Primary cardiac B-cell lymphoma involving sinus node, presenting as sick sinus syndrome

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Introduction

Sick sinus syndrome generally occurs in elderly patients. Two major causes for sick sinus syndrome are fibrosis of the sinus node and surrounding atrial myocardium and external causes to suppress the sinus node function. Fibrosis is mainly age-related, but can be caused by surgery, infiltrative diseases, or congenital disorders. We experienced a patient who presented with sick sinus syndrome and rapidly developed superior vena cava (SVC) syndrome shortly after the pacemaker implant. Autopsy showed B-cell lymphoma destroying the sinus node structure, which was thought to be the cause of sinus node dysfunction.

Case report

A 96-year-old woman without previous medical history presented with recurrent syncope at our hospital. Significant offset pause from atrial fibrillation up to 7 seconds and sinus bradycardia were noted, and she was diagnosed with sick sinus syndrome. Transthoracic echocardiography demonstrated no significant findings (Figure 1A). A single-chamber pacemaker was implanted without technical difficulty or complication, and she was discharged to a nursing home. At 1-month follow-up visit, she complained of anorexia and general malaise, and physical examination showed the swelling of the face, neck, and upper extremity. A chest radiograph revealed massive pleural effusion. Echocardiography showed a poorly mobile mass that partially occupied a right atrium (Figure 1B). Computed tomography with contrast showed an 8-cm mass extending from the high lateral right atrium to the SVC (Figure 2). She was diagnosed with SVC syndrome. Cytology of the pleural effusion showed no malignancy. The patient and her family chose the palliative care, and she died after 2 months.

Autopsy findings showed that the 8-cm tumor infiltrated from a right atrium to the pericardium, SVC, and right upper and middle lobes of the lung. The tumor extended to the right atrial appendage and crista terminalis (Figure 3A). Pathologic findings showed diffusely infiltrated lymphocytes with extreme nuclear atypia and lack of cell junction. Immunochemical staining showed positive CD20 (B-cell marker) and CD79a with negative CD3 (T-cell marker) and CD5. Pathologic findings confirmed the diagnosis of diffuse large B-cell lymphoma. Macroscopic examination and microscopic analysis showed the crista terminalis and sinoatrial node structure including the sinus node artery were completely destroyed owing to the massive infiltration by lymphoma (Figure 3B).

KEY TEACHING POINTS

- Sick sinus syndrome in elderly patients is usually caused by fibrosis of sinus node and surrounding atrial myocardium or external causes to suppress the sinus node function. In rare occasions, it can be caused by primary cardiac lymphoma.
- Cardiac malignancies are a rare disease and delayed diagnosis can lead to a poor prognosis.
- It is useful to repeat echocardiography or computed tomography after the pacemaker implant in suspected patients.

Discussion

Metastatic malignant cardiac tumor is observed in 2.3% of all autopsy cases in general populations, 7.1% in cancer patients, and 24% in malignant lymphoma.[1,2] In contrast, primary cardiac tumor represents 0.33% of autopsies in general populations.[3] Especially, primary cardiac lymphoma is

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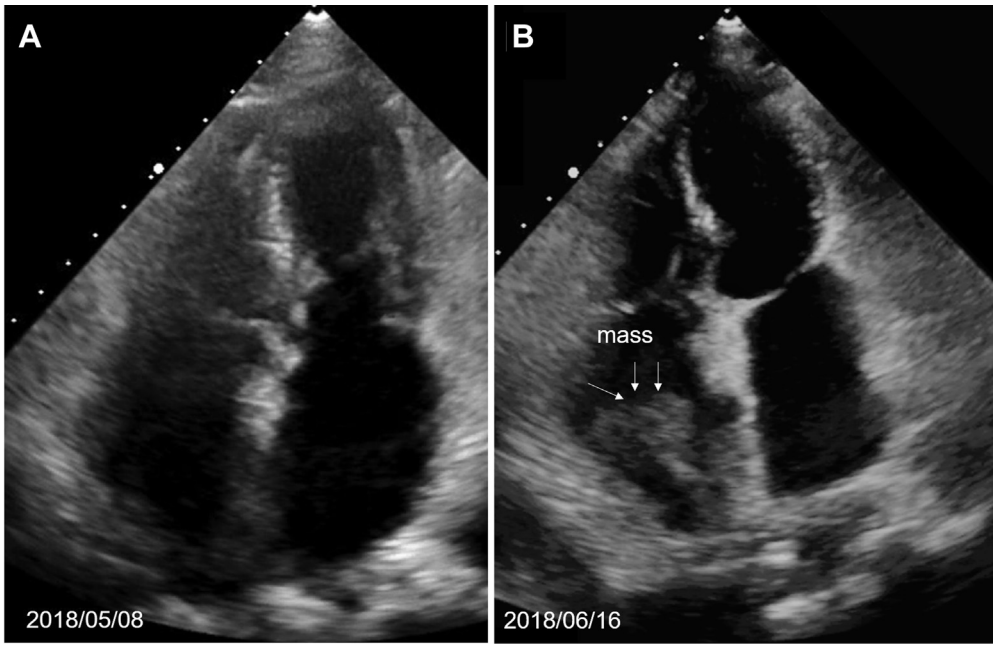

Figure 1 Echocardiogram pre- and 1 month post-implant. **A:** Baseline echocardiogram prior to the implant. No significant finding was found. **B:** Abnormal large mass in the right atrium.

extremely rare, accounting for 1.3% of primary cardiac tumor.[4] Primary cardiac lymphoma was defined as a single cardiac disease and asymptomatic extracardiac site of disease or minimal locoregional disease.[5] We diagnosed our case as primary cardiac lymphoma, based on the major lesion in the right atrium, which extended to the epicardium, SVC, adjacent right lung, and mediastinal fat with asymptomatic, minimally disseminated lesions. As malignant lymphoma often presents with rapid progression, delayed diagnosis is the major factor associated with its poor prognosis. In this case, echocardiography showed rapidly expanding low echogenicity mass over 4 weeks. Common symptoms include heart failure, pericardial effusion, atrioventricular block, SVC syndrome, and precordial pain, but there have been few cases of sick sinus syndrome in previous reports.[6–8] Our patient presented with sick sinus syndrome and developed SVC syndrome, caused by the rapidly grown B-cell malignant lymphoma.

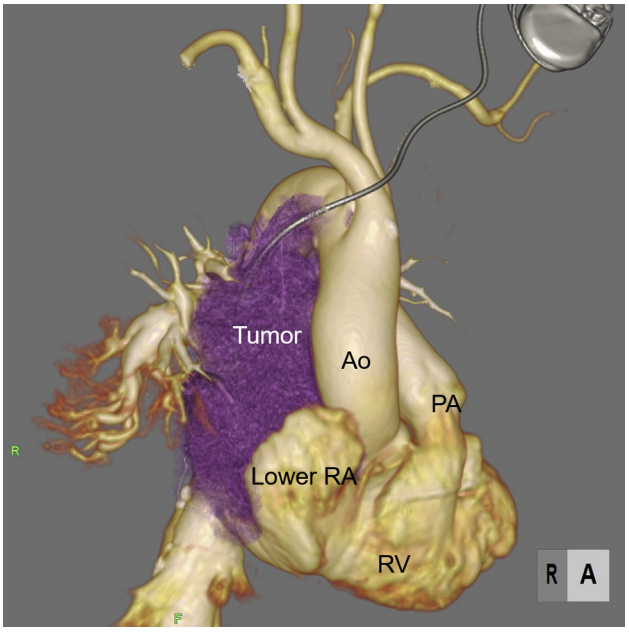

Figure 2 Three-dimensional computed tomography of the heart. Abnormal tumor in the right atrium is depicted with purple, which fills the upper part of the right atrium.

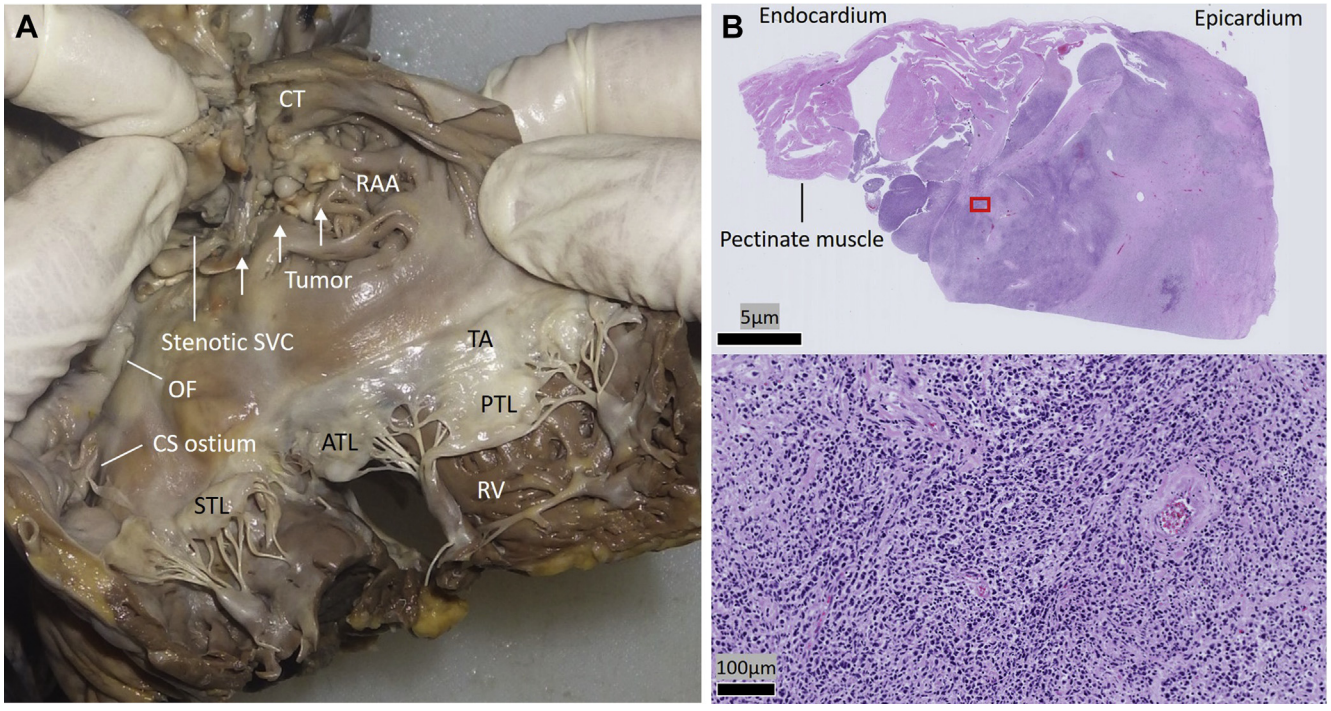

Figure 3 Gross pathology of the heart and pathologic section of sinoatrial node. **A:** Abnormal tumor (*arrow*) extends from the right atrial appendage to the crista terminalis. Right atrium inflow from superior vena cava was stenosed by tumor. ATL = anterior tricuspid leaflet; CS = coronary sinus; CT = crista terminalis; OF = oval fossa; PTL = posterior tricuspid leaflet; RAA = right atrial appendage; RV = right ventricle; STL = septal tricuspid leaflet; TA = tricuspid annulus. **B:** Sinoatrial node structure that had been completely destroyed by lymphoma and pectinate muscle without infiltration.

Conclusions

We report an elderly woman who presented with sick sinus syndrome, which was caused by rapidly growing primary cardiac B-cell lymphoma.

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