A 74-year-old woman was at the emergency department for acute chest pain, dyspnea and severe transient hypotension. History was arterial hypertension and external electrical cardioversion (EEC) for persistent atrial fibrillation (AF) 8 days before admission. At that time echocardiography was normal. The patient underwent coronary angiography with no evidence of significant coronary arteries disease. At echocardiography a large multi-loculated mass occupying most of the left atrial space and obstructing left ventricular inflow was evident. There was mild pericardial effusion. The patient was operated and a large thrombus totally encompassed in the left atrial wall was removed. Initial tearing into the pericardial space was revealed. Post-surgical follow-up was uneventful and at 3–6 months normalization of the atrial cavity with blending of atrial endocardium and epicardium was demonstrated. No apparent etiological factor was found. We have provided evidence of the possible rapid formation of a large intramural atrial hematoma. Spontaneous atrial wall dissection should be considered in the differential diagnosis of chest pain.

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1. Introduction

Atrial wall intramural hematoma is a rare occurrence in clinical practice. There are very few reports that relate it mostly to catheter or surgical cardiac procedures (e.g. transcatheter arrhythmia ablation, bypass or other cardiac surgery) [1, 2]. Intramural bleeding in the atrial wall has also been described in association with cardiac amyloidosis [3], mitral calcification [4], myocardial infarction [5] and blunt chest trauma [6]. Some cases have also been reported without a clear identifiable etiology [7–9]. The variability of the location, the etiology and the dimensions of the mass may account for different modalities of presentation.

We describe the case of a left atrial intramural hematoma that occurred abruptly, mimicking myocardial ischemia and causing pulmonary congestion and transient hemodynamic collapse.

2. Case report

A 74-year-old lady, with no prior thoracic surgery or chest trauma, presented at night to accident and emergency with a history of unrelenting chest pain and dyspnea, that started a few hours before. Past medical history was arterial hypertension and recurrent episodes of atrial fibrillation (AF). An external electrical cardioversion (EEC) for persistent AF was performed 8 days before admission at another institution. The patient was, therefore, under warfarin therapy with an INR of 2.3. At presentation, an ECG showed moderate sinus tachycardia, normal conduction and a slight ST elevation in the anterior leads. Troponin T was mildly positive. X-ray showed signs of pulmonary congestion with a mild increase of the cardiac silhouette. The patient had a single episode of marked hypotension that remitted spontaneously. A coronary angiography was performed and showed only minor atherosclerotic irregularities. The patient was then admitted to the intensive coronary care unit. A trans-thoracic echocardiogram revealed the presence of a mass occupying most of the left atrial cavity; mitral mean gradient was 13 mmHg; there was moderate tricuspid regurgitation with severe pulmonary hypertension; left ventricular dimensions and ejection fraction were normal. Trans-thoracic examination yielded low-quality images, however, due to the robust chest of the patient and no further morphological definition of the mass was possible at this stage. The patient was treated with diuretics for heart failure and no episodes of hypotension occurred in the follow-up. Pain control was obtained only with morphine.

On the following morning a trans-esophageal echocardiogram (TOE) was performed (Fig. 1b) and revealed a large (6.4×5.2 cm) multi-loculated mass occupying most of the left atrial space and obstructing left ventricular inflow. There was no significant obstruction to left venous return.
Fig. 1. (a) TOE performed 8 days before admission showing only a moderately dilated left atrium. (b) TOE at the time of presentation, showing a large (6 cm) multi-loculated mass occupying most of the left atrial space and obstructing left ventricular inflow. (c) Post-surgery TOE showing a totally free left ventricular inflow, the dissected atrial wall with the endothelial layer appearing as a ‘free-floating’ membrane within the atrial lumen. (d) 3 months follow-up trans-thoracic 4-chamber image showing normalization of the atrial cavity.

The mass filled almost all of the atrial space and was delimited by a well-defined wall. Its content was partially fluid with thrombus-like organization. There was no signal over it, at color-Doppler. An acceptable blood flow was maintained through a narrow anterior space. Defining the intra- vs. extra-cardiac origin of the mass was debated among cardiologists and cardiac surgeons at that time, but the ultimate decision was to operate immediately.

Worth mentioning is the fact that the TOE performed, in view of EEC, only 8 days before admission, was normal, except for mild left atrial dilatation (Fig. 1a).

At surgery (Fig. 2a), pericardial opening revealed the presence of 200 ml of blood and the right atrial wall appeared bluish above the inferior vena cava. Left longitudinal atriotomy in front of the right pulmonary veins showed atrial wall thickening containing thrombus (Fig. 2b) and extending to the septum and to the free atrial wall, toward the left pulmonary veins. The right atrium was entered and the atrial septum divided in order to completely remove the mass encompassed in the atrial wall. It was determined that the endocardial wall of the intramural cavity was fully intact; two minimal tears were found on the epicardial layer close to the left pulmonary veins, explaining the presence of blood in the pericardial sac. No evidence of active bleeding was found. Surgical incisions were closed routinely and the dissected epicardial and endocardial layers were not glued. At postoperative TOE (Fig. 1c) the dissected atrial wall appeared like an empty chamber delimited by atrial epicardium and endocardium; the endothelial layer appeared like a free-floating membrane within the atrial lumen.

Follow-up was uneventful and the patient recovered to active life. At 3 months post-surgery, a trans-thoracic echocardiogram revealed normalization of the left atrium with spontaneous blending of the epicardial and endocardial layers (Fig. 1d), thus confirming that the appropriate surgical approach had been chosen. Histology of the mass consisted of a recent thrombus.

3. Discussion

The true incidence of intramural atrial hematomas is unknown, as its presentation with premature death cannot be excluded. We have provided for the first time clinical and iconographic data demonstrating the possible rapid formation of such a life-threatening voluminous mass. Intramural atrial hematomas have already been described, but none of the previous reports provided documented information about their time of development.

The etiology of our case remains undetermined. We do not think EEC a likely cause, because 1) with very large numbers of EECs being performed for AF around the world, there is no description, to our knowledge, of the occurrence of intramural atrial hematomas in the immediate follow-up. 2) Although endothelial dysfunction following therapeutic direct-current shock to the chest has been described, no report of microvascular damage is available. In our case, at histopathology (Fig. 2c) no evidence of active inflammation, myxomatous material, tumor, echynococcus cysts, or localized vascular malformation was present. The specimen was negative for mycobacterial, fungal or bacterial culture. It consisted of a large recent thrombus (Fig. 2b)
and adjacent myocardial wall: the atrial muscle fibers surrounding the clot showed a haphazardly distribution and nuclear enlargement. Red Congo staining for amyloid of the atrial tissue did not show apple-green birefringence (Fig. 2c).

Although more information is needed, we speculate that spontaneous intramural atrial hematomas may represent an ‘idiopathic’ clinical entity. Warfarin and aging may act as favoring agents.

In conclusion, this case confirms the need for considering this disease in the differential diagnosis of prolonged persistent chest discomfort.

References