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Case Report

A Rare Presentation of Wegners Granulomatosis as Complete Heart Block

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ABSTRACT

Cardiac involvement is seen in 6 to 25% Wegener's granulomatosis, but very rarely it can involve the conduction system and manifest as complete heart block. Earlier most of the case reports were with systemic variant rather than a limited variant. We report a case of complete Heart Block (3rd degree) in a case of limited Wegner granulomatosis which was treated with Permanent Pacemaker Implantation. Complete AV block is a rare but treatable manifestation of cardiac involvement of wegners granulomatosis usually associated with early active systemic disease. All patients diagnosed with Wegener's granulomatosis should be screened with a baseline electrocardiogram and a transthoracic echocardiogram to document cardiac involvement and alert clinicians to those at risk of further cardiac complications.

Key words: Wegners granulomatosis, ECG, Complete heart block, Permanent pacemaker implantation, Nasal biopsy.

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INTRODUCTION

Wegener's granulomatosis is a systemic necrotizing granulomatous vasculitis of small- to medium-sized vessels associated with diffuse Cytoplasmic Anti-neutrophil Cytoplasmic Antibodies (cANCA) specific for proteinase-3 (PR3). It typically affects the upper and lower airways, lungs and kidneys. Cardiac involvement manifestation is seen in 6 to 25 % patients and is more common with severe renal involvement. These manifestations include pericarditis, myocarditis and aortitis, although myocardial infarction is also recognized.[1] Cardiac conducting tissue involvement is extremely rare. Although often not clinically apparent, cardiac involvement is associated with increased mortality.[2] We report a case of complete Heart Block (3rd degree) in a case of limited Wegner granulomatosis which was treated with Permanent Pacemaker Implantation.

CASE REPORT

A 44 year old male of south Asian ethnicity admitted to our hospital, in view of multiple episode of epistaxis and diminished vision. His examination revealed bilateral scleritis (Figure 1) with left sided squint and exudative retinal detachment along with tenderness of maxillary sinus. Nasal biopsy (Figure 3) revealed necrotizing and destructive leucocytic angitis of arteries and veins along with ill-formed granulomas reflecting the diagnosis of Wegner granulomatosis. There was no evidence of proteinuria and erythrocyte cast in the urine examination. Also his creatinine and potassium were within normal limit suggesting no renal involvement establishing it as limited variant of Wegners granulomatosis. His blood investigation revealed cANCA 152 (n<12) ESR of 57 mm at the end of one hour, c reactive protein of 9. NCCT of Para nasal sinuses showed bilateral maxillary sinusitis with left CSOM. CECT thorax was normal. On this basis, a diagnosis of limited Wegner granulomatosis was made and the Birmingham Vasculitis activity score (BVAS)[3] which is a method for assessment of activity of vasculitis was calculated to be 12. In view of increasing ophthalmological and other symptoms patient was started on cyclophosphamide, azathioprine and methylprednisolone. During the fifth cycle pt developed ophthalmological flare and syncope with significant bradycardia. ECG showed (Figure 2) heart rate of 34/minute with AV dissociation and ventricular escape rhythm. It was promptly recognised and treated temporary pacemaker. At the initial presentation his ECG revealed presence of LAHB with RBBB which progressed to CHB at the time of disease flare. Later on patient was started on rituximab but CHB still didn't recovered. His trans thoracic echo was within normal limit with LVEF=60% patient was tested for TS t3 t4 and RFT, serum electrolytes, cardiac enzymes which were all normal ruling out causes of reversal Complete heart block. Later patient was considered for permanent pacemaker replacement.

DISCUSSION

Cardiac involvement of Wegener's granulomatosis was first reported by Wegener in 1936, but the first case of associated complete AV block did not appear until 1969.[3] Since then, there have only been 13 cases reported in the published literature and all but one case was associated with systemic disease.[1,3-14] Our case had biopsy proven Wegners granulomatosis limited variant with complete heart block and required permanent pacemaker implantation. Heart block in limited Wegners granulomatosis was earlier reported with one case by Ghaussy *et al.*[11] were they noticed spontaneous resolution. In total Complete AV block was present at disease onset in 50% and in 70% by one year. Half of the cases were asymptomatic, although all had clinical or laboratory evidence of active disease. Echocardiograms were performed in nine of the patients and abnormalities were present in six; most commonly valvular abnormalities and pericardial effusion. Treatment included corticosteroids, cyclophosphamide, temporary pacing wire and pacemaker implantation. The

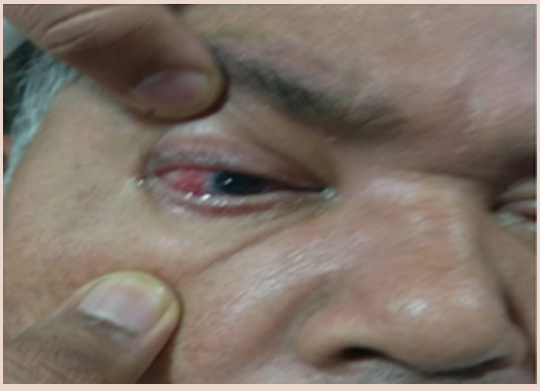

Figure 1: Scleritis.

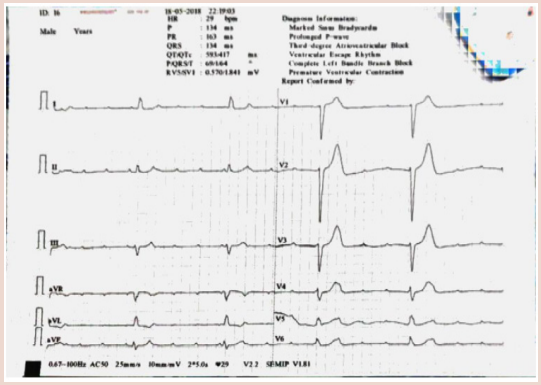

Figure 2: ECG of the patient showing complete heart block with ventricular escape rthm of 34/minute.

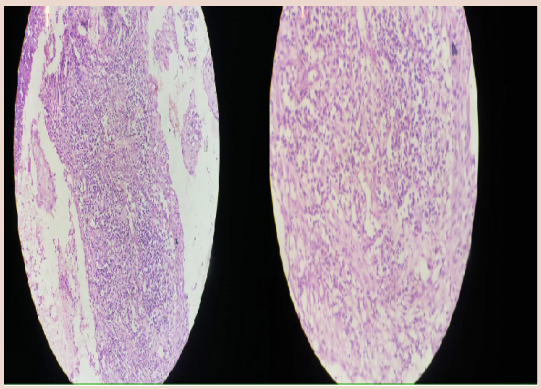

Figure 3: H/E stained sections suggestive of ulcerative squamous epitheilium with foci of pseudostaritified columnar epthelium. The sub epithelial tissue shows necrotizing and destructive leucocytic angitis of arteries and veins along with ill formed granulomas comprising of hisitiocytes, multinucleate gaint cells, plasma cells and eosinophills. Areas of fibrinoid necrosis also noted. Overall findings consistent with wegnres garnulomatosis.

complete AV block resolved in 9 of the 13 cases. Two patients died of cardiac dysrhythmia. Post-mortem examination of the first case reported by Longauer *et al.* showed granulomatous involvement of the conduction system with fibrosis and hyalinization, with no necrosis.[3] In the second case, Ohkawa *et al.* reported small foci of inflammation affecting the sinus and AV nodes with AV nodal arteritis, inflammatory necrotising granulomatous involvement of the conducting system and myocardium associated with an exudative endocarditis and fibrinous pericarditis.[4]

In summary, complete AV block is a rare but treatable manifestation of cardiac involvement usually associated with early active systemic disease. All patients diagnosed with Wegener's granulomatosis should be screened with a baseline electrocardiogram and a transthoracic echocardiogram to document cardiac involvement and alert clinicians to those at risk of further cardiac complications.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

ABBREIVATIONS

ECG: Electrocardiogram.

SUMMARY

Complete AV block is a rare but treatable manifestation of cardiac involvement usually associated with early active systemic disease. All patients diagnosed with Wegener's granulomatosis should be screened with a baseline electrocardiogram and a transthoracic echocardiogram to document cardiac involvement and alert clinicians to those at risk of further cardiac complications.

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