Persistent atrial standstill and idioventricular rhythm in a patient with thalassemia intermedia

Talasemi intermedialı bir hastada kalıcı atriyal felç ve idiyoventriküler ritim

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We present a 57-year old male patient with thalassemia intermedia and right heart failure. He had a 30-year history of anemia and short-term iron therapy without blood transfusion. Hemoglobin level was 7.1 g/dl and hematocrit was 22.7%. White blood-cell and platelet counts, and serum ferritin level were normal. Electrocardiography showed irregular narrow QRS bradyarrhythmia, suggesting slow atrial fibrillation at a mean rate of 35 beats/min. Echocardiographic examination revealed dilatation of the right atrium and ventricle, depressed systolic right ventricular function, advanced tricuspid regurgitation, and mild pericardial effusion. In the electrophysiologic study, no electrical activity was recorded in the right atrium. It was inexcitable at multiple sites and no retrograde conduction to the right atrium could be elicited by ventricular pacing. His bundle (HB) recording showed fixed retrograde HB activation with ventricular rhythm originating from different foci. Retrograde V-H conduction time during ventricular rhythm was 95 msec and did not change. There was no retrograde nodal conduction. A VVIR pacemaker was implanted. During a six-month follow-up, he felt well, his functional capacity was NYHA class II, and his basic rhythm was widened QRS arrhythmia with a rate of 20 beats/min. To the best of our knowledge, atrial electrical inactivity together with right-heart failure and pericarditis confined to the right heart chambers has hitherto not been reported in thalassemic disorders.

Key words: Atrial function, right; beta-thalassemia/complications; echocardiography; electrocardiography; heart failure/etiology; paralysis; pericarditis/etiology.

Bu yazıda, 30 yıl önce talasemi intermedia tanısı konan ve sağ kalp yetersizliği gelişen 57 yaşında bir erkek hasta sunuldu. Hastanın kısa süre demir tedavisi gördüğü, ancak hiç kan transfüzyonu yapılmadığı öğrenildi. Hemoglobin düzeyi 7.1 gr/dl, hematokrit 22.7% idi. Beyaz küre ve platelet sayısı ve serum demir düzeyi normaldi. Elektrokardiyogramda, yavaş hızlı atriyal fibrilasyon olarak düşünülen, 35/dk hızda dar QRS kompleksli ritim izlendi. Ekokardiyografide, sağ atriyum ve ventrikül genişlemesi, sağ ventrikül sistolik fonksiyonunda azalma, ileri derece triküspit yetersizliği ve hafif perikardiyal efüzyon saptandı. Elektrofizyolojik çalışmada sağ atriyumda elektriksel aktivite kaydedilemedi. Sağ atriyumun çeşitli bölgelerine yapılan uyarılara yanıt alınamadı ve sağ ventriküler pacing ile sağ atriyuma retrograt iletim gözlenmedi. His bölgesi kayıtlarında, farklı odaklardan köken alan ventriküler aktivitenin retrograt olarak His bölgesini aktive ettiği görüldü. Ventriküler ritim sırasında retrograt V-H iletim zamanı 95 msn ile değişiklik göstermedi. Retrograt nodal iletim yoktu. Hastaya VVIR kalp pili takıldı. Altı aylık takip sırasında hasta kendini iyi hissettiğini belirtti; fonksiyonel durumu NYHA sınıf II idi. Temel ritmi, 20/dk ile genişlemiş QRS aritmisi şeklindeydi. Bildiğimiz kadarıyla, sağ kalple sınırlı perikardiyal efüzyon ve sağ kalp yetersizliğinin eşlik ettiği atriyal elektriksel inaktivite, talasemik bozukluklarda ilk kez bildirilmektedir.

Anahtar sözcükler: Atriyal fonksiyon, sağ; beta talasemi/komplikasyon; ekokardiyografi; elektrokardiyografi; kalp yetersizliği/etyoloji; paralizi; perikardit/etyoloji.

Beta-thalassemias represent a group of genetically inherited hemoglobin disorders leading to chronic anemia.[1] Two clinical forms have been defined: thalassemia major (TM) and thalassemia intermedia (TI). The former presents as severe anemia from the first year of life requiring life-long transfusion therapy, whereas TI presents as mild anemia of late clinical onset, which may not necessitate regular transfusions.

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The pathogenesis of cardiac involvement is not certain but is probably related to iron overload secondary to overtransfusions especially in TM.[2] Left-sided cardiac failure, arrhythmias, and pericarditis may be seen.[3,4] In TI, age-related pulmonary hypertension (PHT) and high cardiac output state with left ventricular (LV) remodelling have been described.[5] The absence of regular transfusions in TI may preserve systolic LV function, but pulmonary hypertension may develop, which also leads to heart failure, especially after the fourth decade, a decade later than in those having TM.[6] Even though systolic LV function is preserved, pulmonary pressure increases, representing the typical finding in TI patients.

In this report, a case of TI is presented in which right-sided failure and degenerative process in the electrical system of the myocardium developed in the absence of pulmonary hypertension, left ventricular failure, and iron overload.

CASE REPORT

A 57-year-old man was referred to our department with dyspnea, orthopnea, easy fatigue, dizziness, and syncope. He was diagnosed as having TI when he was 20 years old. There was a history of short-term iron replacement therapy, but no transfusions. Physical examination showed jaundiced conjunctiva, a pale face, apparent forehead, no lymphadenopathy, jugular venous distention (10 cm), hepatosplenomegaly, and edema of the lower limbs. His pulse was irregular with a rate of 34 beats/min and his blood pressure was 75/40 mmHg. Cardiac examination revealed vivid parasternal impulse, gallop rhythm, and a grade V/VI pansystolic ejection murmur over the left forth and fifth intercostal spaces at the parasternal border. Hemoglobin level was 7.1 g/dl and hematocrit was 22.7%. White blood-cell and platelet counts, and serum ferritin level were normal. Peripheral blood smear showed microcytic and hypochromic anemia and a high number of erythroblasts and platelets. The mean cell volume was 74 fl and the mean cell hemoglobin was 24 pq. Hemoglobin electrophoresis showed elevated levels of hemoglobin A2 and hemoglobin F.

Twelve-lead electrocardiography showed no atrial activity and a distal irregular escape rhythm with QRS duration of 120 milliseconds at a mean rate of 35 beats/min (Fig 1). Chest radiography showed cardiomegaly, and dilatation of the main pulmonary artery. Echocardiographic examination showed left atrial dilatation, normal left ventricular size and function, posterior mitral valve and annular calcification, dilatation of the right ventricle and atrium, decreased right ventricular systolic function, and pericardial effusion (15 mm) localized to the right heart chambers. There was severe tricuspid regurgitation and pulmonary artery pressure derived from the tricuspid regurgitation trace was 35 mmHg. Doppler echocardiography documented the absence of A wave both in the tricuspid and mitral valve flows. During right heart catheterization, pulmonary artery pressure was 28/18 mmHg (mean 22 mmHg), pulmonary capillary wedge pressure was 14 mmHg, and pulmonary vascular resistance was 178 dyn·s/cm^5. Coronary angiography showed normal coronary arteries. Electrophysiologic study demonstrated electrical inactivity in the right atrium. Atrial pacing with maximum output yielded no atrial response. Coronary sinus recordings were not obtained due to technical reasons. His bundle recording revealed a ventricular rhythm and a retrograde conduction from the ventricle to His with a conduction time of 95 msec (Fig 2). The V-V intervals ranged from 902 msec to 1630 msec. The QRS and QT intervals were 120 msec and 406 msec, respec-

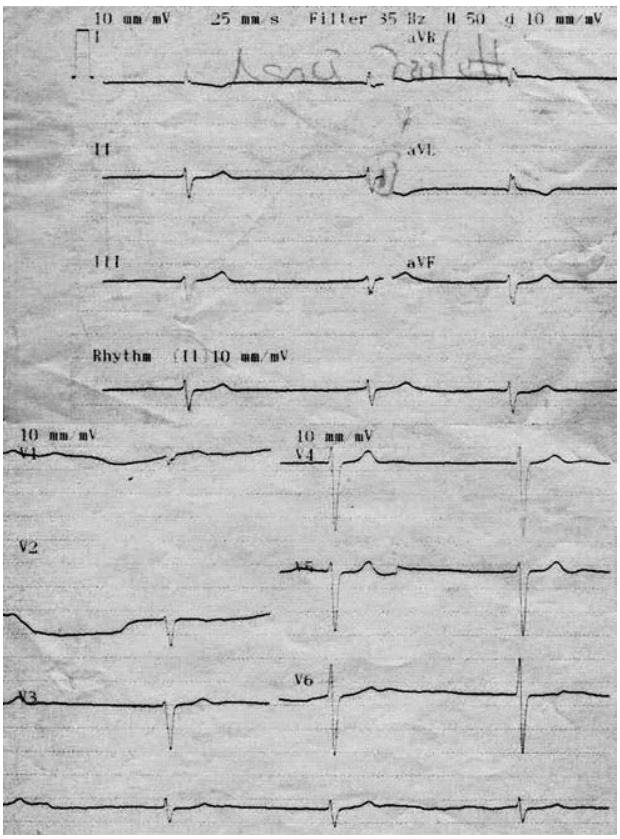

Figure 1. Admission 12-lead electrocardiogram. An escape distal rhythm with QRS duration of 120 milliseconds. The heart rate was 35 beats/min and no P wave was demonstrated.

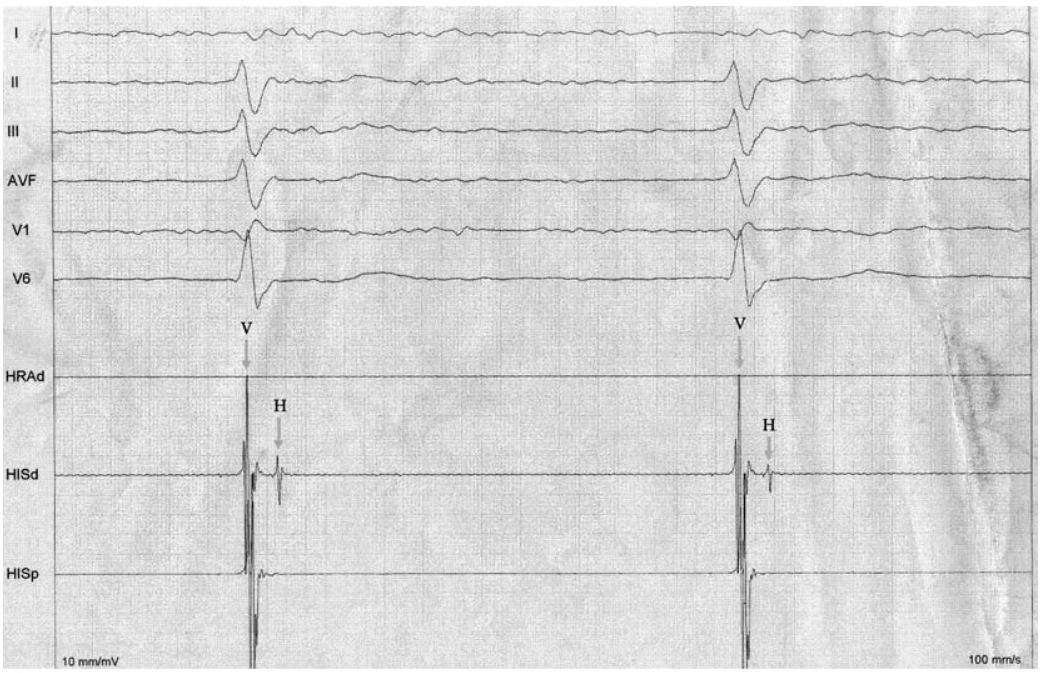

Figure 2. Intracardiac electrogram showing right atrial inactivity and retrograde His bundle activation in His bundle recordings.

tively. These findings necessitated implantation of a VVIR pacemaker. Implantation was performed after erythrocyte transfusion without any complications. During a six-month follow-up, the patient felt well, his functional capacity was NYHA class II, and his basic rhythm was widened QRS arrhythmia with a rate of 20 beats/min.

DISCUSSION

Atrial paralysis or standstill refers to the absence of electrical and mechanical activity in the atria. It may be transient or persistent, and complete or partial, with idiopathic, sporadic, or familial forms. Association with Emery-Dreifuss muscular dystrophy (X-linked)[7] or Kugelberg-Welander syndrome (autosomal recessive)[8] have been shown. It may often be associated with muscular dystrophy-related cardiomyopathies. Three cases of atrial standstill associated with a familial Ebstein's anomaly have been reported.[9,10] Tsutsugamushi myocarditis, congestive heart failure, and persistent atrial standstill have been reported in one case.[11] Demiralp et al.[12] described electrical activity in the left atrium, no electrical activity in the right atrium, no P wave on the surface ECG, and a wide QRS escape rhythm in a young patient with partial atrial standstill. The disease may be progressive or it may primarily be confined to the right atrium and atrioventricular junction region.

Compared to TM, cardiac involvement may be different in TI because patients generally have low hemoglobin levels and lower iron loads. Aessopos et al.[5] evaluated 110 patients with TI and concluded that PHT was the leading cause of congestive heart failure (CHF). High carbon monoxide resulting from chronic tissue hypoxia and increased pulmonary vascular resistance were the main contributing factors. The main electrocardiographic findings in 110 patients with TI were right or left ventricular hypertrophy in 13.6% and 7.2%, respectively, biventricular hypertrophy in 6.3%, right QRS axis deviation in 6.3%, and right bundle branch block in 8.1%. Premature atrial contraction and atrial fibrillation were observed in 16.3%, and 6.3%, respectively. Pericarditis was found in 8.1%. Pericardial thickening was detected in 38 patients (34.5%), 16 of whom also had small or moderate effusion without acute symptoms. Valvular leaflet thickening was found in 48.1% and endocardial calcification was present in 20.9%. Regurgitation was encountered at all valvular sites. Pulmonary acceleration time was lower than 120 msec in 64.5%, indicating increased pulmonary artery pressure. Tricuspid gradient higher than 30 mmHg indicating PHT was found in 59.1%. Pulmonary hypertension was the main cardiac finding. The authors concluded that PHT developed with age, caused right ventricular deterioration, and was the main cause of CHF.[5]

To the best of our knowledge, this is the first case of atrial standstill associated with TI. We based our diagnosis of atrial standstill on the lack of electrical and mechanical activities and failure to pace the right atrium during electrophysiologic study, along with an idioventricular escape rhythm and retrograde His bundle activation and the lack of P waves on the surface ECG. These findings suggested that the pathogenesis of atrial standstill might be related to atrial myocardial involvement in TI. In our case, iron overload was not a primary cause of atrial involvement because there was no history of transfusion and the ferritin level was normal. Serum ferritin level is the most widely used marker of iron load, although it is not the best. Pericarditis and myocarditis may be the other possible causes of atrial involvement and standstill in our case. Right ventricle dilatation, advanced tricuspid regurgitation, and pericardial effusion localized to the right heart chambers, together with atrial standstill suggest that right heart involvement in TI may occur primarily.

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