



Case Report

Occult etiologies of complete atrioventricular block: Report of two cases

Neeta Bachani^a, Gopi Krishna Panicker^b, Jaipal Jadwani^a, Yash Lokhandwala^{c,*}^a Holy Family Hospital, Mumbai, India^b Quintiles Cardiac Safety Services, Mumbai, India^c Arrhythmia Associates, Mumbai, India

ARTICLE INFO

Article history:

Received 11 March 2016

Accepted 27 July 2016

Available online 6 August 2016

Keywords:

Complete atrioventricular block

Sarcoidosis

Non-Hodgkin's lymphoma

ABSTRACT

In patients presenting with complete atrioventricular (AV) block, the common causes are degeneration of the conduction system, acute myocardial infarction, congenital and metabolic disorders (such as azotemia). However, at times, no cause can be ascribed and the label congenital or degenerative is applied depending on the patient's age and the QRS complex width. We present two cases of patients with complete AV block, who were subsequently found to have rare etiologies – sarcoidosis (with isolated feature of AV block) and non-Hodgkin's lymphoma.

© 2016 Cardiological Society of India. Published by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Complete atrioventricular (AV) block is regarded as a serious condition requiring prompt therapy. Studies evaluating its incidence and prevalence amongst patients with known heart disease and screening of normal cohorts confirm that the most common causes of complete AV block are degeneration of the conducting system, congenital, acute myocardial infarction and metabolic disorders (such as azotemia). However, in a significant number of cases, no specific cause can be ascribed and a congenital or degenerative cause is inferred based on the patient's age and the QRS complex width. We present two cases in which the patients had complete AV block due to rare etiologies.

2. Case 1

A 30-year-old man of Indian origin without any comorbidities was admitted in a hospital in Dubai with a history of unexplained giddiness and chest heaviness since 1 day. The ECG showed a complete AV block with a wide QRS escape rhythm (Fig. 1a) and the patient underwent temporary transvenous pacing along with supportive therapy (atropine/dopamine/IV fluids). The troponin I evaluation showed an increasing trend for 2 days and then it decreased. The echocardiogram was normal. The escape rhythm

gradually changed with the QRS becoming relatively narrow (Fig. 1b) and after 4 days the AV conduction recovered, albeit with a prolonged PR interval of 240 ms. The pacing electrode was withdrawn and the patient was discharged.

The patient subsequently came to Mumbai and was referred to the hospital for further evaluation. The coronary angiogram evaluation was normal while a cardiac MRI evaluation showed mid-myocardial scarring along with inflammatory changes in the basal septal myocardium (Fig. 2); there was significant mediastinal and hilar adenopathy with sub-pleural and peri-bronchovascular nodules. Overall, these findings suggested sarcoidosis with cardiac involvement. Further work-up showed the following: SGOT-42 IU/l, SGPT 82 IU/l, Serum calcium 9.6 mg%, ESR 16 and mildly elevated ACE level of 58 µg/l. The Mantoux test was negative.

A CT-guided sub-pleural lymph node biopsy revealed non-necrotizing granulomatous inflammation of undetermined etiology. A 24-h Holter showed no evidence of AV block and an average HR of 72 bpm. The patient was started on oral prednisolone 40 mg once a day. Over the next 2 weeks his PR interval normalized; prednisolone was then tapered off. At 3 months follow-up, he is on prednisolone 5 mg daily and his ECG remains normal.

3. Case 2

A 68-year-old man had been diagnosed to have idiopathic thrombocytopenic purpura (ITP) since 2012 and had undergone splenectomy after inadequate response to medical therapy. Eight months after the surgery, the patient started again having

* Corresponding author at: 502 A, Leela Business Park, M.V. Road, Andheri (East), Mumbai 400 059, India.

E-mail address: yashlokhandwala@hotmail.com (Y. Lokhandwala).

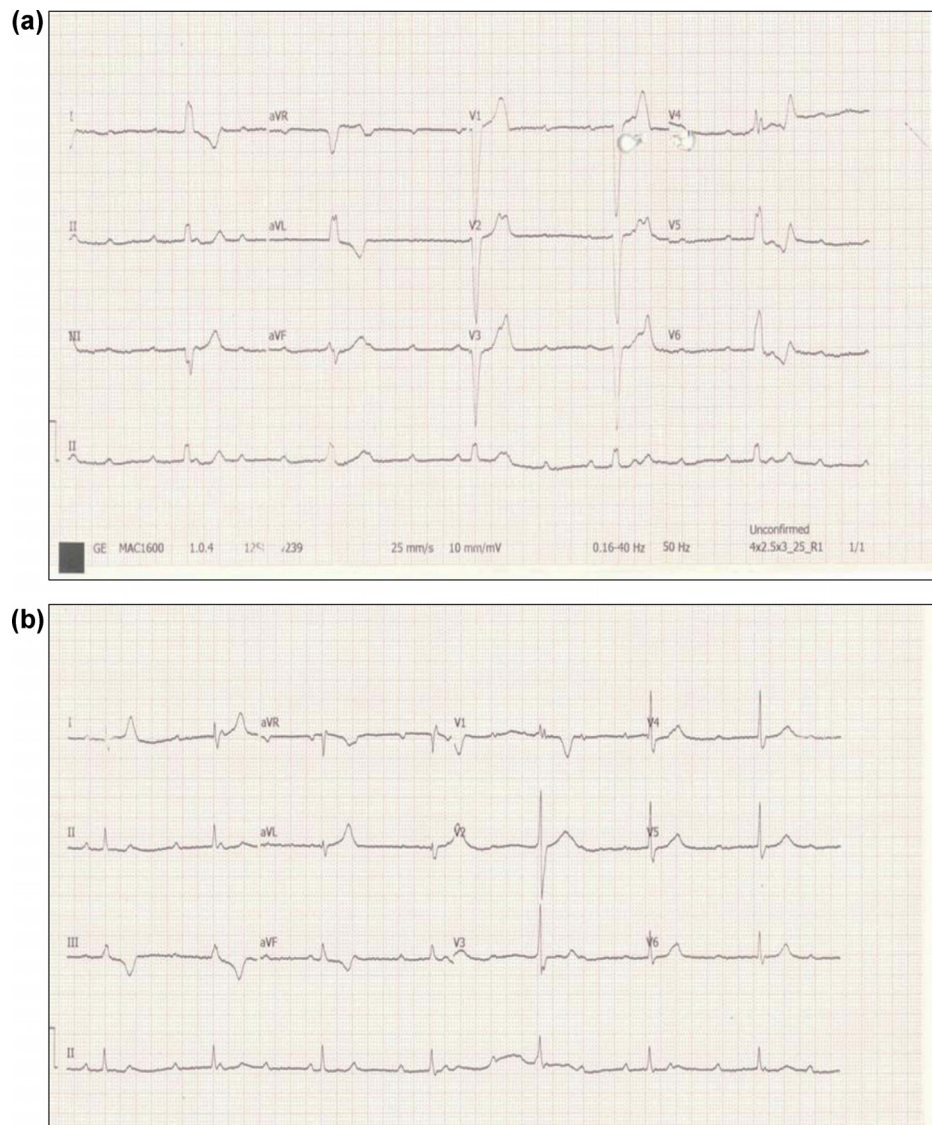


Fig. 1. (a) ECG on presentation. Sinus tachycardia, complete AV block, wide QRS escape rhythm. (b) Sinus tachycardia and complete AV block persist, but the escape rhythm shows relatively narrow QRS complexes (IRBBB).

symptomatic episodes of thrombocytopenia. In January 2014, he was re-evaluated by a hematologist and started on Eltrombopag (Revolad), which was used intermittently as per the platelet count.

The patient was admitted in November 2014 for fever after getting repeated bouts of fever with thrombocytopenia. The etiology of fever could not be established following an extensive work-up including echocardiography and whole body PET CT scan. However, the patient responded to broad-spectrum antibiotics and was subsequently discharged.

The patient was admitted in March 2015, for weakness and giddiness and was found to have complete AV block with a narrow QRS escape rhythm. Initially a conservative therapeutic approach was adopted since there appeared to be a stable ventricular rate of 40 beats/min. A repeat echocardiogram was normal. Since the rhythm did not settle over 7 days, an AV sequential pacemaker was implanted.

The patient was re-admitted in May 2015 with fever and thrombocytopenia. As a part of work up for identifying the cause of the fever, the patient again underwent an echocardiography, which surprisingly revealed a large echo-dense mass attached to the left atrial wall above the mitral annulus. There was also a large

echo-dense mass in the AV groove (Fig. 3a), which was seen extending outwards on both sides. There was also infiltration of the interatrial septum and interventricular septum (Fig. 3b) along with a small pericardial effusion. The cardiac CT evaluation revealed encasement of proximal right and left coronary arteries; the mass also invaded the central fibrous body area and eroded the left atrial wall, thence protruding into its cavity (Fig. 4).

The whole body PET CT scan was repeated which showed:

- Active disease demonstrating high grade metabolic activity involving soft tissue nodular masses in the interatrial septum and left atrium extending into mitral valve, interventricular septum, aorto-pulmonary recess and along posterior wall of right atrium.
- Metabolically active nodular soft tissue lesions in abdomino-pelvic region.
- Focal inflammatory activity involving wall thickening of abdominal aorta.

A CT-guided biopsy of the peritoneal deposit showed non-Hodgkin's lymphoma (NHL) of T cell variety.

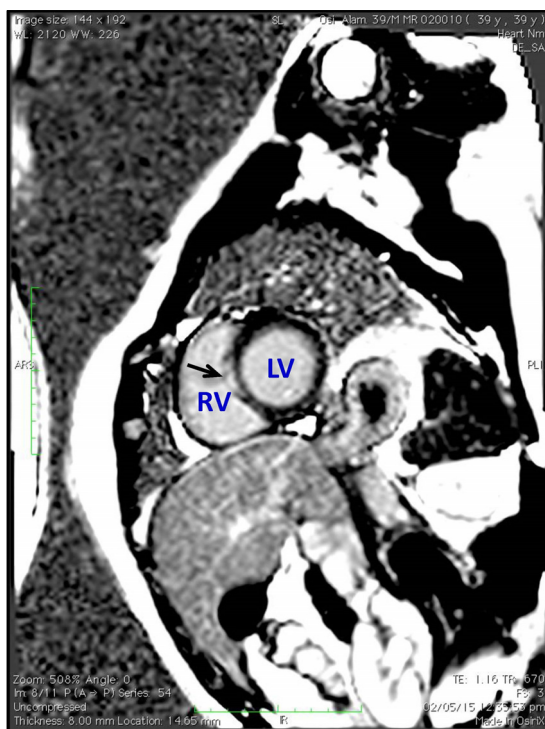


Fig. 2. Cardiac MRI: phase sensitive inversion recovery image taken in the short axis plane shows septal scar (solid black arrow) as an area of persistent enhancement at 15 min delayed post-contrast imaging. LV, left ventricle; RV, right ventricle.

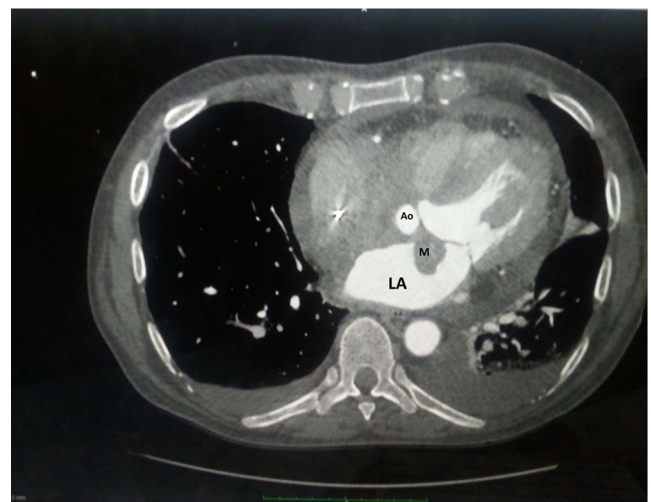


Fig. 4. Cardiac CT scan image showing the mass around the aortic root and invading the left atrial wall.

4. Discussion

In patients with symptomatic AV block, permanent pacing is needed unless there is a reversible cause, such as inferior wall myocardial infarction or metabolic disturbance. These are usually easily diagnosed. Even viral myocarditis would be picked up on echocardiography. As seen in our patient, sarcoidosis would have been missed, if not thought of and assiduously looked for.

The common organs involved in sarcoidosis include lymph nodes, skin, lungs and eye. The heart is involved in up to 30% of patients with sarcoidosis.¹ Only 40–50% of patients with cardiac sarcoidosis diagnosed at autopsy have the diagnosis made during their lifetime. Cardiac sarcoidosis can manifest itself as AV block, ventricular arrhythmias, heart failure, pericardial effusion, pulmonary hypertension and ventricular aneurysms. Presentation with symptomatic AV block is rare and is seen in less than 2% of patients. The mechanism is either involvement of the interventricular septum or of the AV nodal artery. Complete AV block is considered a high-risk category with the prognosis depending on the site of the block. If the site of block is the AV node, the escape rhythm may recover; the AV block in this setting may respond to vagolytic or adrenergic drugs. If the site of the block is the His bundle or the right or left bundle branches, then it is less likely to be reversible and is associated with higher risk of ventricular fibrillation, heart failure and increased mortality. Pacemaker implantation is the only reasonable treatment when reversible causes cannot be identified.

Cardiac involvement in malignancy is more commonly secondary and the pericardium is the commonest location.² These are usually obvious at echocardiography. In our patient, the unique feature was that the septal involvement which led to AV block was not apparent at initial presentation. Primary cardiac lymphomas are often Hodgkin's variety of lymphoma. In this case, the patient was assessed as having primary NHL (extra-cardiac involvement was minimal) and the mass possibly started from area of central fibrous body, which clinically manifested as complete AV block. Eltrombopag is a thrombopoietin receptor agonist. It improves the platelet count stimulating the cascade of megakaryocytes formation and differentiation. The US FDA approved eltrombopag in 2008 for Immune/idiopathic thrombocytopenia purpura with inadequate response to steroids, immunotherapy, immunoglobulins and/or splenectomy. It is supposed to be used intermittently to improve platelet count >50,000/mm.³ A recently concluded "EXTEND" trial has proven its efficacy and safety in long-term

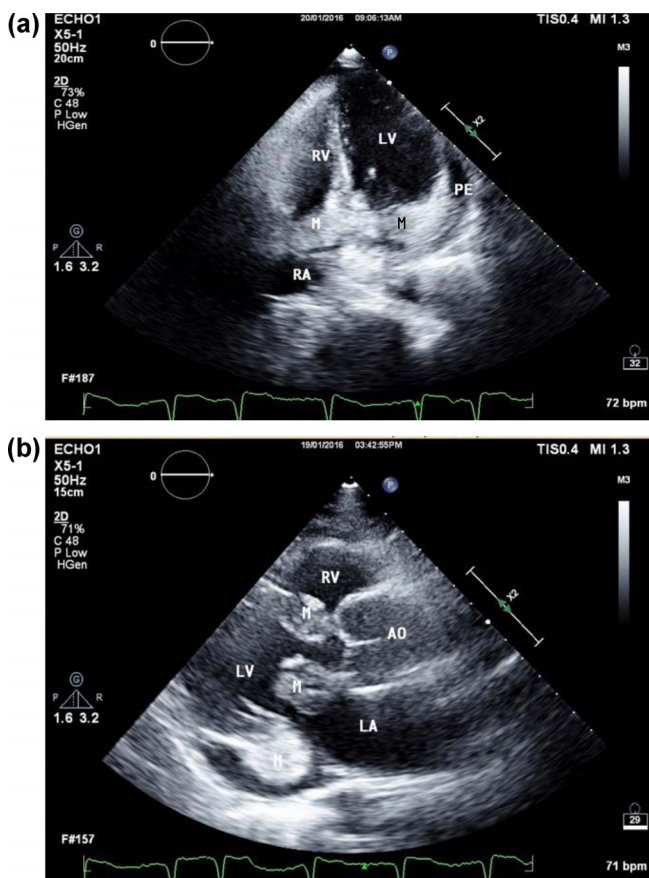


Fig. 3. (a) Apical 4 chamber view. Mass (M) along the AV groove. LV, left ventricle; RV, right ventricle; RA, right atrium; PE, pericardial effusion. (b) Parasternal long axis view. Masses seen around the mitral annulus and interventricular septum; LA, left atrium; Ao, aorta.

use.³ There is concern about increased incidence of lymphomas with long term use. In an evaluation of 419 patient years of long-term use (>2 years), 2 patients developed lymphoma.⁴ Our patient used eltrombopag continuously for more than 14 months. The dilemma remains whether the NHL was incidental or was related to prolonged use of eltrombopag.

In conclusion, one must be alert for unusual etiologies of AV block, especially when there is a fluctuant course or atypical presentation. This would allow for disease modifying treatment and sometimes eliminate the need for pacing.

Conflicts of interest

The authors have none to declare.

Acknowledgement

Dr Alpa Bharati, Radiologist, Lokmanya Tilak Municipal General Hospital, Mumbai.

References

1. Ipek E, Demirelli S, Ermis E, Inci S. Sarcoidosis and the heart: a review of the literature. *Intract Rare Dis Res*. 2015;4:170–180.
2. Grebenc ML, Rosado de Christenson ML, Burke AP, Green CE, Galvin JR. Primary cardiac and pericardial neoplasms: radiologic–pathologic correlation. *Radiographics*. 2000;20:1073–1103.
3. Saleh M, Bussel J, Cheng G, et al. Safety and efficacy of eltrombopag for treatment of chronic immune thrombocytopenia: results of the long-term, open-label EXTEND study. *Blood*. 2012;121(3):537–545.
4. Laroussi L, Badhwar N. Atrioventricular conduction disease and block. *Card Electrophysiol Clin*. 2014;6:445–458.