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RAPID COMMUNICATION

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Third-degree atrioventricular block and pacemaker implantation in a man with severe hemophilia A: A case report

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Abstract

Hemophilia A (HA) is a rare bleeding disorder characterized by reduced factor VIII (FVIII) activity and consequently spontaneous bleeding. Since the introduction of prophylaxis with safer FVIII concentrates, people with HA are ageing. Interestingly, they are developing cardiovascular diseases as their non-hemophilia counterparts. We describe a 48-year-old patient with severe HA who presented a third-degree atrioventricular block. A DDDR pacemaker was implanted under supervision of the Hematology Clinics. There were no adverse events during the procedure. The procedure was safe, and it should be performed under the supervision of a hemophilia expert.

KEYWORDS

hemophilia A, pacemaker, third-degree atrioventricular block

1 | INTRODUCTION

Hemophilia A (HA) is a hereditary bleeding disorder characterized by a reduced/abolished activity of factor VIII (FVIII). A 48-year-old man was admitted to the hospital, after having a syncope. He had severe HA (FVIII activity < 1%), and self-infused plasma-derived (pd) FVIII 20 IU/kg twice weekly as prophylaxis. He was a smoker and he had arterial hypertension and gout. Admission electrocardiogram showed an atrioventricular dissociation with P frequency of 100 bpm and QRS frequency of 40 bpm. P-wave was normal (approximately 2.0 mm of height on leads II and III; 0.08 s of duration), and QRS suggested a right bundle branch block (0.13 s of duration, a terminal R-wave in lead V1, and an S-wave longer than R-wave in lead V6). A third-degree atrioventricular block was diagnosed without instabilities. A DDDR pacemaker was implanted via left subclavian vein. He received a *bolus* of 35 IU/kg of pdFVIII few minutes before the procedure, and maintenance with 30 IU/kg twice daily for 3 consecutive days, according to international guidance, which recommends an initial FVIII *bolus* dose of 20-40 IU/kg, to reach plasmatic activity of ~40-80 IU/dL, and daily maintenance of 10-40 IU/kg, to reach plasmatic activity of ~20-80 IU/dL, for at least 3 days.[1] Then regular prophylaxis with pdFVIII was resumed. No abnormal bleeding was reported during or after the procedure. Small hematomas at the puncture and the battery sites resolved in 1 week.

Before discharging, Holter analysis demonstrated a normal mean heart rate, with complete ventricular capture (v-paced 100% of the time) and < 1% of ventricular ectopia. Transthoracic echocardiography showed normal cardiac chambers dimensions, eccentric hypertrophy of the left ventricle, without segmental deficits, and normal systolic-diastolic functions. He was discharged home with the diagnosis of myocardiopathy of unknown etiology. Since third-degree atrioventricular among people with hemophilia is a rare event, we could not predict a specific cause for

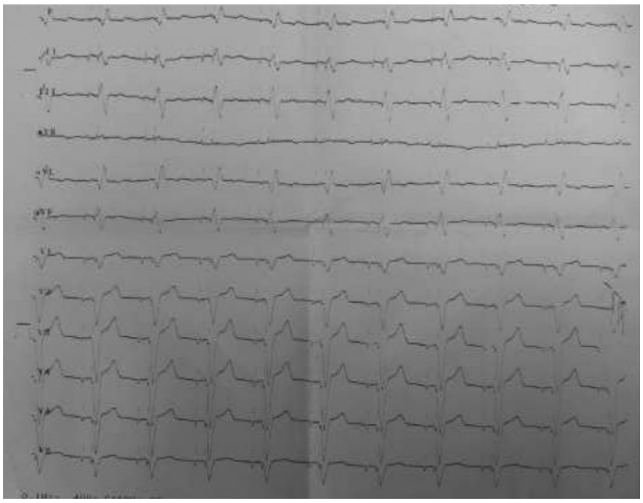

FIGURE 1 Control electrocardiogram of a severe hemophilia A man who developed a symptomatic idiopathic third-degree atrioventricular block who had a DDDR pacemaker implanted (device settings: 6 channels at 10.0 mm/mV and 25.0 mm/s)

it, different from the causes of third-degree atrioventricular block among non-hemophilia people (eg, idiopathic fibrosis, ischemia, or toxicity). Semesterly visits to the Cardiologist were scheduled (Figure 1).

There are few reports of pacemaker implantations in people with HA in the literature. A DDD pacemaker was implanted in a 62-year-old man patient due to an ischemic cardiac disease with symptomatic bradycardia.[2] Other three people with HA 60 years or older had a permanent pacemaker implantation, but those indications were not described.[3] Pacemakers are not contraindicated in hemophilia patients. However, until the first half of the 20th century median life expectancy among people with HA was about 20 years,[4] and they would seldomly present a cardiovascular disease, which is known to be more prevalent as the person gets older.[5] Life expectancy of people with HA has increased, mainly due to the introduction of prophylaxis, the faster access to FVIII to home therapy of bleeding episodes, and the use of safer FVIII together with more effective antiviral treatments.[6] With fewer patients dying from hemorrhages or viral infections, they are growing older and developing cardiovascular diseases,[4,7] among them symptomatic bradycardias. Therefore, Cardiologists should be aware in the near future, because some people with HA may need cardiovascular interventions. The close contact to a hemophilia treatment center can help solving doubts and ensuring safe and successful results.[8]

CONFLICT OF INTEREST

RMC received honoraria for participating as a speaker at scientific/educational meetings and travel support for scientific meetings from Takeda and Hoffman-La Roche. AMV received honoraria for participating as a speaker at scientific/educational meetings from Takeda and travel support for scientific meetings from Takeda, Hoffman-La Roche, Novo Nordisk and BioMarin. BPD and AMNJ declare they have no interests which might be perceived as posing a conflict of bias.

ETHICAL APPROVAL

HemoCardio Study was approved by the local Committee on Ethics in Research (CAAE 86067818.6.0000.5195). The patient signed the Informed Consent Form and permitted us to publish his case.

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