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A Rare Case of Primary Biatrial and Coronary Venous Sinus Rhabdomyosarcoma

A 26-year-old male presented with sudden onset of chest tightness, and chest pain with syncope for 1 day. He had a history of aortic valve replacement 6 years ago. Cardiac auscultation revealed a moderate diastolic rumble murmur at the apex. Electrocardiogram and laboratory examinations were unremarkable. Transthoracic echocardiography revealed a lobulated, heterogeneous, and solid mass invading biatrial and coronary venous sinus, measuring 5.1 × 4.0 cm, 3.4 × 2.5 cm, and 5.8 × 3.6 cm, respectively (Figure 1A and B), and moderate pericardial effusion. Cardiac-enhanced computed tomography showed multiple low-density soft tissue masses in both atria and coronary sinus and moderate pericardial effusion (Figure 1C and 1D). The patient underwent successful removal of the tumor under extracorporeal circulation. Intraoperative findings of extensive pericardial adhesions and moderate hemopericardium effusion show that the tumor was located in both atria with invading inferior posterior left atrial wall and coronary sinus. Postoperative histopathological examination confirmed that the tumor was an embryonal rhabdomyosarcoma (Figure 1E and F). The patient underwent postoperative adjuvant chemotherapy and there was no recurrence or metastasis after 1-year follow-up.

Primary cardiac rhabdomyosarcoma is an aggressive tumor with a dismal prognosis. The cardiac sites involved usually include the left atrium (55%), left ventricle

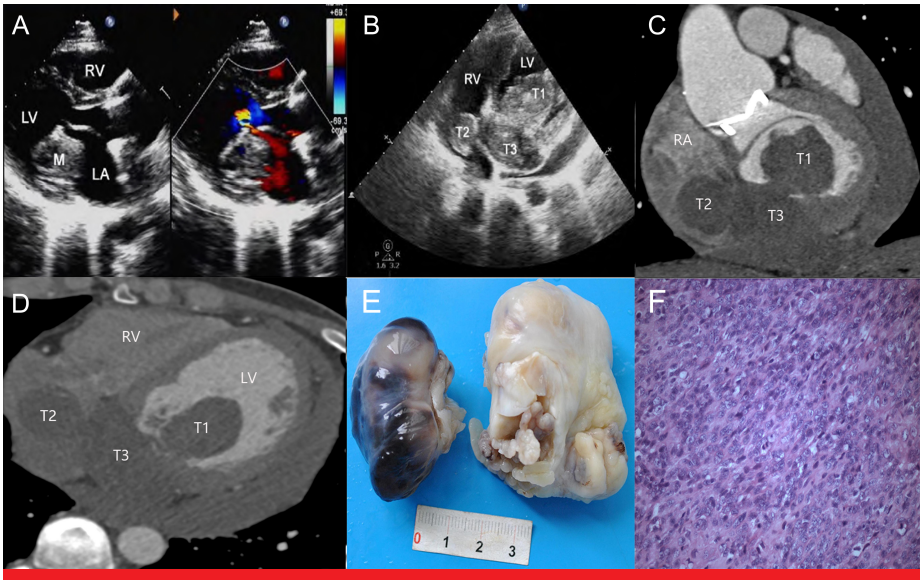

Figure 1. (A and B) Transthoracic echocardiography shows a solid mass invading the biatrial and coronary venous sinus and moderate pericardial effusion. (C and D) Cardiac-enhanced computed tomography shows multiple low-density soft tissue masses in both atria and coronary sinus, and moderate pericardial effusion. (E and F) Postoperative histopathology confirmed the tumor as an embryonal rhabdomyosarcoma. LA, left atrium; LV, left ventricle; M, mass; RA, right atrium; RV, right ventricle; T1-T3, tumor1-tumor3.

E-PAGE ORIGINAL IMAGE

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(15.7%), right ventricle (15.7%), and right atrium (13%). Biatrial rhabdomyosarcoma is an extremely rare occurrence.[1,2] The tumor is characterized by intraluminal growth and expressed invasion in the myocardium and cardiac valves.[3] The rapid progression of this tumor may cause intracardiac obstruction, arrhythmias, or pericardial effusion with tamponade as well as systemic embolization.[4] The differential diagnosis between benign cardiac tumors and sarcoma is important for determining appropriate treatment. It is difficult due to the lack of specific clinical symptoms and typical findings of preoperative examinations.[5] Multimodality images play an important essential diagnosing and assessing tumor size, shape, attachment, and mobility.[6] There is no consensus on treating cardiac rhabdomyosarcoma because the benefits of surgery and postoperative chemotherapy or radiotherapy are unclear, and the long-term prognosis is poor even after multidisciplinary treatment.[7] Although the mid-term prognosis of the tumor is poor, surgical resection should be recommended as a cornerstone of these therapeutic modalities to clarify the diagnosis, relieve symptoms, and improve short-term survival.[1]

Informed Consent: Informed consent was obtained from the patient for this study.

Declaration of Interests: The authors have no conflict of interest to declare.

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