Case Report

A Case of Atrioventricular Complete Block Due to Behcet's Disease

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SUMMARY

The cardiovascular system is involved in 5% of the cases of Behcet's disease. Thrombophlebitis, aneurysm in arteries, pericarditis, myocarditis, valvular disease, ventricular arrhythmia, and conduction system disorders may occur. A case of Behcet's disease with complete proximal atrioventricular block is presented. (Jpn Heart J 2002; 43: 697-701)

Key words: Behcet's disease, Atrioventricular block

BEHCET'S DISEASE is a diffuse vasculitis progressing with recurrent oral aphtha and genital ulceration, uveitis, and iritis. Cardiac involvement is rare; but pericarditis, myocarditis, valvular disease, arrhythmia, and conduction impairment may occur.¹⁻³⁾ Although cases of atrioventricular complete block at the level of the His bundle in Behcet's disease have been reported,⁴⁾ to our knowledge, no cases of Behcet's disease with complete block, the localization of which is the atrioventricular node (proximal type), have been reported so far. Here we present a case of Behcet's disease with complete proximal atrioventricular block.

CASE REPORT

A 28-year-old Turkish woman (married with 2 children) presented to our dermatology outpatient clinic with complaints of recurrent oral aphtha and genital ulceration which had started 2 years previously, and swelling in the knee joint. She was subsequently hospitalized. Atrioventricular block did not exist in the ECG examination performed that day (Figure 1). In the clinical laboratory tests, hemoglobin was found to be 12.6 g/dL, hematocrit 36%, WBC 9000/mm³, sedimentation rate 34 mm/hour, ASO: 180 IU/mL (normal <200 IU/mL), CRP: 48 mg/L (normal <6 mg/L), rheumatoid factor: 7 (normal <20 IU/mL), ANA: (-), Anti-ds DNA: (-), and patergy test (+). There was no family history of collagen vascular disease. Considering the existing clinical findings and laboratory tests, a

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Figure 1. Twelve-lead ECG of this patient at her first visit was normal.

diagnosis of Behcet's disease was made and therapy was initiated with colchicine and antiinflammatory drugs. The patient, who had been diagnosed as having Behcet's disease and followed up for the previous 2 years, visited the cardiology outpatient clinic with complaints of dizziness, presyncope, and sluggishness. In a physical examination, her pulse rate (45/min) and other systemic findings were within normal ranges (WBC 9100/mm³, haemoglobin 14.1 g/dL, hematocrit 39.5%, thrombocyte count 307,000/mm³, sedimentation rate 18 mm/hour). The telecardiography and color Doppler echocardiography findings were all normal, but her ECG and Holter recording revealed atrioventricular complete block (Figure 2). Ambulatory ECG with 12 leads (Rozzin Electronics, Inc.) and thallium-201 scintigraphy, performed in order to determine the silent myocardial ischemia, was normal. Possible factors in block formation, other than Behcet's disease, were examined through clinical and laboratory studies (angiocardiography, etc.) but no disease was detected. Complete atrioventricular block within the AV node was detected in the electrophysiological study performed to localize the block (Figure 3). When atropine was administered the location of the block remained the same. However, both the atrial rate and the escape rhythm rate increased (Figure 4). Considering the symptoms, a permanent DDD-R pacemaker was implanted.

Figure 2. Twelve-lead ECG of this patient with dizziness showed complete AV block with escape rhythm of narrow QRS complex.

Figure 3. Intracardiac recordings revealed that the site of complete AV block was within the atrioventricular node. HIS md, px, ds=His-bundle with bipolar recordings from the mid, proximal, and distal, poles; A=Atrial potential; H=His potential; V=Ventricular potential.

Figure 4. After atropine, the location of the block remained the same, but increased both the atrial and escape rhythm rates. hRA=high right atrial electrogram; HIS md, px, ds=his-bundle with bipolar recordings from the mid, proximal, and distal, poles; A=atrial potential; H=his potential; V=ventricular potential.

DISCUSSION

Behcet's disease is common in eastern Mediterranean countries and Japan. It is a vasculitis which often involves both large and small arteries.¹⁻³⁾ Neutrophil activation caused by immune complex build-up in small arteries gives rise to necrosis on the walls of large arteries, blocking vasa vasorum. Cardiac involvement arises as pericarditis, myocardial infarction, coronary arteritis, valvular regurgitation (particularly aortic valve regurgitation), and impaired conduction.^{2,5,6)} Although the etiology of the conduction defect in Behcet's disease is not exactly known, it has been suggested that inflammation affects the conduction system and causes the impairment of conduction.^{3,7)} Sun, *et al* reported one case of Behcet's disease in which they detected intra-hisian block, and aortic regurgitation.⁴⁾ Nojiri, *et al* reported one case with aortic regurgitation, valsalva aneurysm rupture, first degree atrioventricular block, and incomplete right bundle branch block.⁷⁾ In one case reported by Or, *et al* obstruction of the vena cava superior together with atrioventricular complete block were observed.⁸⁾ Stucchi, *et al* reported one case having atrioventricular complete block with atrial fibrillation,⁹⁾ and Kansu, *et al* a case with atrioventricular complete block and renal artery involvement.¹⁰⁾ Coronary artery disease and silent myocardial ischemia¹¹⁾ due to

microvascular disease can induce conduction block. However, our case did not have ischemia. To our knowledge, no cases with proximal atrioventricular complete block have been reported thus far. In our case, the location of the block, the atrioventricular node, necessitates that congenital block be considered in the differential diagnosis since patients with congenital complete block can also continue to live for a long time asymptotically without any other cardiac pathology and generally with an escape rhythm rate of 40-60/min. Such a congenital block and Behcet's disease may coexist. However, in our case, first the normal cardiovascular system and ECG examination findings 2 years previously, second, the absence of a family history of collagen vascular disease, and finally, the lack of involvement of any other factor which may potentially cause atrioventricular complete block in the 2-year period, indicate that the condition may have developed predominantly as a result of Behcet's disease.

In conclusion, proximal atrioventricular complete block may develop without other signs of cardiac involvement in Behcet's disease, which, we think, may have resulted from inflammation of the conduction system.

REFERENCES

1. Wechsler B, Du LT, Kieffer E. Cardiovascular manifestations of Behcet's disease. *Ann Med Interne (Paris)* 1999; 150: 542-54.
2. Koç Y, Güllü I, Akpek G, Akpolat T, Kansu E, Kiraz S. Vascular involvement in Behcet's disease. *J Rheumatol* 1992; 19: 402-3.
3. James DG. Behcet's syndrome. *N Engl J Med* 1979; 301: 431-2.
4. Sun RL, Wang FZ, Hu SJ. Intra-Hisian block associated with unusual etiologies. *PACE* 1987; 10: 1117-24.
5. Bowles CA, Nelson AM, Hammill SC, O'Duffy JD. Cardiac involvement in Behcet's disease. *Arthritis Rheum* 1985; 28: 345-8.
6. James DG, Thomson A. Recognition of the diverse cardiovascular manifestation in Behcet's disease. *Am Heart J* 1982; 103: 457-8.
7. Nojiri C, Endo M, Koyanagi H. Conduction disturbance in Behcet's disease Association with ruptured aneurysm of the sinus of Valsalva into the left ventricular cavity. *Chest* 1984; 86: 636-8.
8. Or I, Roguin N, Yahalom M, Birjiniuk V, Amikam S. Pacemaker implantation in a patient with Behcet's disease associated with superior vena cava obstruction. *Cardiovasc Intervent Radiol* 1986; 9: 13-4.
9. Stucchi C, Vollenweider A. Maladie de Behcet et manifestations atypiques. *Ophthalmologica* 1958; 135: 573.
10. Kansu E, Deglin S, Cantor RI, Burke JF Jr, Cho SY, Cathart RT. The expanding spectrum of Behcet's syndrome: a case with renal involvement. *JAMA* 1977; 237: 1855-6.
11. Güllü IH, Benekli M, Müderrisoğlu H, *et al.* Silent myocardial ischemia in Behcet's disease. *J Rheumatol* 1996; 23: 323-7.