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MINI-FOCUS ISSUE ON ELECTROPHYSIOLOGY AND PACING

BEGINNER

CASE REPORT: CLINICAL CASE

Primary Cardiac Lymphoma Manifesting as an Atrioventricular Block in a Renal Transplantation Recipient

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ABSTRACT

Our report illustrates an atrioventricular block due to primary cardiac lymphoma attached to the right atrial septum that was rapidly reversible by surgical debulking and effective chemotherapy without the need for a permanent pacemaker. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:600–3) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 56-year-old man was admitted to our hospital for fever and dyspnea on exertion, which lasted for 1 week.

PAST MEDICAL HISTORY

The patient was diagnosed with hypertension and end-stage renal disease in 1989 and received a right kidney transplant from his father in 1992. In 2017, he received a left kidney transplant from his spouse owing to chronic allograft injury. After the second transplantation, he suffered from acute rejection and was treated with immunoglobulin infusion, steroid pulse therapy, and plasmapheresis.

LEARNING OBJECTIVES

- Rarely, cardiac tumors cause atrioventricular blocks.
- When patients are treated with immunosuppressive therapies (i.e., post-transplant), there is a high risk of lymphoproliferative disorders, including lymphoma.
- When atrioventricular blocks are caused by an infiltration of lymphoma, they can be resolved by an effective therapy for the tumor.

INVESTIGATIONS

Chest radiography detected marked cardiomegaly. His electrocardiogram (ECG) showed a 4:3 Mobitz 1 atrioventricular (AV) block which was not seen 2 months earlier (**Figures 1A and 1B**). Transthoracic echocardiography (TTE) detected a large amount of pericardial effusion without hemodynamic compromise and a mobile heterogeneous echogenic mass (2 × 3 cm) attached to the septal wall and another mass in the pericardial space attached to the right atrium (**Figure 2**). Cardiac magnetic resonance failed to perform properly due to poor patient cooperation (Supplemental Figure 1), and positron emission tomography-computed tomography (PET-CT) revealed a large hypermetabolic mass in the right

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atrium (Figure 3). No other significant hypermetabolic uptake was seen throughout the entire body.

MANAGEMENT

After consultation with a thoracic surgeon, we decided to perform a surgical right atrial mass removal for debulking and improvement of mass effects, in addition to the histological diagnosis. His AV block progressed to a 2:1 AV block with a long PR interval of 330 ms (Figure 1C). On the fourth hospital day, the ECG showed a high-grade AV block (Figure 1D). Cardiac mass removal and surgical pericardial drainage were performed on the fifth hospital day. Temporary pacemaker insertion via the femoral vein was performed for a complete AV block at the time of surgery because of the capture failure of the epicardial pacing lead. On post-operative day (POD) 1, intermittent conducted beats were noted. On POD 2, complete AV block improved to a 2:1 AV block, and on POD 3, 1-to-1 conduction with a long PR interval (320 ms) was shown (Figure 1E), but an intermittent 2:1 AV block was still noted on telemetry. The temporary pacemaker was removed at POD 5. After POD 8, there was no second-degree AV block on telemetry monitoring. Telemetry monitoring was performed for 2 weeks. Pathologic examination revealed diffuse large B cell lymphoma (Figure 4). Treatment with combination chemotherapy (R-CHOP [rituximab, cyclophosphamide, anthracycline, vincristine, and prednisone]) was started on POD 15 by a hematologist. After the first cycle of chemotherapy, the ECG showed a sinus rhythm with a mildly prolonged PR interval of 240 ms. After the fourth cycle of chemotherapy, ECG showed normal sinus rhythm with a PR interval of 150 ms (Figure 1F), and PET-CT revealed no hypermetabolic uptake in entire body.

FOLLOW-UP

After the sixth cycle of chemotherapy, we detected complete remission of the primary cardiac lymphoma (PCL) through PET-CT and TTE.

ABBREVIATIONS AND ACRONYMS

AV = atrioventricular

ECG = electrocardiogram

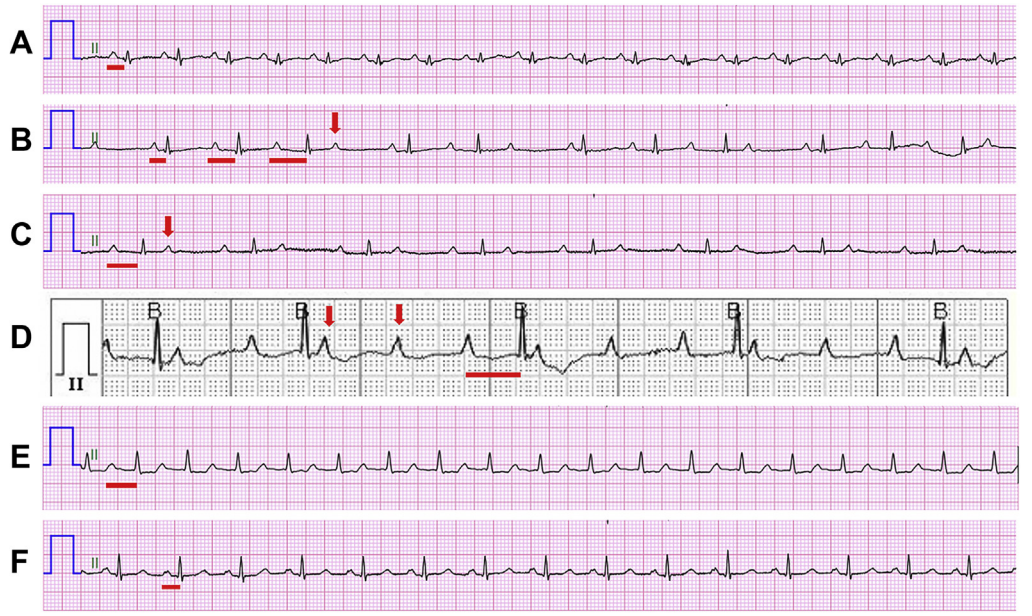
PCL = primary cardiac lymphoma

PET-CT = positron emission tomography-computed tomography

POD = postoperative day

TTE = transthoracic echocardiography

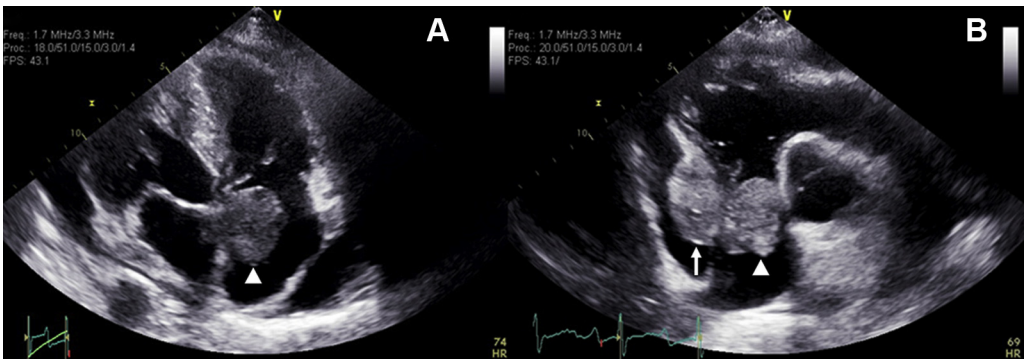
FIGURE 1 Electrocardiogram Changes



(A) Normal sinus rhythm. PR interval **(red bar)** is within the normal range. **(B)** 4:3 Mobitz 1 atrioventricular (AV) block. PR interval **(red bars)** is progressively increasing, and the fourth P-wave **(red arrow)** is blocked. **(C)** AV block progressed to a 2:1 AV block, with a long PR interval of 330 ms (**red bar** indicates conducted PR interval, **red arrow** indicates blocked P-wave). **(D)** Telemetry strip on the day of surgery shows a high-grade AV block. The conduction ratio was aggravated to a 3:1 ratio (**red bar** indicates conducted PR interval, **red arrows** indicate blocked P-wave). **(E)** Sinus rhythm with a mildly prolonged PR interval of 240 ms (**red bar** indicates conducted PR interval). **(F)** Sinus rhythm with a PR interval of 150 ms (**red bar** indicates conducted PR interval).

JACC: CASE REPORTS, VOL. 2, NO. 4, 2020
APRIL 2020:600–3

FIGURE 2 Transthoracic Echocardiography

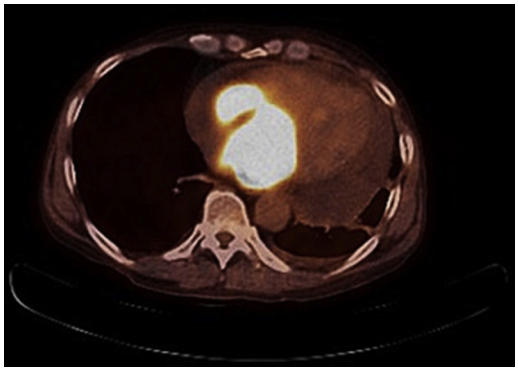


A mobile mass attached to the atrial septal wall **(arrowheads)** and another mass in the pericardial space attached to the right atrium **(arrow)**. **(A)** Apical 4-chamber view. **(B)** Parasternal short-axis view.

DISCUSSION

PCL is a rare tumor that comprises only <2% of all primary cardiac tumors (1). PCL is more likely to involve the right heart than the left heart, and the clinical manifestations of a cardiac mass are determined by its size, anatomical location, rate of growth, and invasiveness (1,2). PCL is frequently associated with an immunocompromised status, such as human immunodeficiency virus, or immunosuppressive medications (3,4). PCL commonly manifests as an AV block. Most previous case reports showed that an AV block improves after prompt diagnosis and effective chemotherapy; therefore, permanent pacemaker implantation is not necessary (3,5–8). However, in some cases, a permanent pacemaker is required due to the difficulty and delay in diagnosis and treatment (9–11). For prompt diagnosis and treatment, an interdisciplinary approach consisting of radiologists, oncologists or hematologists, cardiac surgeons, and cardiologists is required. In our case, the AV block rapidly progressed to a high-grade AV block. Although we had a backup temporary pacemaker after surgery for 5 days, the AV block improved after debulking of the right atrial mass, and the sinus rhythm improved with a normal PR interval after the first chemotherapy session.

FIGURE 3 Positron Emission Tomography-Computed Tomography

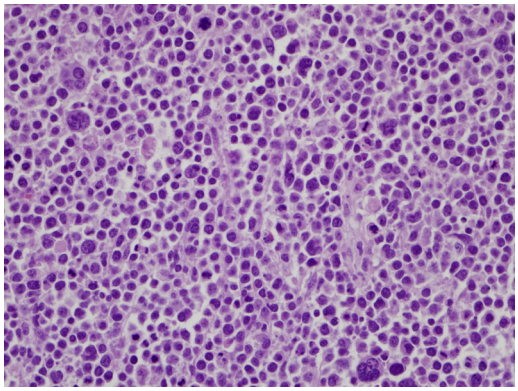


Large hypermetabolic bilobed mass in the right atrium.

Diagnostic work-up of cardiac masses involves identifying the masses by multimodality noninvasive imaging. TTE is the first diagnostic approach, and cardiac magnetic resonance is the best available diagnostic tool for predicting cardiac tumor malignancy. PET can be used to evaluate the nature of the tumor and staging (12,13). Histopathological confirmation through an endomyocardial or surgical biopsy remains the diagnostic gold standard (2,14).

The standard treatment option of PCL is not well established, and various treatment modalities, such as surgical resection, systemic chemotherapy, radiation therapy, and combined treatments, have been attempted. Sudden death may result from ventricular rupture, ventricular arrhythmias, embolization, or valve obstruction, which explains why approximately 50% of patients die within 2 months from diagnosis. About 40% to 50% of patients eligible for chemotherapy combined with immunotherapy (anty-CD20 antibodies) achieve prolonged survival or complete recovery. The long-term disease-free survival rate in patients with PCL treated with systemic chemotherapy combined with or without post-chemotherapeutic radiation therapy has been

FIGURE 4 Histologic Findings



Diffuse infiltration of intermediate to large-sized atypical lymphoid cells in the myocardium (hematoxylin and eosin staining, 100×).

reported (15). In cases of PCL with a concerning tumor size and symptoms owing to cardiac obstruction, early surgical debulking could improve hemodynamics, prevent sudden death, and confirm immunopathological diagnosis needed to determine further chemotherapy, which is proven to improve survival (16). In our case, first, we decided to perform a surgical right atrium mass excision because the patient's AV block progressively worsened, so we thought that there was not much time for diagnosis and treatment. Second, we expect that the AV block may improve after surgical debulking by mitigating mass effects. Third, we wanted to prevent the risk of tumor embolism (17,18). After completion of the initially planned diffuse large B cell lymphoma treatment, the patient should be evaluated to determine the disease response to treatment through PET-CT and followed longitudinally for relapse (19).

CONCLUSIONS

Although it is rare, the possibility of a cardiac tumor should be considered in patients with sudden AV block. In patients with PCL and AV block, it may be reasonable to delay the implantation of a permanent pacemaker because the AV block can rapidly be reversed after the appropriate chemotherapy with or without surgical removal.

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KEY WORDS cardiac mass, diffuse large B cell lymphoma, immunosuppressant, primary cardiac lymphoma

APPENDIX For a supplemental figure, please see the online version of this paper.