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 Wegener 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.

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. Cardiac conducting tissue involvement is extremely rare. Although often not clinically apparent, cardiac involvement is associated with increased mortality. 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 Wegens granulomatosis. His blood investigation revealed cANCA 152 (<12) ESR of 57 mm within normal limit with LVEF=60% patient was tested for TS 13 14 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. Since then, there have only been 13 cases reported in the published literature and all but one case was associated with systemic disease. Our case had biopsy proven Wegens 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 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 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 13 14 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.
Figure 2: ECG of the patient showing complete heart block with ventricular escape rhythm of 34/minute.

Figure 3: H/E stained sections suggestive of ulcerative squamous epithelium with foci of pseudostatified columnar epithelium. The sub epithelial tissue shows necrotizing and destructive leucocytic angitis of arteries and veins along with ill-formed granulomas comprising of histiocytes, multinucleate giant cells, plasma cells and eosinophils. Areas of fibrinoid necrosis also noted. Overall findings consistent with Wegener's granulomatosis.

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CONFLICT OF INTEREST
The authors declare no conflict of interest.

ABBREVIATIONS
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.

REFERENCES