Left Atrial Thrombus Mimicking a Myxoma in a Patient with Systemic Lupus Erythematosus

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Atrial masses are uncommon, and mainly consist of tumors, vegetations and thrombi. Despite the availability of several imaging modalities, it may still be difficult to distinguish between them. Atrial thrombi seen in patients with systemic lupus erythematosus (SLE) usually occur in the setting of anti-phospholipid antibody syndrome, which confers a hypercoagulable state. We report a case of left atrial thrombus mimicking a myxoma in a patient with SLE but negative anti-phospholipid antibodies.

The differential diagnosis of an atrial mass includes tumor, vegetation and thrombus. Atrial myxoma accounts for up to half the cases of all atrial tumors; the remaining ones include cardiac rhabdomyoma, cardiac papillary fibroelastoma, cardiac fibroma, hemangioma, cardiac lipoma, and cardiac paraganglioma. Vegetations can mimic an atrial mass, but they are typically attached to the valve leaflet. Intra-atrial thrombi can sometimes present as atrial masses, with a predilection for the right atrium. They may or may not be associated with a hypercoagulable state. Numerous case reports have described the occurrence of atrial thrombi in patients with systemic lupus erythematosus (SLE). However, most of these cases also have positive anti-phospholipid antibodies. We describe a unique case of left atrial thrombus in a patient with SLE but negative anti-phospholipid antibodies.

Case presentation

Our patient was a 33-year-old Hispanic female, with a seven year history of SLE, who was being treated with multiple regimens, including steroids, cyclophosphamide, rituximab, and mycophenolate. This was complicated by end-stage renal disease secondary to SLE, treated by hemodialysis for the last two years. She presented to the emergency room with a two-day history of fever, chills and cough. Blood cultures were drawn and she was started on intravenous antibiotics. The chest radiograph depicted right sided pleural effusion. Computed tomography confirmed these findings, but also revealed an incidental finding of a left atrial mass (Figure 1) that was also seen on the transthoracic contrast echocardiogram. Subsequently, a transesophageal echocardiogram revealed a low-density, echogenic, wide-based peduncular mass attached to the atrial aspect of the posterior mitral valve annulus near the A1-P1 scallops (Figure 2). It was homogeneous and non-mobile, with no evidence of calcification or central necrosis. There was no color Doppler signal noted in the mass, making it less likely to be a vascular rich structure. The mass was not found to obstruct the mitral inflow on multiple views. For better characterization of this left atrial mass, cardiac magnetic resonance imaging (CMR) was performed, and showed a 2.4 × 1.8 cm mass attached to the mitral annulus near the A1-P1 scallops (Figure 3). The mass was well-defined with smooth
borders. Unfortunately, gadolinium contrast could not be administered because of the patient’s renal failure. Given these findings, an atrial myxoma was the most likely diagnosis, and the patient underwent successful resection.

Intraoperatively, a round, firm, well-defined left atrial mass adjacent to the mitral valve annulus was seen. The mitral valve annulus showed extensive calcification. Pathologic examination revealed an organized thrombus with extensive hemorrhage, hemosiderin and hemosiderin-laden macrophages. The specimen was negative for calretinin and S100. This confirmed the diagnosis of left atrial thrombus. A complete hypercoagulable work up, including international normalized ratio (INR), prothrombin time, partial thromboplastin time, lupus anticoagulant panel, anticardiolipin antibody, protein C, protein S, factor II prothrombin (G20210A) mutation, factor V Leiden, and antithrombin III revealed no abnormalities. The patient did not have a history of atrial fibrillation. Postoperatively, she was started on warfarin and discharged. A six-month follow-up echocardiogram was normal.

Discussion

Atrial masses are a rare occurrence, and primarily consist of tumors, vegetations and thrombi.\textsuperscript{1} Echocardiography can be used for characterization of the mass by morphologic shape, appearance, site of attachment, type of margin, and presence or absence in the left atrial appendage.\textsuperscript{4} Transesophageal echocardiography is superior to transthoracic echocardiography.\textsuperscript{3-7}

Atrial myxomas are by far the most common benign cardiac primary tumors. On the echocardiogram, they usually appear as a mobile mass attached to the endocardial surface by a stalk, which typically arises from the fossa ovalis. If the stalk is not visualized, further imaging with CMR is indicated, which is the test of choice for the evaluation of atrial masses.\textsuperscript{4,8} Once the diagnosis of atrial myxoma is made, surgery is indicated because of the risk of embolic complications. Rarely, coronary myxemboli can lead to myocardial infarction.\textsuperscript{8}

Generally, atrial thrombi can present with thoracic pain, heart murmur, congestive heart failure symptoms,\textsuperscript{3} or may even be asymptomatic—as in our patient, where it was diagnosed as an incidental finding. Right atrial thrombi may lead to pulmonary embolism, whereas left atrial thrombi may lead to stroke or peripheral embolic phenomena. Atrial thrombi are more commonly seen on the right side.\textsuperscript{3} As far as left atrial thrombi are concerned, they are classically found in the atrial appendage, but can also be found in the body of the left atrium,\textsuperscript{9} as in our patient. They may sometimes have a stalk, which can lead to their being diagnosed as myxomas, as in our patient, and subsequently unnecessary surgical resection.\textsuperscript{10,11} More rarely, the thrombus may even be entrapped in a patent foramen ovale, again mimicking a myxoma.\textsuperscript{12} Any atrial mass is more likely to be a thrombus in the presence of atrial fibrillation, enlarged atrial chamber, stenotic or prosthetic mitral and tricuspid valves, low cardiac output state or spontaneous atrial contrast echoes.\textsuperscript{9,13}

The treatment of atrial masses typically includes surgical resection. In cases of atrial thrombi, anticoagulation therapy along with a hypercoagulable workup is recommended.\textsuperscript{14-15} In our patient, a complete hypercoagulable workup, including INR, prothrombin time, partial thromboplastin time, lupus anticoagulant panel, anticardiolipin antibody, protein C, protein S, factor II prothrombin (G20210A) mutation, factor V Leiden, and antithrombin III, was negative. The patient did not have a known history of atrial fibrillation at any point. Numerous case reports describing atrial thrombi and SLE patients have been published, but they are primarily in reference to antiphospholipid antibody syndrome.\textsuperscript{1,3,15-17} However, as reported previously,\textsuperscript{2} and in our patient, it can also occur in SLE patients in the absence of antiphospholipid antibody syndrome, raising concerns that a hypercoagulable state may still be present with a negative lupus anticoagulant panel.

Figure 1. Computed tomography with intravenous contrast of the thorax, demonstrating a well-circumscribed hypodense lesion measuring 2.4 cm in diameter within the left atrium.
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References


Figure 2. Transesophageal echocardiogram depicting a wide-based peduncular mass attached to the anterior aspect of the posterior mitral valve annulus.

Figure 3. Cardiac magnetic resonance imaging showing a 2.4 × 1.8 cm mass attached to the mitral annulus near the A1-P1 scallops.