CASE REPORT

Cystic tumour of the atrioventricular node: treatment dilemma

Wael Abuzeid, Robert B H Myers

Sunnybrook Health Sciences
Centre, Toronto, Ontario,
Canada

Correspondence to

Dr Wael Abuzeid,
wael.abuzeid@mail.utoronto.ca

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SUMMARY

An asymptomatic 18-year-old black woman was found to have an incidental finding of third-degree atrioventricular (AV) block on a routine ECG. 2-Dimensional transthoracic echocardiography showed a heterogeneous mass in the region of the peri-membranous septum in the AV node area. A cardiac MRI showed a well-defined cystic mass arising from the right side of the interatrial septal wall. An MRI compatible permanent pacemaker was implanted with plans to monitor the tumour with non-invasive imaging. Cystic tumour of the AV node is a rare primary cardiac tumour. It is known as the smallest and most common primary cardiac tumour that can cause sudden death. All previous six cases of living patients with cystic tumour of the AV node reported in the literature were females and symptomatic. We present a rare case of a patient with cystic tumour of the AV node, and we highlight the treatment dilemmas for this condition.

BACKGROUND

There are no established guidelines for the management of patients with cystic tumours of the atrioventricular (AV) node. Patients can present with sudden cardiac death due to arrhythmias. They can also present in an asymptomatic state (ie, incidental finding). This is a rare condition, and management is based on expert opinion. Reporting these rare cases is crucial to aid in the clinical management of these patients.

CASE PRESENTATION

An 18-year-old black woman was referred for cardiac assessment after an incidental finding of third-degree AV block on a routine ECG ordered by her family physician. She was asymptomatic with no history of syncope, palpitations or prior hospitalizations. Her medical and family histories were unremarkable.

Physical examination revealed an 18-year-old woman with a blood pressure of 90/60 mm Hg and a regular heart rate of 42 bpm. Cardiovascular examination demonstrated normal jugular venous pressure, carotid impulse and heart sounds with no extra sounds or murmurs.

INVESTIGATIONS

The 12-lead ECG showed third-degree AV block and a narrow complex junctional escape with right axis and incomplete right bundle-branch block (figure 1). 2-Dimensional transthoracic echocardiography showed a heterogeneous mass measuring

16 mm in the region of the peri-membranous septum at the base of the septal leaflet of the tricuspid valve (figure 2). A cardiac MRI showed a well-defined cystic mass measuring 23×15 mm arising from the right side of the interatrial septal wall immediately above the medial leaflet of the tricuspid valve and in continuity to the orifice of the coronary sinus (figure 3).

DIFFERENTIAL DIAGNOSIS

- ▶ Lyme disease
- ▶ Medication effects
- ▶ Hypothyroidism
- ▶ Congenital AV block
- ▶ Teratoma
- ▶ Mesothelial cysts
- ▶ Bronchogenic cysts

TREATMENT

After consultation with cardiac surgery and electrophysiology services, it was decided to implant an MRI compatible permanent pacemaker and monitor the tumour with non-invasive imaging for signs of tumour progression or other mechanical complications such as compression of surrounding structures.

OUTCOME AND FOLLOW-UP

The patient is followed up for tumour progression with imaging. She received an MRI compatible dual-chamber permanent pacemaker. She is doing well on follow-up.

DISCUSSION

Cystic tumour of the AV node is a rare primary cardiac tumour that was first described in 1911 in a 5-year-old boy and has been known as the smallest and most common primary cardiac tumour that can cause sudden death.¹ It is difficult to determine the incidence of this disease, as most cases are not discovered until autopsy with careful microscopic examination of the conduction system.² All previous six cases of living patients with cystic tumour of the AV node reported in the literature were females with an average age of 41 years.² Unlike our asymptomatic patient, the symptoms described by these patients included palpitations, dyspnoea and dizziness.

Tumour size can range from 0.5 to 30 mm with no relationship between size and occurrence of arrhythmia.³ These patients can have variable degrees of heart block that can progress to complete

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Figure 1 ECG showing complete heart block. Red arrow showing P waves marching along.

heart block.³ Sudden cardiac death is known to occur in patients with primary cardiac tumours. In a time span from 1994 to 2012, a UK database identified 164 primary cardiac tumours. Eleven of those 164 cases (9.7%) were associated with cardiac death. Four of those 11 cases comprised cystic tumours of the AV node.² Echocardiography, cardiac CT and MRI have been used to diagnose this condition.

Given the limited number of living cases with this condition, treatment decisions are based on case reports. Complete resection of the tumour has been suggested, although it is unclear if tumour resection would reduce the risk of sudden cardiac death once AV nodal destruction has taken place. Treatment with permanent pacemaker implantation was used in several cases with varying degrees of heart block. The role of implantable cardioverter defibrillator for primary prevention for ventricular arrhythmia is unclear.

Figure 2 Short-axis 2-dimensional transthoracic echocardiographic view showing the atrioventricular tumour (arrow). AV, aortic valve; LA, left atrium; RA, right atrium.

Figure 3 MRI view showing the atrioventricular tumour (green label). LA, left atrium; RA, right atrium; RV, right ventricle; LV, left ventricle.

In summary, we present the seventh reported living case of a patient with cystic tumour of the AV node. After discussing the risks and benefits of treatment options, the patient opted for close follow-up for tumour progression with imaging versus surgical resection. She received an MRI compatible dual-chamber permanent pacemaker and is currently being closely followed.

Learning points

- ▶ Cystic tumour of the atrioventricular node is a rare condition that can present incidentally with no symptoms or can present with sudden cardiac death.
- ▶ Treatment decisions are based on expert opinion and prior case reports. This includes tumour resection, pacemaker implantation and/or implantable cardioverter defibrillator implantation.
- ▶ Multimodality imaging with echocardiography, CT and MRI is crucial in diagnosis and follow-up for tumour progression.

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