Complete heart block in thalassemia major: a case report

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SUMMARY: Küçükosmanoğlu O, Özbarlas N, Şaşmaz İ. Complete heart block in thalassemia major: a case report. Turk J Pediatr 2002; 44: 261-262.

Cardiac complications of iron overload are the most common cause of death in patients with thalassemia major. These complications include recurrent pericarditis, refractory congestive heart failure and rhythm disorders. The usual rhythm disturbances are supraventricular or ventricular premature contractions and first-or second-degree heart block. Complete heart block is a very rare complication of thalassemia major. Herein, we report a case of complete heart block with thalassemia major. The patient also had serious congestive heart failure. Management of the heart block with pacemaker brought no clinical improvement, and she died in the second month of hospitalization.

Key words: thalassemia major, complete heart block, endocardial pacemaker.

The cardiac complications of iron overload are the most common causes of death in patients with thalassemia major[1]. These complications include recurrent pericarditis, refractory congestive heart failure, supraventricular or ventricular premature contractions and various forms of heart block Although many authors mentioned "various forms" of heart block, first- and second-degree block are the most common types. Complete heart block is a very rare condition in patients with thalassemia major[2-5].

In this paper, we report a 15-year-old girl with complete atrioventricular block and severe congestive heart failure due to cardiac hemosiderosis who died in spite of medical therapy and pacemaker implantation.

Case Report

A 15-year-old girl was referred to the Pediatric Cardiology Department with generalized edema, dyspnea, fatigue and generalized weakness. She had been followed at the Pediatric Hematology Department with the diagnosis of thalassemia major since her neonatal period, and she had received many blood transfusions and irregular deferoxamine therapy. Physical examination revealed a regular pulse of 60 bpm and blood pressure of 120/70 mmHg. She had generalized edema, ascites, marked hepatosplenomegaly and chromatic skin discoloration. Her hemoglobin level was 10.2 g/dl and ferritin level 4,520 ng/ml. Heart sounds were normal. Chest X-ray showed marked cardiomegaly with a cardiothoracic index of 0.62. Echocardiography showed severe systolic dysfunction (ejection fraction 38%, fractional shortening 18%) and dilatation of cardiac chambers. Twelve-lead ECG showed complete atrioventricular block. Holter recording revealed complete atrioventricular block with a ventricular rate of 50-60 bpm, frequent multiform ventricular premature complexes and slow, nonsustained (3-11 beats) idioventricular rhythm episodes (Figs. 1a and 1b). Decongestive therapy was

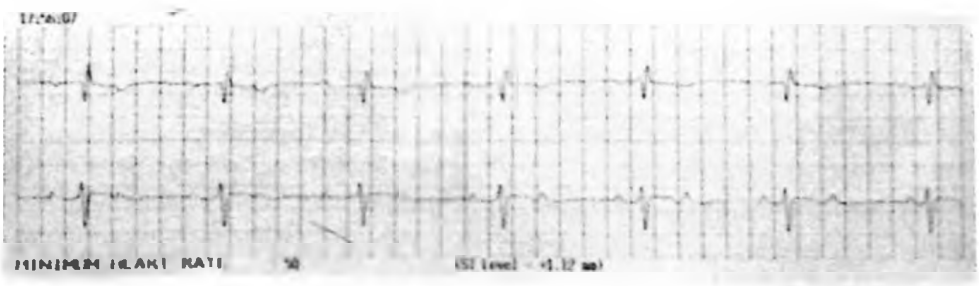

Fig. 1. Holter electrocardiogram obtained before pacemaker implantation, with a) complete heart block and,

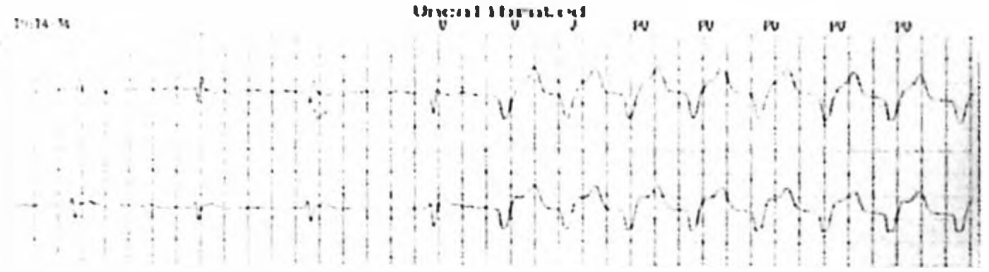

Fig. 1. Holter electrocardiogram obtained before pacemaker implantation, with b) idioventricular rhythm episode.

given and an endocardial DDD pacemaker was implanted successfully. She was discharged a week after pacemaker implantation with ejection fraction of 48% and fractional shotening of 24%. However, four days later she admitted to hospital with worsening symptoms of heart failure. The pacemaker functions were found normal. In spite of further intensive medical therapy, she died in the second month of hospitalization.

Discussion

Repeated blood transfusions to prevent severe anemia are necessary in patients with thalassemia major. But these frequent transfusions lead to chronic iron overload and related cardiac complications. Extent of cardiac impairment mostly depends on the quantity of iron deposition in the ventricular myocardium. When clinical findings of cardiac failure become apparent, patients with thalassemia major have a poor prognosis with a short life expectancy from six months to one year[6-7]. However, in a recent study, Kremastinos et al.[2] found that the five-year survival rate in patients with thalassemia and heart failure was greater (48%) than previously reported. Regular subcutaneous deferoxamine infusion therapy may protect patients from iron overload and related cardiac toxicity. Nevertheless, patients who receive deferoxamine are not entirely risk-free of cardiac iron deposition[5]. Serum ferritin levels are not correlated with survival in patients with thalassemia major and cardiac involvement[2]. Besides ventricular and atrial myocardium, conduction tissue is another site of iron deposition. The usual cardiac rhythm disturbances are supraventricular or ventricular premature contractions, and first-or second-degree heart block. Engle et al.[3] reported two cases of complete heart block due to hemochromatosis in the pre-deferoxamine era. Kremastinos et al.[2] reported that one of 52 patients who received iron chelation therapy with deferoxamine before the age of five years had complete heart block, and they emphasized that patients with rhythm disorders had poor prognosis. In our patient, heart block was an additional cause of congestive cardiac failure, but the management of the heart block with pacemaker brought no clinical improvement and she died in the second month of hospitalization. Insufficient iron chelation therapy was the main reason for the severe cardiac involvement. High-dose continuous deferoxamine infusion could be effective in decreasing the serum ferritin level, but this does not provide clinical improvement in patients with established clinical findings of heart failure[1,2].

We conclude that complete atrioventricular block may develop in the terminal stage of thalassemia major and that pacemaker therapy may not be effective in this condition because of the severe involvement of the myocardium. Regular cardiovascular evaluation of the patients and timely diagnosis of cardiac involvement may be beneficial in the management of thalassemia major.

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