011; Accepted: February 22, 2011*  
**Iran Cardiovasc Res J 2011;5(1):32-34**

## Introduction

Wegener's granulomatosis (WG) is a necrotizing vasculitis that mainly affects the respiratory tract and kidneys but it may also involve other organs. Cardiovascular manifestation of the disease may arise from involvement of coronary system, pericardium, myocardium, endocardium, valves, conduction system, or great vessels; but clinically notable cardiac involvement is only rarely encountered and symptomatic conduction defects in WG are quite uncommon. There are several reported cases of complete heart block (CHB) in WG in the literature,<sup>1</sup> of which only few have been in limited WG.<sup>2,3</sup> We hereby report a case of symptomatic atrioventricular block due to WG that was responsive to medication and reversing to normal sinus rhythm.

## Correspondence:

**M Zabihyeganeh**

Department of Rheumatology, Firouzgar General Hospital, Tehran University of Medical Sciences, Tehran, Iran

Tel: +98-21-22770699

E-mail: mozhdehzhabihi@gmail.com

## Case Report

A 62-year-old woman was admitted to coronary care unit of a University hospital in pre-syncope state. Her electrocardiography (ECG) showed CHB with narrow QRS and heart rate of 40/min which was resistant to atropine. Temporary pacing was applied. Her past medical history was negative for any cardiac problems and positive to surgeries for management of recurrent subglutic stenosis, a functional endoscopic sinus surgery, and also two recent dacryocystorhynchostomy (DCR); latest had only been performed ten days before new admission. Her recent medical file of the second DCR showed that both of her pre and postoperative ECGs were completely normal.

She complained from paresthesia in distal part of all four limbs, and also from shoulders pain. On physical examination, her blood pressure was 120/80 mm/Hg; oral temperature and pulse rate were 37.8°C and 40/min; respectively. Cardiopulmonary examination was normal, and her complete general physical examination was normal except for inflammation of nasal mucosa with multiple

crusts. Relevant laboratory tests were carried out and showed the following data:

Complete blood count, fasting blood glucose, blood urea nitrogen, serum creatinine, and urinalysis were normal. Cardiac enzymes (Troponin, CPK MB, LDH), serum ALT, AST, and angiotensin converting enzyme levels were normal. C-reactive protein (CRP) was elevated; erythrocyte sedimentation rate (ESR) and serum Ferritin were 65 mm/h, and 102 ng/ml; respectively. c-ANCA (Anti-neutrophil cytoplasmic antibody) was positive and serum anti PR 3 was reported as 200 (normal  $\leq 20$ ). Rheumatoid factor, ANA, p-ANCA, anti ds DNA, and anti Myeloperoxidase were all negative.

Computed tomography (CT) of chest showed a small speculated pulmonary nodule in left lung without calcification. Mucosal thickening was found in ethmoid and maxillary sinuses by CT scan. Echocardiography revealed ejection fraction as 60%, normal interventricular septum, and no wall motion abnormality or pericardial effusion. Electromyography and nerve conduction velocity showed mixed motor and sensory peripheral polyneuropathy. Finally, the presence of granuloma was detected in biopsy samples from nasal septum and maxillary sinus.

According to her history of recurrent subglottic stenosis and dacryocystitis, chronic sinusitis, bloody nasal discharge, limbs paresthesia, and musculoskeletal problems, and considering her pulmonary nodule, the presence of granuloma in mucosa, high serum anti PR 3 level and positive cANCA, and normal cardiac enzymes and echocardiography, she was diagnosed as a case of WG. Echocardiography helped ruling out differentials such as myocarditis and other cardiac infiltrative disease. She was then started on oral Prednisolone 60 mg/d and oral Cyclophosphamide 125 mg/d. She was awaited pacemaker insertion while the diagnostic tests and procedures were taking place. Surprisingly four days after initiation of immunosuppressive therapy, her ECG returned to normal sinus rhythm and she was discharged safely. Her ECG has remained normal in the last 2 years, as well as her renal system, during our follow-up visits.

## Discussion

In this case report we described a patient with complete heart block due to undiagnosed limited WG which rapidly responded to the treatment of the WG.

Cardiac involvement in WG, including its subclinical forms, is being increasingly recognized and

ranging between 6 to 44% of all WG cases, and is not as rare as previously considered.<sup>1</sup> This wide variation may be due to the nature of the disease itself, the medical subspecialty of the investigating team, the effect of medications on the natural course of disease, or to the number of procedures used to diagnose subclinical cases.<sup>2</sup> Of all cases of heart involvement in WG, 50% represent with coronary arteritis, 25% with myocarditis, 21% with valvulitis, 17% with conduction system granulomata, 13% with arteritis of the blood supply to the atrioventricular node, 11% with myocardial infarction, and 8% with epicarditis.<sup>4</sup> The probable pathogenesis of atrioventricular block associated with WG is granulomatous lesions within the conduction system, or arteritis of atrioventricular nodal artery. Symptomatic cardiac conduction system abnormalities in WG are rare. Supraventricular arrhythmias are much more common than ventricular arrhythmias. Complete heart block (CHB) in WG has been only reported in 6 cases (In English literature).<sup>2-7</sup>

Limited WG is a subtype of WG that involves upper respiratory tract or lung without glomerulonephritis; also there have been reported cases of limited WG with salivary, ear, orbital and cardiac involvement.<sup>8</sup> The paucity of reported cases of CHB in limited WG is worthy of attention. To our knowledge, beside present report, there have only been only two more cases published in English literatures.<sup>2,3</sup> In the first case presented by Ghaussy et al., CHB was the first manifestation of the disease in a previously undiagnosed limited WG in a male. This was similar to our patient not only in its presentation but also in its rapid response to treatment with Prednisolone and Cyclophosphamide; indeed Ghaussy was the first to use a gallium scan fused with myocardial perfusion scan which was compared with cardiac MRI for the diagnosis of CHB in a case of WG and showed that the initially increased uptake of Gallium near AV node became normal four months after therapy.<sup>3</sup> The second report, published by Lisitsin et al., described CHB in a 43-year-old woman, who was a previously known case of limited WG with inadequate response to conventional steroid and cyclophosphamide therapy, that finally responded to treatment with Infliximab.<sup>2</sup>

Because of our unawareness about this rare complication of WG before this case, the patient had been improperly elected for early permanent pacemaker insertion. We were unfortunately unable to define the possible pathologic reason of CHB in our case. However, considering the rapid resolution

of the block with immunosuppressive therapy we hypothesize that arteritis of the blood supply to the atrioventricular node has been a main mechanism of CHB.

Considering a published report of nine WG cases without any symptomatic cardiac involvement who, by using routine echocardiography, later proved to have some cardiac abnormalities.<sup>9</sup> It is suggested that more widespread use of Holter monitoring and echocardiography would lead to detecting an increasing number of asymptomatic, but important, cardiac complications in WG patients.

The present case showed that in regard to the

unusual presentation of limited WG, such as CHB, it is essential to make a correct decision on the treatment of CHB which maybe reversible by immunosuppressive therapy without any need to pacemaker insertion.

#### Acknowledgement

We would like to thank B Farahani, MD, assistant professor of cardiology, Tehran University of Medical Sciences, for his kindly consultations. The authors declare that they have no conflicts of interest.

#### References

- 1 Korantzopoulou P, Papaioannides D, Siogasa K. The Heart in Wegener's Granulomatosis. *Cardiology* 2004; **102**:7-10. [14988611]
- 2 Lisitsin S, Farah R, Shay M. Limited Wegener's granulomatosis, is it Limited? *Clin Rheumatol* 2007; **26**:1999-2000. [17401514]
- 3 Ghaussy NO, Du Clos TW, Ashley PA. Limited Wegener granulomatosis presenting with complete heart block. *Scandinavian J Rheumatol* 2004; **33**:115-8. [15163113]
- 4 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; **10**:148-54. [7292019]
- 5 Handa R, Wali JP, Aggarwal P, Wig N, Biswas A, Kumar AK. Wegener's granulomatosis with complete heart block. *Clin Exp Rheumatol* 1997; **15**:97-9. [9093782]
- 6 Wilcke JT, Nielsen PK, Jacobsen TN. Reversible complete heart block due to Wegner granulomatosis. *Int J Cardiol* 2003; **89**:297-8. [12767556]
- 7 Suleymenlar G, Sarikaya M, Sari R, Tuncer M, Sevinc A. Complete heart block in a patient with Wegner granulomatosis in remission- a case report. *Angiology* 2002; **53**:337-40. [12025922]
- 8 Stone JH, Wegener's Granulomatosis Etanercept Trial Research Group. Limited versus severe Wegener's granulomatosis: baseline data on patients in the Wegener's granulomatosis, etanercept trial. *Arthritis Rheum* 2003; **48**: 2299-309. [12905485]
- 9 Morelli S, Gurgo Di Castelmanardo AM, Conti F, Sgreccia A, Alessandri C, Bernardo ML, et al. Cardiac involvement in patients with Wegener's granulomatosis. *Rheumatol Int* 2000; **19**:209-12. [11063289]