CASE REPORT

Complete Heart Block with Ventricular Tachycardia in a Patient with Hemochromatosis

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ABSTRACT

Hereditary hemochromatosis is an inherited condition of dysregulated iron absorption, and usually presents with clinical features of hepatic dysfunction. Cardiac involvement as the presenting manifestation of hereditary hemochromatosis is rare. We report a young male who presented with complete heart block and ventricular tachycardia and was subsequently diagnosed as hemochromatosis. He was managed with permanent pacemaker implantation, oral antiarrhythmic and chelation therapy.

Key words: Complete heart block, hemochromatosis, ventricular tachycardia

INTRODUCTION

Incidence of cardiac arrhythmias is only marginally increased in asymptomatic subjects with hereditary hemochromatosis.[1] Complete atrioventricular block and ventricular tachycardia as presenting manifestations are very rare. We report a young male with complete AV block and non-sustained ventricular tachycardia who was found to have hemochromatosis.

CASE REPORT

A 30-year-old male presented with a 2-day history of recurrent syncope. He noticed diffuse pigmentation of the skin since the age of 20 years. Physical examination revealed smooth, diffusely pigmented skin with absence of chest, facial, axillary and pubic hair. There was no hepatosplenomegaly and his testes and penis were small. His heart rate was 40 beats/min with a blood pressure of 120/70 mmHg. Examination of the cardiovascular system showed intermittent cannon waves, cardiomegaly, variable intensity of first heart sound and normal second heart sound.

Laboratory investigations showed a hematocrit value of 50%, hemoglobin of 11 gm/dL, platelet count of 200,000/mm³, absolute neutrophil count of 7600/mm³, serum creatinine of 0.6 mg/dL, sodium of 140 meq/L, potassium of 3.4 meq/L, calcium of 9.4 mg/dL and magnesium of 2.1 meq/L. Liver function test and thyroid function tests were normal. His blood sugar was elevated. Serum iron was elevated to 312 μg/dL (N = 50-150 μg/dL) with a total iron binding capacity of 268 μg/dL (N = 250-400 μg/dL), transferrin saturation of 70.5% (N = 15-50%) and serum ferritin of 6727 ng/mL (N = 10-150 ng/mL). His electrocardiogram at admission showed complete heart block with a wide QRS escape rhythm at a rate of 36 beats/min. During hospital stay, he developed several episodes of non-sustained monomorphic ventricular tachycardia [Figure 1]. There was cardiomegaly in the...
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overload with secondary tissue damage in a wide range of organs. Initial symptoms often include lethargy, arthralgia, change in skin color, loss of libido and features of diabetes mellitus. Hepatomegaly, splenomegaly, arthropathy, congestive heart failure and cardiac arrhythmias are common in advanced disease. Cardiac involvement is the presenting manifestation in about 15% of the symptomatic patients.[2] The most common manifestation is congestive heart failure, which occurs in about 10% of young adults with the disease, especially those with juvenile hemochromatosis. Symptoms of congestive heart failure may develop suddenly, with rapid progression to death if untreated. The heart is diffusely enlarged; this may be misdiagnosed as idiopathic cardiomyopathy if other overt manifestations are absent.

He was managed with permanent pacemaker implantation. Post-implantation, he again developed recurrent episodes of monomorphic non-sustained VT [Figure 2], for which he was started on oral amiodarone therapy. He improved with treatment and was discharged on oral iron chelation therapy and amiodarone.

**DISCUSSION**

Hemochromatosis is a common inherited disorder of iron metabolism, characterized by excessive iron deposition in parenchymal cells that can lead to total-body iron overload with secondary tissue damage in a wide range of organs. Initial symptoms often include lethargy, arthralgia, change in skin color, loss of libido and features of diabetes mellitus. Hepatomegaly, splenomegaly, arthropathy, congestive heart failure and cardiac arrhythmias are common in advanced disease. Cardiac involvement is the presenting manifestation in about 15% of the symptomatic patients.[2] The most common manifestation is congestive heart failure, which occurs in about 10% of young adults with the disease, especially those with juvenile hemochromatosis. Symptoms of congestive heart failure may develop suddenly, with rapid progression to death if untreated. The heart is diffusely enlarged; this may be misdiagnosed as idiopathic cardiomyopathy if other overt manifestations are absent.

**Figure 1:** Electrocardiogram showing complete AV block and non-sustained VT

**Figure 2:** Electrocardiogram showing ventricular paced beats with ventricular premature contractions in couplets
Cardiac arrhythmias include premature supraventricular beats, paroxysmal tachyarrhythmia, atrial flutter, atrial fibrillation and varying degrees of atrioventricular block.\cite{3-5} Various A-V blocks and atrial arrhythmias are reported in patients with hemochromatosis, but complete heart block with ventricular tachycardia is a rare occurrence.\cite{6,7} A study among the urban male population of Sweden showed a prevalence of iron overload in the pacemaker-treated population, where high-grade AV block was less than 2%.\cite{8} Animal studies have shown that chronic iron overload decreases CaV1.3-dependent L-type Ca$^{2+}$ currents, leading to bradycardia, altered electrical conduction and atrial fibrillation.\cite{9}

**CONCLUSION**

Our patient had the classical clinical and laboratory manifestations of hemochromatosis, and he completely responded to permanent pacemaker implantation followed by oral amiodarone therapy. Asymptomatic male with hemochromatosis presenting with complete heart block and ventricular tachycardia are the highlights in this case.

**REFERENCES**


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