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CASE 2. Right Ventricular Lesion As Presenting Feature of Acute Promyelocytic Leukemia

A 22-year-old white man presented to the cardiologist because of a 3-week history of palpitations. A transthoracic echocardiography (TTE) revealed a mobile heterogenous bilobate mass with a small stalk in the right ventricle adjacent to the tendon chordal attachment of the tricuspid valve (Fig 1A, arrows). The cardiac magnetic resonance imaging showed a spherical 2 cm lesion with heterogenous signal intensity on T1-weighted images, not enhancing in contrast-enhanced scans, occupying the right ventricle, and originating from the tendon chordal of the anterior papillary muscle (Fig 1B). It resembled a papillary fibroelastoma (PF). Presurgical laboratory tests revealed leukopenia, thrombocytopenia, and prolongation of clotting times. The patient was referred to the hematologist. On admission he showed platelet count of 53×10^9/L, hemoglobin level of 120 g/L, and WBC count of 1.9×10^9/L with 10% blasts. Coagulation studies were: prothrombin time, 57%; partial thromboplastin time, 1.28 seconds; fibrinogen, 1.78 g/L; and D-dimer, 40,000 ng/mL. Bone marrow aspirate showed 90% blasts with heavily granulated cytoplasm and numerous Auer rods (Fig 1C). Immunophenotyping analysis showed CD13−, CD33−, *c-KIT* positive, and *HLA-DR*–negative blast cells. Cytogenetic studies revealed the presence of chromosomal translocation t(15;17) in all examined metaphases. The diagnosis was acute promyelocytic leukemia (APL), WHO classification M3,[1] with t(15;17) and disseminated intravascular coagulation. The patient was treated with idarubicine and *all-trans* retinoic acid (ATRA) according to the Italian Group for Hematologic Diseases in Adults (GIMEMA) protocol AIDA 2000. Because of disseminated intravascular coagulation and the risk of embolization from the cardiac lesion, the patient was started on heparin to maintain partial thromboplastin time in therapeutic ranges of 1.5 to 2.5 times. Platelets were transfused for a platelet count of $> 50 \times 10^9$/L to reduce the hemorrhagic risk. A complete remission of the leukemia was obtained. The intracardiac lesion was unmodified at the end of the therapy. After complete bone marrow recovery, the patient underwent surgical removal of the cardiac mass. Histologic examination revealed amorphous eosinophilic material with reticular fibers and inflammatory cells (Fig 1D, arrows) consistent with an organized thrombus related to APL (Fig 1D, endothelial cell hyperplasia, arrow heads). He remains in complete hematologic and molecular remission. Subsequent TTE examinations showed no alterations of cardiac motion and empty heart chambers.

A PF was considered in the differential diagnosis of our patient's cardiac abnormality. PFs are rare benign primary cardiac tumors (PCTs) predominantly involving cardiac valves and affecting patients with mean age lower than myxomas or malignant PCT.[2] Most are papillary lesions less than 1 cm, connected to the valve or mural endocardium by a small stalk. Typical TTE features of PFs include a round, oval, or irregular mass with well demarcated borders and a homogenous texture; nearly half have a stalk and are mobile.[3] Magnetic resonance imaging usually shows spherical or ovoid lesions on valve leaftles or on the endocardial surface of the affected cardiac chamber, with heterogeneous signal intensity on T1-weighted images.[4] Although often asymptomatic, embolization from the lesion or attached thrombus may cause serious neurological or cardiac events. All symptomatic PF should be removed unless there are compelling contraindications, in which case anticoagulation is an acceptable but unreliable alternative.[5]

APL is a distinct subtype of acute myelogenous leukemia characterized by a balanced chromosomal translocation between chromosomes 15 and 17, young age of patients at presentation, and unique response to the ATRA treatment. Furthermore, the disease typically presents with a life-treating hemorrhagic diathesis. ATRA usually induces resolution of the bleeding symptoms. Nonetheless, recent studies report an incidence of early hemorrhagic deaths of approximately 10% to 20% in APL.[6] Thromboses are a rare complication of APL and are almost invariably associated with ATRA treatment. A review of the literature disclosed 10 cases of arterial (seven cases),[7,8] venous (one case)[9] or intracardiac (two cases)[10] thrombosis in the course of differentiation therapy with ATRA. Some authors have referred the procoagulant tendency to the overexpression of cellular surface adhesion molecules induced by ATRA.[7] Only two cases so far have been reported of arterial occlusions as presenting features of APL, independently from ATRA administration. In the first case, the patient presented with an acute myocardial infarction. A fibrin-platelet

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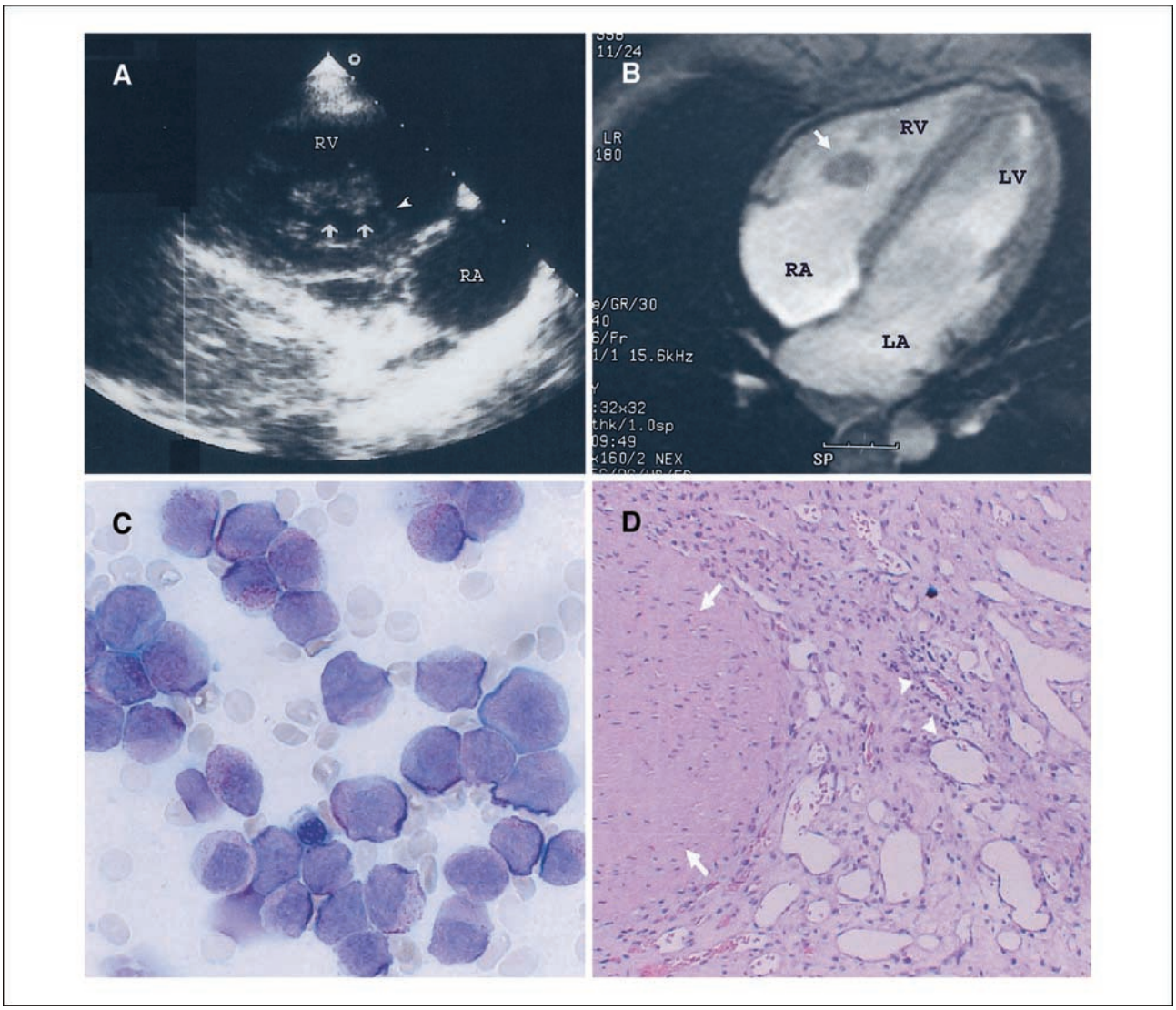

Fig 1.

thrombus was demonstrated postmortem in the anterior descending branch of the left coronary artery.[11] In the second case, the patient presented with pain and bluish discoloration of his left great toe. An aortogram revealed a clot in the left external iliac artery. The thrombus was surgically removed but was not sent for histologic examination. TTE showed a sessile mass in the right ventricle not attached to the tricuspid valve, the possible focus of the embolus. ATRA plus chemotherapy and heparin resulted in safe and efficacious management and resolution of both thrombosis and leukemia. Thrombi were attributed on a clinical basis to leukemic cell thrombosis.[12] Here we report the second case of intraventricular thrombosis as a presenting feature of APL. Furthermore, the radiologic appearance of the thrombus was elusive and suggestive of a PCT. Histologic examination disclosed the nature of the cardiac mass. Thrombotic events may occur in APL independently from ATRA treatment. Heparin may be safe and useful in the management of coagulation abnormalities in the course of APL.

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Authors' Disclosures of Potential Conflicts of Interest

The authors indicated no potential conflicts of interest.

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CASE 3. Primary Cardiac Lymphoma

A 41-year-old female developed acute onset of right shoulder pain followed by anterior pleuritic chest pain on the morning of admission. She had had a similar episode approximately 1 month prior, which resolved spontaneously and was not as intense in nature. Her past medical history was only significant for well-controlled asthma and endometriosis. Labs were notable for a normocytic anemia. Chest x-ray showed an indistinct superior right heart border and right hilum. Cardiac magnetic resonance image showed a large mass filling the majority of the right atrium (Fig 1, arrow). This measured 3.5 × 6 × 5 cm and appeared fixed to the wall of the right atrium along the right anterior aspect. The mass was separate from the intra-atrial septum and did not involve the right ventricle. It extended superiorly, involving the junction of the superior vena cava and the right atrium. A transesophageal echocardiagram demonstrated a large, sessile right atrial mass with a multilobular appearance, with a broad base attached to the right atrial free wall and the wall of the superior vena cava. Ventilation perfusion scan showed no evidence of pulmonary emboli. The patient underwent surgical resection of the atrial mass and reconstruction of the right atrium with a patch graft. Pathology demonstrated an 8 × 6.8 × 3 cm diffuse large B-cell lymphoma involving the full thickness of the right atrium (Fig 2A and B). Immunoperoxidase stains were positive for leukocyte common antigen (CD45) and CD20/L26 B-cell marker (Fig 2C). Postoperatively, a computed tomography scan of the abdomen was notable for a complex left uterine mass, measuring 7.8 × 5.7 × 6.1 cm. Biopsy revealed only RBCs, possibly secondary to endometriosis. Positron emission tomography scan showed no residual disease in the right atrium and no areas of increased uptake.

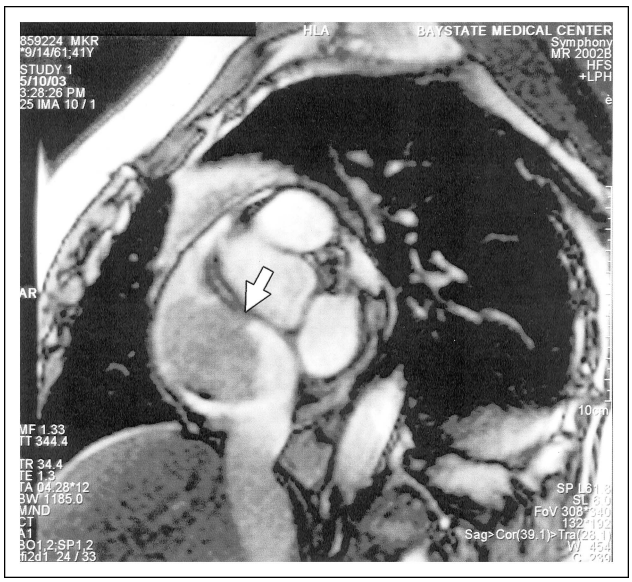

Fig 1.

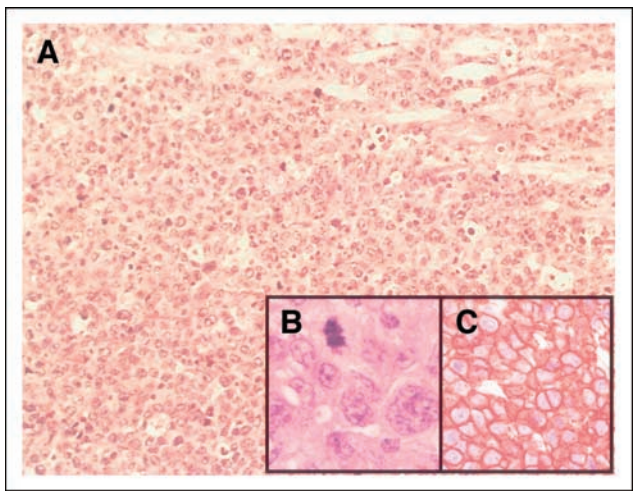

Fig 2.

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