Case Reports

Coronary Embolization from a Left Atrial Myxoma

Containing Malignant Lymphoma Cells

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Systemic embolization from a primary cardiac tumor is a relatively frequent presentation. However, an acute myocardial infarction due to coronary embolization is rarely seen. We offer an unusual case of a 50-year-old man who presented with severe angina and was diagnosed with an inferolateral ST-segment-elevation myocardial infarction. Aside from otherwise healthy coronary arteries, his coronary angiogram revealed an acute occlusion of the first obtuse marginal branch, which was treated with balloon angioplasty. Because no residual plaque or dissection was found after the angioplasty, an embolic source was suspected. An echocardiogram then revealed a large mobile left atrial myxoma prolapsing into the left ventricle, so the patient underwent minimally invasive resection. Detailed pathologic examination of the myxoma revealed a concomitant high-grade B-cell lymphoma. ***(Tex Heart Inst J 2015;42(6):565-8)***

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The pathophysiology of an ST-segment-elevation myocardial infarction involves, in most cases, the rupture of an atherosclerotic plaque with superimposed thrombus formation.[1] Cardiac tumors are among the possible embolic sources of coronary embolization, and can present with an acute myocardial infarction.[2] Embolic causes must be suspected in patients who have otherwise healthy coronary arteries. We present the case of a 50-year-old man who presented with myocardial infarction due to an acute occlusion of the first obtuse marginal branch—most likely by a coronary embolus from a large mobile left atrial myxoma. In addition, a concomitant malignant lymphoma was found within the tumor.

Case Report

A 50-year-old white man presented at the emergency department at our institution with a 1-hour history of severe retrosternal pain, dyspnea, and diaphoresis. His medical history was noteworthy for hypertension and dyslipidemia, both of which had been treated with diet and exercise, and there was no family history of coronary artery disease. He drank alcohol occasionally, and had no history of smoking or illicit-drug use. He reported no symptoms before the presentation, such as dyspnea on exertion, fever, night sweats, or weight loss. On presentation, his blood pressure was 148/92 mmHg; pulse, 102 beats/min; respiratory rate, 22 breaths/min; and oxygen saturation, 97%. The patient was anxious and diaphoretic, but the results of his cardiopulmonary examination were generally within normal limits, without murmurs or signs of congestive heart failure. The initial electrocardiogram (Fig. 1) showed inferolateral ST-segment elevation with reciprocal changes in the anteroseptal leads, so the patient was taken for emergency coronary angiography. His angiogram revealed an acute occlusion of the first obtuse marginal branch in its mid segment (Fig. 2A), with otherwise normal coronary arteries. After an attempted aspiration recovered no débris, balloon angioplasty was performed, and a repeat angiogram showed no significant residual plaque or dissection at the level of the original occlusion; but it did reveal distal branch embolization (Fig. 2B). Therefore, an embolic source was suspected. The patient was admitted to the coronary care unit for further evaluation and conventional postinfarction management. His echocardiogram showed normal left ventricular (LV) function with no wall-motion abnormalities or valvular disease of substance; however, it revealed a large polypoid left atrial mass attached to the interatrial septum. Notably mobile and prolapsing into the LV, it was suspect for myxoma (Figs. 2C and D). In light of these findings, the patient underwent minimally invasive resection

of a 6.5 × 3-cm left atrial myxoma (Fig. 3A) via a right minithoracotomy. Pathologic analysis confirmed myxoma (Fig. 3B), but at the tumor's edge there were also atypical lymphoid cells (Fig. 3C–E) with high mitotic activity—these stained positive for CD3, CD20, Ki67, and PAX5, consistent with a concomitant high-grade B-cell lymphoma. Further diagnostic testing failed to detect additional areas of malignant involvement or an immunodeficiency. The patient had an uneventful postoperative recovery and was discharged from the hospital after 9 days, with oncologic follow-up for further management.

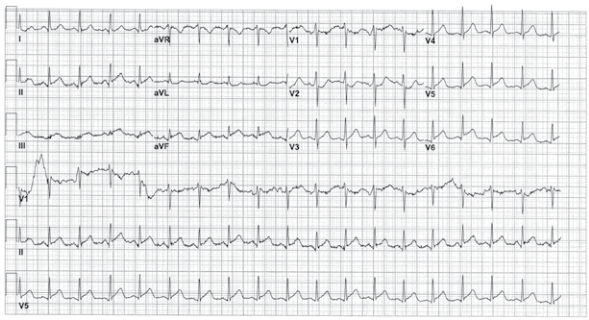

Fig. 1 *Initial electrocardiogram shows inferolateral ST-segment elevation with reciprocal changes in the anteroseptal leads.*

Discussion

Primary cardiac tumors are rare, with a calculated prevalence of 0.3%, according to autopsy studies.[3] Of those tumors, 75% are benign. Myxomas account for 50% of primary cardiac tumors; most are sporadic, and are found more commonly in women.[2,4] The most frequent

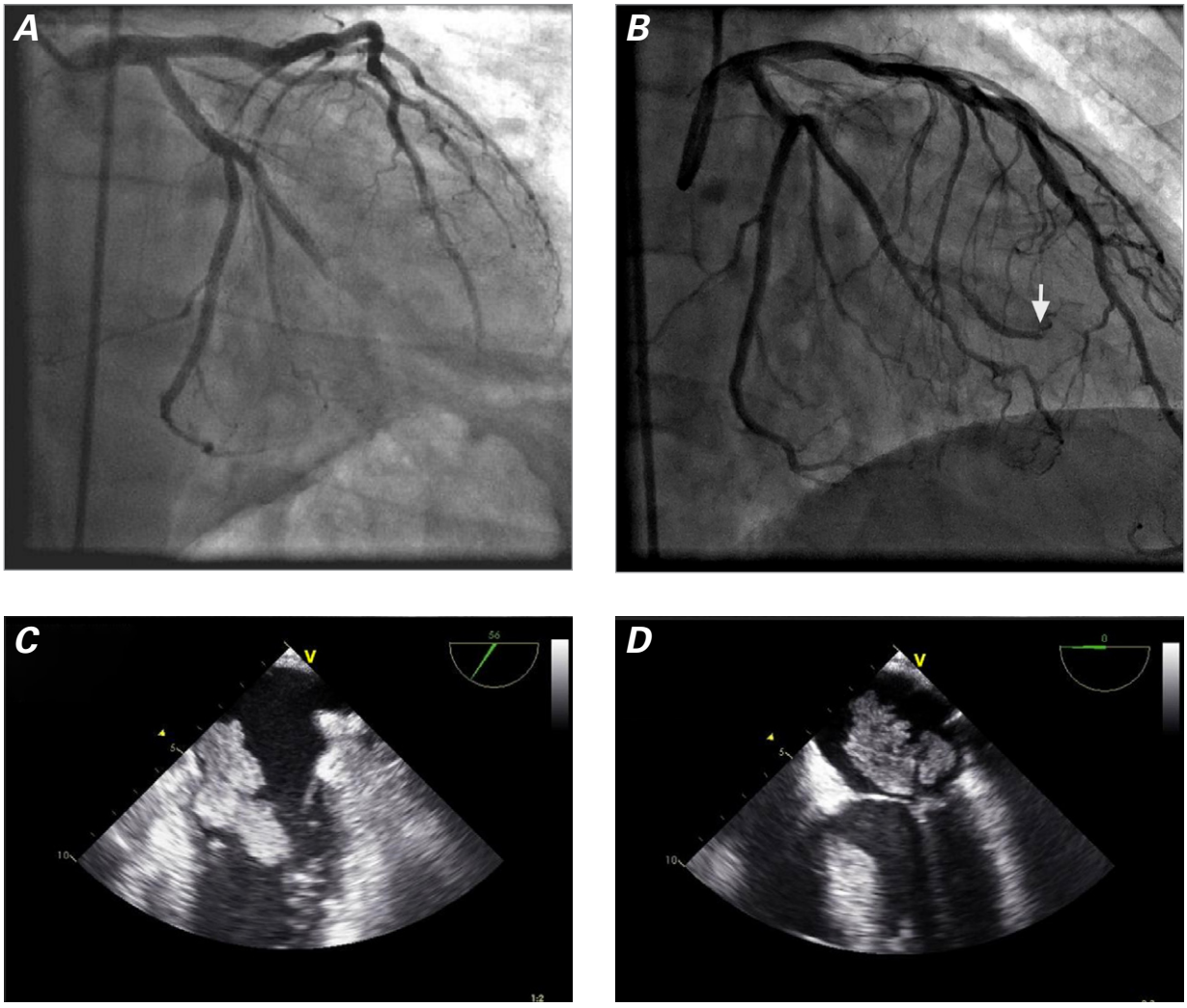

Fig. 2 ***A****) Coronary angiogram shows acute occlusion of the first obtuse marginal branch in its mid segment.* ***B****) Repeat coronary angiogram after aspiration and balloon angioplasty shows no substantial residual plaque or dissection at the level of the original occlusion, but it does reveal distal branch embolization (arrow). The echocardiogram shows* ***C****) a left atrial mass attached to the interatrial septum and prolapsing into the left ventricle, and* ***D****) a large polypoid left atrial mass.*

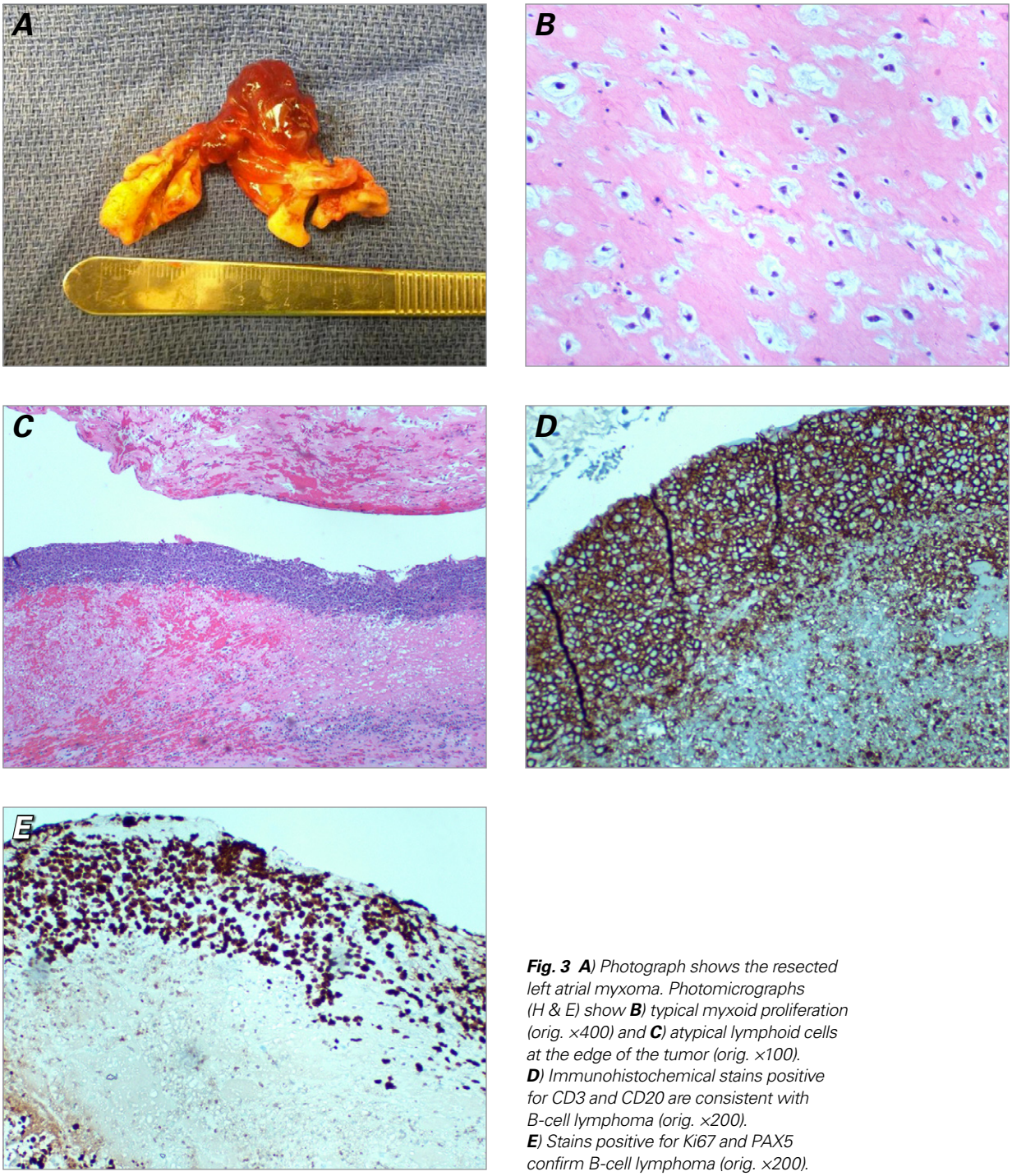

Fig. 3 ***A****) Photograph shows the resected left atrial myxoma. Photomicrographs (H & E) show* ***B****) typical myxoid proliferation (orig. ×400) and* ***C****) atypical lymphoid cells at the edge of the tumor (orig. ×100).* ***D****) Immunohistochemical stains positive for CD3 and CD20 are consistent with B-cell lymphoma (orig. ×200).* ***E****) Stains positive for Ki67 and PAX5 confirm B-cell lymphoma (orig. ×200).*

location (75%–85%) is the left atrium, where they typically arise from the interatrial septum. Ninety percent of myxomas are pedunculated, with 2 morphologic patterns: round or polypoid.[2,4] There are 3 distinct groups of clinical manifestations. Constitutional symptoms, which have been attributed to tumoral cytokine production, include fatigue, fever, weight loss, and myalgias. Atrioventricular valve obstruction can present with symptoms of mitral or tricuspid valve regurgitation or stenosis, and with left- or right-sided heart failure.[2,4] Finally, in up to 65% of left-sided myxomas, systemic embolization causes arterial occlusion and organ ischemia or infarction, which typically manifests itself as ischemic stroke or renal infarction.[2,4] However, the incidence of coronary embolization is only 0.06%, which might be explained by the perpendicular orientation of the coronary ostia to the aortic flow and to their consequent protection from the aortic leaflets

during systole.[2,4-7] The right coronary artery is most often affected; however, embolization to the left anterior descending and left circumflex coronary arteries has also been reported. Systemic embolization is more frequently associated with polypoid tumors that have an irregular and friable surface, and with tumors that prolapse into the ventricle.[2,4-7]

Malignant neoplasms account for 25% of primary cardiac tumors. Primary cardiac lymphomas are extremely rare (<2% of primary cardiac tumors and <0.5% of lymphomas) in immunocompetent individuals, are usually invasive, and carry a very poor prognosis.[8,9] Petrich and colleagues[8] combined the data from all available cases of primary cardiac lymphoma in the literature up to 2011 and reported that the most common presenting symptoms were heart failure and constitutional symptoms. In their series, only 6% of the patients had an embolic event, and there were no cases of coronary embolization.[8]

The treatment of primary cardiac lymphomas includes standard chemotherapy for B-cell lymphomas, which typically includes cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) with or without rituximab, in combination with surgical resection, radiation therapy, or both.[8,10] The reported median survival period ranges from 1 to 22 months. Apart from the immunologic status of the individual, the response to therapy and the survival of patients with primary cardiac lymphomas appears to relate to extracardiac extension of the tumor, to LV involvement, and to arrhythmias.[8]

To our knowledge, only 3 cases of a localized malignant cardiac lymphoma arising from a myxoma have been reported.[5-7] In all 3 cases, the patient presented with symptoms attributable to a large left atrial myxoma, including mitral valve obstruction with heart failure or ischemic stroke, and the more troublesome aspect of this dual pathologic condition—a concomitant malignant lymphoma—was found incidentally. Unlike the prior reported cases, and despite the large tumor size, our patient did not have typical constitutional symptoms or heart failure secondary to atrioventricular valve obstruction; instead, he had an atypical presentation with an acute myocardial infarction secondary to coronary embolization.

Summary

Here, we present an unusual case of a large left atrial myxoma and a concomitant malignant B-cell lymphoma presenting with an ST-segment-elevation myocardial infarction most likely caused by coronary embolization. This case highlights not only the importance of suspecting an embolic source of acute myocardial infarction (especially when the coronary arteries are otherwise normal), but also the extremely rare occurrence of this dual-tumor pathologic disorder.

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