The Surgical Treatment of Primary Cardiac B-Cell Lymphoma of Clinically Unstable Patient

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The incidence rate of primary cardiac tumors is 13.8 per 1 million inhabitants per year, and 2.4% of them consist of primary cardiac lymphoma. Primary cardiac lymphoma is a fatal malignancy. Echocardiography and whole-body computed tomography are useful tools for diagnosis. Although chemotherapy has been previously described as the standard treatment for primary cardiac lymphoma, surgical treatment can be used for clinically unstable patients. Herein we report the first surgically treated case of a 57-year-old man with biatrial involvement of primary cardiac B-cell lymphoma.

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The incidence rate of primary cardiac tumors is 13.8 of 1 million inhabitants per year, and 2.4% of them consist of primary cardiac lymphoma [1]. Primary cardiac lymphoma is accepted as a type of non-Hodgkin's lymphoma located in the heart without any involvement of other parts of the body [2]. Primary cardiac lymphoma is a fatal malignancy in immunocompetent patients. Herein we report a surgically treated case of a primary cardiac B-cell lymphoma. To our knowledge, our patient is the first case to be surgically treated for primary cardiac lymphoma with biatrial involvement [2].

A 57-year-old man with superior vena cava syndrome and severe dyspnea was admitted to our hospital. Neck veins were prominent during physical examination. Electrocardiogram showed no abnormality. Transthoracic echocardiography was performed and revealed a 60 × 45 mm mass in the right atrium that extended to the left atrium. The mass was subtotally obstructing the superior vena cava. Coronary angiogram was performed to visualize concomitant coronary artery disease. A branch of circumflex artery was supplying the tumor (Fig 1A). Coronary arteries were normal. Thorax computed tomography and magnetic resonance imaging confirmed the echocardiographic findings and showed the biatrial tumor with pericardial involvement (Fig 1B, 1C). Mediastinal and cervical lymphoid tissues were normal. Exacerbation of vena cava superior syndrome led to the intubation of the patient in the intensive care unit. Emergent surgical treatment was decided on because of hemodynamic instability.

The operation was performed with median sternotomy and under cardiopulmonary bypass with aortic arterial and bicaval selective venous cannulations. Both left and right atria, interatrial septum, proximal portion of superior vena cava, and pericardium were infiltrated by the tumor (Fig 2A). Myocardial protection was maintained during the operation by using both antegrade and retrograde blood cardioplegic solution. Right atriotomy was performed. The tumor originated from the superior part of the fossa ovalis and had a lobular, solid structure and yellowish color (Fig 2B). Both right and left atria with superior vena cava were infiltrated. The superior vena cava was almost totally obstructed by the tumor. The major left atrial involvement was in the interatrial septum. In addition, the superior part of the left atrium—or more precisely, the left atrial portion anterior and superior to the right upper pulmonary vein orifice—was also infiltrated. The affected area including both atria, proximal portion of superior vena cava, and the tumor were totally removed (Fig 2C). Coronary sinus, mitral, and tricuspid valves were intact.

Left and right atrial chambers were reconstructed using bovine-pericardium patch. A 10-mm Dacron (C.R. Bard, Haverhill, PA) graft was interposed between the reconstructed right atrium and distal superior vena cava (Fig 2D). Deep hypothermia at 20°C and total circulatory arrest were used during reconstruction. Both atrial and ventricular temporary pacemaker wires were placed. The early postoperative period in the intensive care unit was uneventful except for total atrioventricular block. The temporary pacemaker was replaced with a DDD mode permanent pacemaker. The late postoperative period was uneventful. A rituximab plus cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) chemotherapy protocol was used postoperatively.

The macroscopic evaluation revealed that the tumor had almost totally obstructed the superior vena cava (Fig 3A). Pathology specimens showed infiltration of the lymphocytic cells in myocardial muscular tissue (Fig 3B). Infiltrated cells had large nuclei, medium with cytoplasm, and rough chromatin material (Fig 3C). The lymphocytic cells were confirmed to be B cells after strong cytoplasmic CD20 positivity in immunohistochemical staining (Fig 3D).

Comment

Primary cardiac lymphoma is a clinical entity accounting for 2.4% of primary cardiac malignancies [1]. Its estimated incidence is higher among immunocompromised patients [2]. Most common sites of cardiac involvement are right atrium, right ventricle, left atrium, and left ventricle. To our knowledge, our patient is the first case treated surgically for primary cardiac lymphoma with biatrial involvement [2].

Symptoms and clinical presentations of primary cardiac lymphoma are usually nonspecific and related to the

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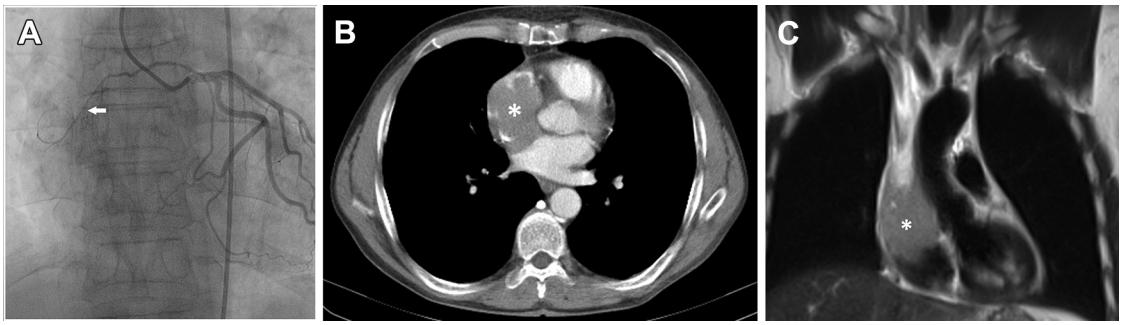


Fig 1. (A) Coronary angiogram (arrow indicates coronary artery branch of tumor). (B) Transverse section of computed tomographic view (white asterisk indicates tumor). (C) Coronal section of magnetic resonance imaging showed atrial involvement (white asterisk indicates tumor).

extension of the tumor [2]. Most common symptoms are dyspnea and palpitations. Superior vena cava syndrome is not common but may be present owing to obstruction of the superior vena cava by the tumor. Echocardiography is a useful tool for diagnosis of a cardiac tumor [3]. Both whole-body computed tomography and magnetic resonance imaging are also helpful to visualize the extension of the tumor. Most of the reported cases of biatrial primary cardiac lymphomas were B-cell lymphoma that consisted of diffuse large cell type. Although chemotherapy is the major treatment option for primary cardiac lymphoma, surgical treatment can be used for

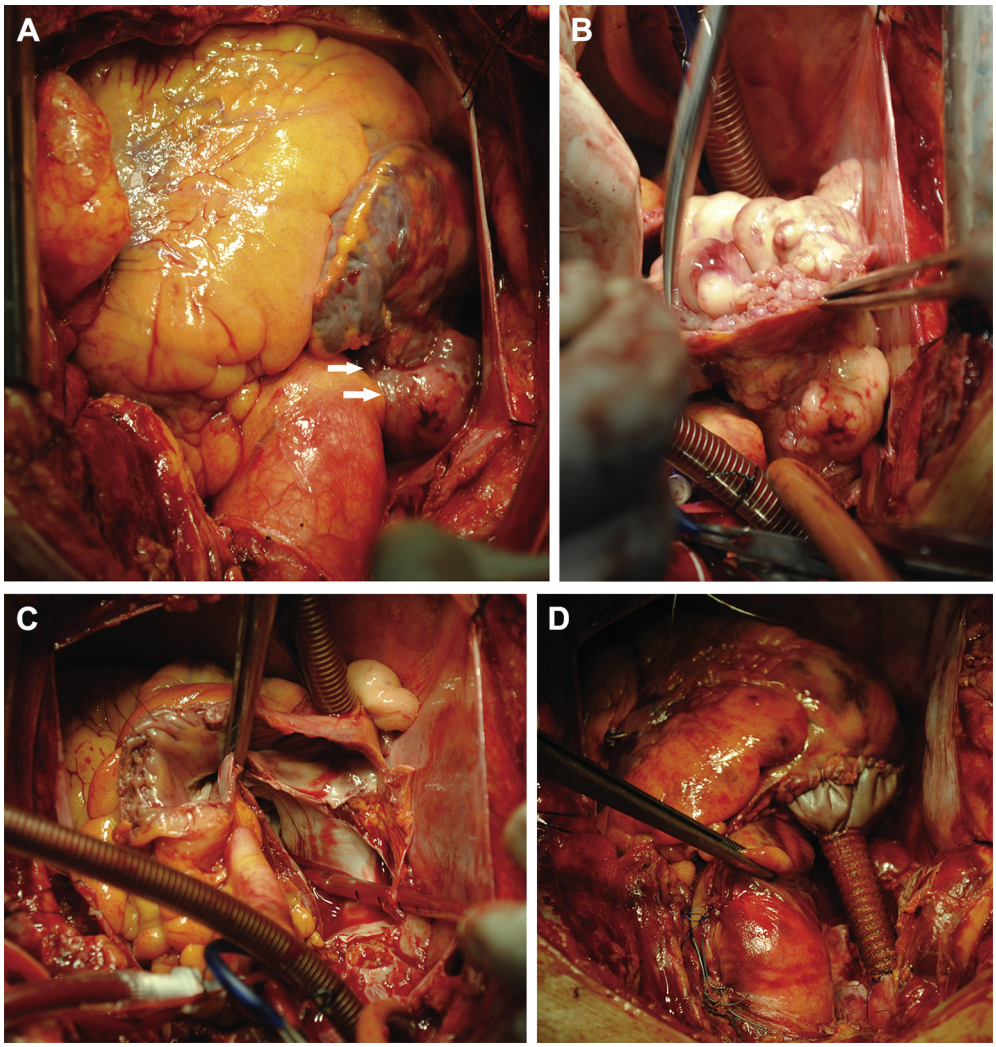


Fig 2. Intraoperative view: (A) after pericardiotomy (arrows indicate external view of tumor); (B) after right atriotomy; (C) after removal of tumor from both right and left atrium; and (D) reconstructed view of atriums and superior vena cava.

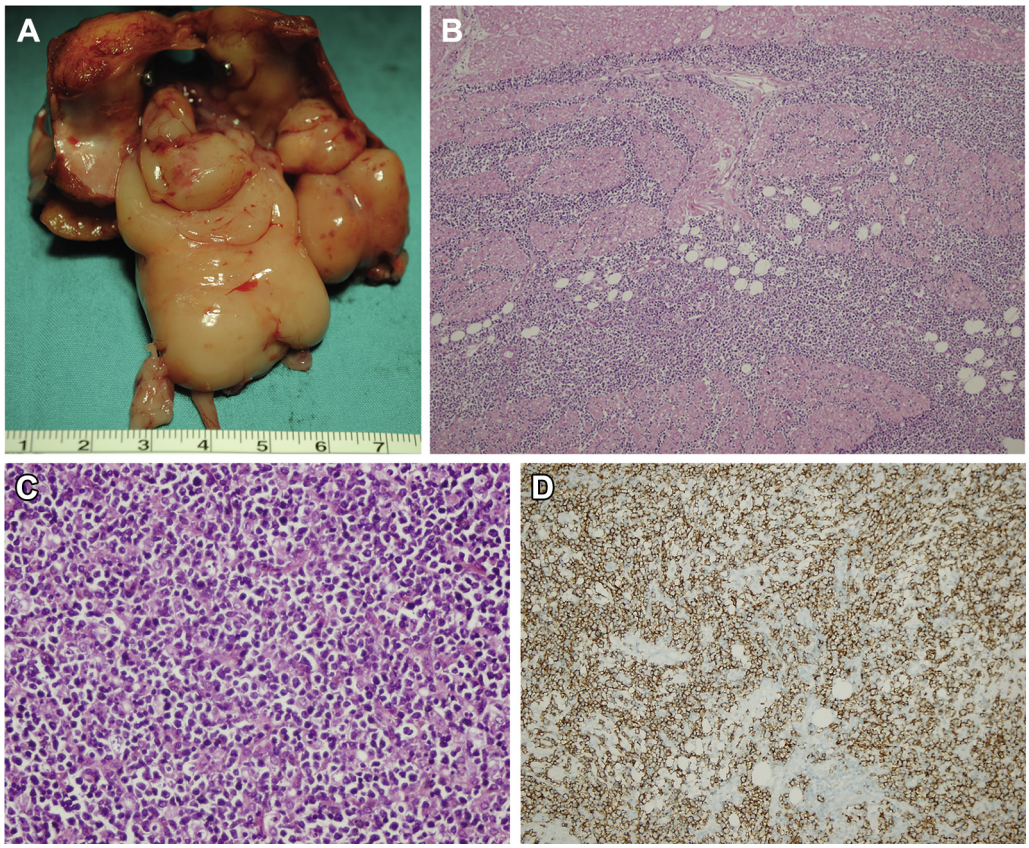


Fig 3. Pathology specimens: (A) macroscopic view of removed tumor from right atrial side (surgical dissector is passing through subtotally obstructed superior vena cava); (B) microscopic view of tumor showing infiltration of tumor in myocardial tissue (hematoxylin-eosin stain; original magnification ×100); (C) lymphocytic cells with large nucleus, medium width cytoplasm, and with rough chromatin material (hematoxylin-eosin stain; original magnification ×400); and (D) immunohistochemical stain showing strong cytoplasmic CD20 positivity (original magnification ×200).

clinically unstable patients like our patient [4, 5]. Total resection of the tumor and reconstruction of both atria can be successfully used for patients with biatrial involvement of primary cardiac lymphoma.

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