Case Report

A patient with Wegener’s granulomatosis in apparent remission presenting with complete atrioventricular block

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Received 7 November 2010; received in revised form 16 January 2011; accepted 24 January 2011

KEYWORDS
Wegener’s granulomatosis; Complete atrioventricular block; Cardiac involvement; Multisystem disorder; Pacemaker

Introduction

Wegener’s granulomatosis is a systemic necrotizing granulomatous vasculitis of small- to medium-sized vessels typically affecting upper and lower airways, lungs, and kidneys. Cardiac involvement is less common and conducting tissue involvement is extremely rare. Cardiac manifestations are often not clinically apparent, but are associated with increased mortality. We report the case of a 36-year-old female with Wegener’s thought to be in remission, presenting in complete atrioventricular (AV) block, with echocardiographic evidence of basal interatrial septum and basal lateral left atrial wall thickening. Despite immunosuppression therapy a permanent pacemaker was required for recurring complete AV block. Although rare, this case emphasizes the need for careful and regular screening for cardiac involvement in this multi-system condition.

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Case report

A 36-year-old woman with a three-month history of presyncopal symptoms presented with two syncopal episodes...
in May 2010. At the initial presentation her electrocardiogram showed sinus rhythm with first degree atrioventricular (AV) block and a PR interval of 250 ms. Overnight monitoring did not reveal any other abnormality and she was discharged with plans for further outpatient cardiac investigation. Six days later she re-presented with a second episode of syncope and at this time electrocardiogram showed complete AV dissociation with a ventricular rate of 28 beats per minute (Fig. 1). No other symptoms or signs of systemic illness were elicited.

In June 2008 she had been diagnosed with Wegener’s granulomatosis after presenting with a two-week history of nasal crusting, loss of hearing, polyarthralgia, and small-vessel ischemia of the fingers and toes. At that time erythrocyte sedimentation rate (ESR) was 86 mm/h, C-reactive protein (CRP) 140 mg/l, and cANCA immunofluorescence was positive with anti-PR3 titre of 103 units (normal range 0—7 units) by enzyme-linked immunosorbent assay. An electrocardiogram had been performed showing sinus rhythm with a normal PR interval and QRS duration, but regrettably no other cardiac investigation, including echocardiography, had been undertaken at the time of first diagnosis. She was treated with high-dose steroids, cyclophosphamide, prostacyclin, and co-trimoxazole. Clinical and laboratory remission was achieved by September 2008, and she was maintained on azathioprine therapy.

At the time of this admission, her inflammatory markers were mildly elevated (ESR of 19 mm/h and CRP of 23 mg/l) and the anti-PR3 titre was normal (4.5 units). Serum potassium was increased at 7 mmol/l with a creatinine of 118 μmol/l, without erythrocyturia, urinary casts, or proteinuria. Although she initially settled into a sinus rhythm with first-degree AV block following correction of her hyperkalaemia, complete AV block recurred the next day despite normal electrolyte levels.

A transthoracic echocardiogram showed markedly thickened basal segments of her left atrium, including the basal interatrial septum (Figs. 2 and 3). Left ventricular chamber size and wall thickness were normal, with normal systolic and diastolic function. It was felt, given the raised inflammatory markers, that she may have active granulomatous disease affecting the region of the atrioventricular node and she was treated with intravenous methylprednisolone and cyclophosphamide. Despite this, she continued to experience symptomatic high-grade AV block with prolonged episodes of ventricular standstill and so a dual chamber pacemaker was implanted.

At three-month follow-up she remained asymptomatic. However, her transthoracic echocardiographic appearances were essentially unchanged and she had been pacing dependent for 85% of the time in the month up to review.

**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 previously reported in the published literature and all but one case was associated with systemic disease (Table 1) [1,3—14]. Complete AV block was present at disease onset in 50% and in 70% by one year. Half...
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levels [15,16]. Given this uncertainty, we opted to treat 
of patients with limited active disease may have negative 
ity of cANCA without adverse outcome, whilst up to 20%
to local reference values. The significance of this is unclear 
our patient, this would be considered as normal according 
to the previous publication cases, our patient was 
unusual in that she had relatively longer disease duration 
of 24 months at diagnosis, she had previously been treated 
with intravenous cyclophosphamide, and she had no clinical 
or laboratory evidence of extra-cardiac increased disease 
activity. Although a low level cANCA level was detected in 
her with further immunosuppression on the basis of raised 
inflammatory markers and cardiac imaging suspicious for 
complete AV block, however, she is young and otherwise 
well and so we would feel that an association between these 
abnormalities and her presentation is likely. We had hoped 
to arrange magnetic resonance imaging to try and clarify if 
disease was also present at the AV node and His conducting 
tissue, but her need for pacing support prevented this.

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. Echocardiography 
frequently detects abnormalities suggesting cardiac 
involvement in asymptomatic individuals and this case highlights 
that later presentation of cardiac abnormalities than 
previously reported can occur and lifelong surveillance 
needs to be considered given the poorer prognosis in these 
individuals. In turn, all patients presenting with cardiac 
abnormalities and evidence of systemic inflammation should 
be screened for Wegener’s by clinical, laboratory, and radiographic 
assessment.

References

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complications of Wegener granulomatosis: a case report of complete 
[3] Longauer F, Takac M, Halasova K. On damage of the heart conduc-
Ohtsubo K. Extensive involvement of the myocardium and the 
cardiac conduction system in a case of Wegener’s granulomato-

Table 1  Summary of reported cases of complete heart block.

<table>
<thead>
<tr>
<th>Case</th>
<th>Authors</th>
<th>Year</th>
<th>Age/sex</th>
<th>Type</th>
<th>Duration (months)</th>
<th>Previous treatment</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Longauer [3]</td>
<td>1969</td>
<td>47, F</td>
<td>syst</td>
<td>0</td>
<td>–</td>
<td>cs, ppm</td>
<td>died</td>
</tr>
<tr>
<td>2</td>
<td>Forstot [1]</td>
<td>1980</td>
<td>26, M</td>
<td>syst</td>
<td>11</td>
<td>cs, cyc, tpw, ppm</td>
<td>resolution</td>
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<tr>
<td>9</td>
<td>Wilcke [10]</td>
<td>2003</td>
<td>63, M</td>
<td>syst</td>
<td>0.3</td>
<td>–</td>
<td>cs, cyc, pph, tpw</td>
<td>resolution</td>
</tr>
<tr>
<td>11</td>
<td>Elikowski [12]</td>
<td>2006</td>
<td>52, M</td>
<td>syst</td>
<td>14</td>
<td>cs, cyc, cic, cs, cyc, tpw, ppm</td>
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<td></td>
</tr>
<tr>
<td>13</td>
<td>Sarlon [14]</td>
<td>2010</td>
<td>77, M</td>
<td>syst</td>
<td>0</td>
<td>–</td>
<td>cs, cyc, ppm</td>
<td>died</td>
</tr>
</tbody>
</table>

Abbreviations used: F, female; M, male; lim, limited; syst, systemic; cs, corticosteroids; cyc, cyclophosphamide; cic, ciclosporin; aza, azathioprine; ppm, permanent pacemaker; tpw, temporary pacing wire.


