

INTRODUCTION

- Fabry disease is a lysosomal storage disorder that can result in the accumulation of globotriaosylceramide in the heart¹.
- We present the case of a patient with Fabry disease who developed an advanced inter-atrial conduction delay and atrial fibrillation, a condition known as Bayés' syndrome^{2,3}.

CASE REPORT

- A 72-year-old female with genetically confirmed Fabry disease who had previously declined enzyme replacement therapy presented for evaluation of a recent episode of palpitations.
- The episode was brief, resolved spontaneously, and had no associated symptoms. She was otherwise asymptomatic and had a normal cardiovascular physical exam.
- Her 12-lead electrocardiogram revealed P-wave prolongation of greater than 120 milliseconds measured in lead V1, notched P-waves, and no ischemic changes.
- Subsequent ambulatory rhythm monitoring (Fig. 1) was notable for sinus bradycardia with first-degree atrioventricular block, sinus P-wave terminal delay with biphasic morphology, and 14 episodes of non-sustained atrial tachycardia.

CASE REPORT (continued)

- A cardiac MRI demonstrated normal-sized atria, progressive asymmetric septal hypertrophy, and progressive patchy mid-myocardial late gadolinium enhancement within the left ventricle compared to prior scans.

FIGURES



Figure 1: Panel A - ambulatory rhythm monitor showing sinus bradycardia with first-degree AV block and sinus P-wave terminal delay with biphasic morphology. Panel B - ambulatory rhythm monitor showing sinus bradycardia with first-degree AV block and a pair of PACs with wide, fragmented P-waves.

DECISION MAKING

- Her electrocardiography demonstrated advanced inter-atrial block (A-IAB), a result of impairment of Bachmann's bundle⁴.
- Given her A-IAB and history of atrial tachycardia, she was diagnosed with Bayés' syndrome.
- She began treatment with enzyme replacement therapy given this evidence of progressive intracardiac accumulation of globotriaosylceramide.
- Months later, she followed up in our clinic where asymptomatic atrial fibrillation with slow ventricular response was found. Anticoagulation was initiated.

CONCLUSION

- While Fabry disease is an established cause of cardiac conduction disease, Bayés' syndrome due to Fabry disease has never been reported.
- In addition to considering A-IAB and Bayés' syndrome consequences of Fabry disease, providers need to be able to recognize the electrocardiogram findings of A-IAB and understand the elevated risk of developing atrial arrhythmias and subsequent thromboembolic events that it entails.

REFERENCES AND DISCLOSURE

References are available upon request.
The authors report no conflict of interest.

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