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An unexpected and tumultuous diagnosis of a left atrial mass

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Abstract

This case presents a curious diagnosis in a young male presenting with chest pain. The first imaging tests suggested the presence of a hypovascular left atrial tumor. After cardiac magnetic resonance and the exclusion of extra-cardiac lesions, sarcoma emerged as the main diagnostic hypothesis. Unexpectedly, the histopathological study revealed the absence of malignancy, identifiable inflammatory and cardiac muscle tissue, and fibrosis. This pattern was compatible with inflammatory myofibroblastic tumor diagnosis, a rare entity with uncertain behavior but a known risk of recurrence and/or potentially fatal complications.

This is a unique case of an unexpected finding at presentation, as well as a complex diagnostic work-up and a surprisingly unusual final diagnosis. It also highlights the increasing importance of the multimodality imaging approach, as well as the critical role of multidisciplinary discussion in optimizing patient management in such complex cases.

Key words: cardiac imaging, cardiac masses, inflammatory myofibroblastic tumor, multimodality imaging.

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Case Report

Patient presentation

A 40-year-old healthy male presented to the emergency department (ED) with a 3 day-history of chest pain. The first episode had onset while resting after dinner. It was a constrictive, intense, and apparently prolonged episode with spontaneous resolution during the night. On the following morning, the patient presented with pleuritic chest pain. Intensification of these complaints motivated the ED visit. He had no cardiovascular risk factors, and his past medical and family history were unremarkable. The patient was not on any regular medication. At ED admission, he presented with residual pleuritic chest pain complaints and hemodynamic stability. Physical examination was normal, and the electrocardiogram (ECG) showed diffuse and nonspecific repolarization abnormalities.

Initial workup

Blood samples were collected, and serial high-sensitivity cardiac troponin testing was performed, showing elevated inflammatory markers (leukocytosis and high C-reactive protein value), as well as elevated troponin (maximum reported value of 300 ng/L; normal values <34 ng/L). At this point, two main differential diagnoses (DD) were being considered – myocardial infarction (MI) *vs.* myopericarditis. A complementary diagnostic work-up was performed to clarify the chest pain etiology. ECG showed no dynamic changes. A transthoracic echocardiogram (TTE) was performed (Figure 1a and b) and revealed preserved biventricular ejection fraction, absence of changes in segmental contractility, small circumferential pericardial effusion, and, unexpectedly, a left atrium (LA) mass. The mass had noteworthy dimensions, though no auriculoventricular flow disturbances were present. Regarding its appearance, it was a heterogeneous and infiltrative mass. A transesophageal echocardiogram (TEE) was the next step, aiming to confirm and better characterize the finding. TEE (Figure 1c and d) corroborated TTE data, showing a vacuolized, polycystic mass infiltrating the LA roof, inferior and posterior walls, as well as the interauricular septum. Thoracic computed tomography angiography (CTA) was also requested to help clarify mass etiology, revealing its hypovascular nature (Figure 1e and f). Altogether, imaging findings pointed to a hypovascular neoplastic mass as the most reasonable diagnostic hypothesis. The patient was admitted to the cardiology unit with the diagnosis of myopericarditis in the context of a cardiac neoplastic mass of unclear etiology.

Diagnosis and management

The patient was observed and managed in a multidisciplinary way by the cardiology, cardiothoracic, and oncology teams. According to the myopericarditis diagnosis, anti-inflammatory therapy was initiated [nonsteroidal anti-inflammatory drugs (NSAIDs) and colchicine], with a favorable response, translated by chest pain resolution and normalization of inflammatory markers. Additional testing was performed to pursue a definitive LA mass etiology. Cardiac magnetic resonance (CMR) showed a regular LA mass (69×30×38 mm) with a lobulated outline appearance. There was an intimal relation with the LA inferior, posterior, and lateral walls, without clear cleavage plans. There was also an extension to the inferior pulmonary veins.

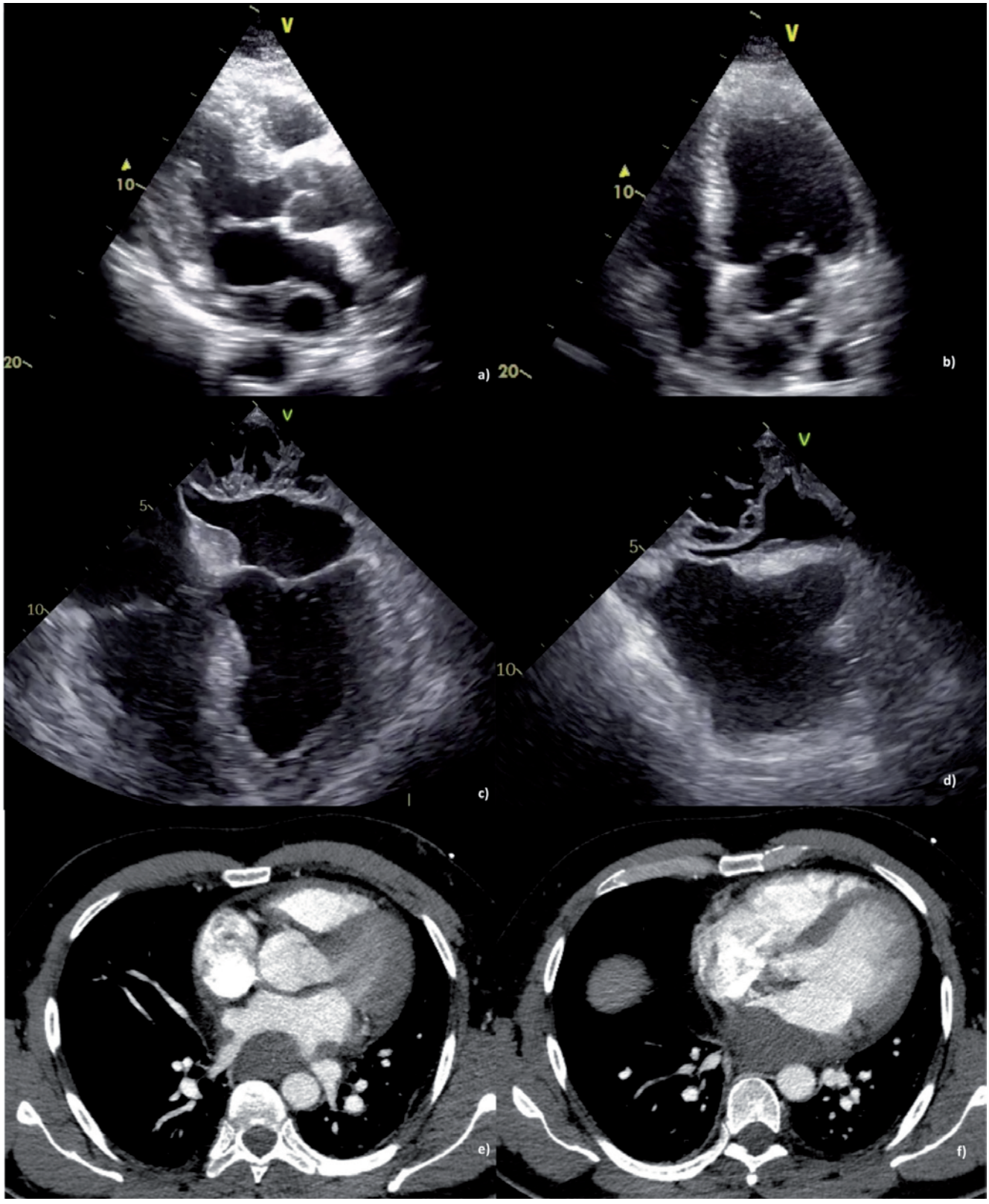

Figure 1. Echocardiographic and computed tomography images at admission showing the left atrial mass. a) Parasternal long-axis view; b) transthoracic four-chamber view; c) mid-esophageal four-chamber view; d) mid-esophageal bicaval view; e,f) computed tomography angiography images highlighting the hypovascularized nature of the mass.

Concerning tissue characterization, T1-isointensity and global T2-hypointensity with some hyperintense areas were observed; late gadolinium enhancement was present in the peripheral region of the mass and absent at its central core (Figure 2a-c). Altogether, CMR pointed out signs of malignancy and sarcoma as the most likely diagnostic hypothesis. Simultaneously, an abdominopelvic CT was also requested to exclude the presence of extra-cardiac lesions. After a multidisciplinary discussion of the case, histopathologic characterization of the mass was decided as the most appropriate next step. Thus, a surgical mass biopsy was performed. Extemporaneous histopathological examination of surgical samples showed the presence of non-malignant inflammatory tissue. After this result, LA mass resection was attempted in the same surgical time (Figure 2d). Complete removal was not possible due to the absence of dissection plans. More tumoral samples were collected for further histopathological study. The patient presented a good post-operative evolution and was discharged after 5 days with a possible inflammatory myofibroblastic tumor (IMT) diagnosis. Acute pericarditis standard of care treatment was maintained at discharge (NSAID therapy with a tapering plan, a 3-month colchine course, and gastroprotection during the treatment). The patient was referred to outpatient assessment after discharge for clinical and imaging reevaluation.

Follow-up

The patient was serially evaluated in an outpatient setting. At the 1-year follow-up, he presented no recurrence of symptoms, and a new CMR showed a small reminiscent of the LA mass, without contrast enhancement (Figure 3a-c). In the meantime, histopathological study of the additional surgical specimens became available and corroborated the extemporaneous findings, revealing the presence of fibrosis and cardiac muscular tissue, in a hypocellular, scar-like pattern [1] (Figure 2e). Overall, these results sustained the IMT hypothesis.

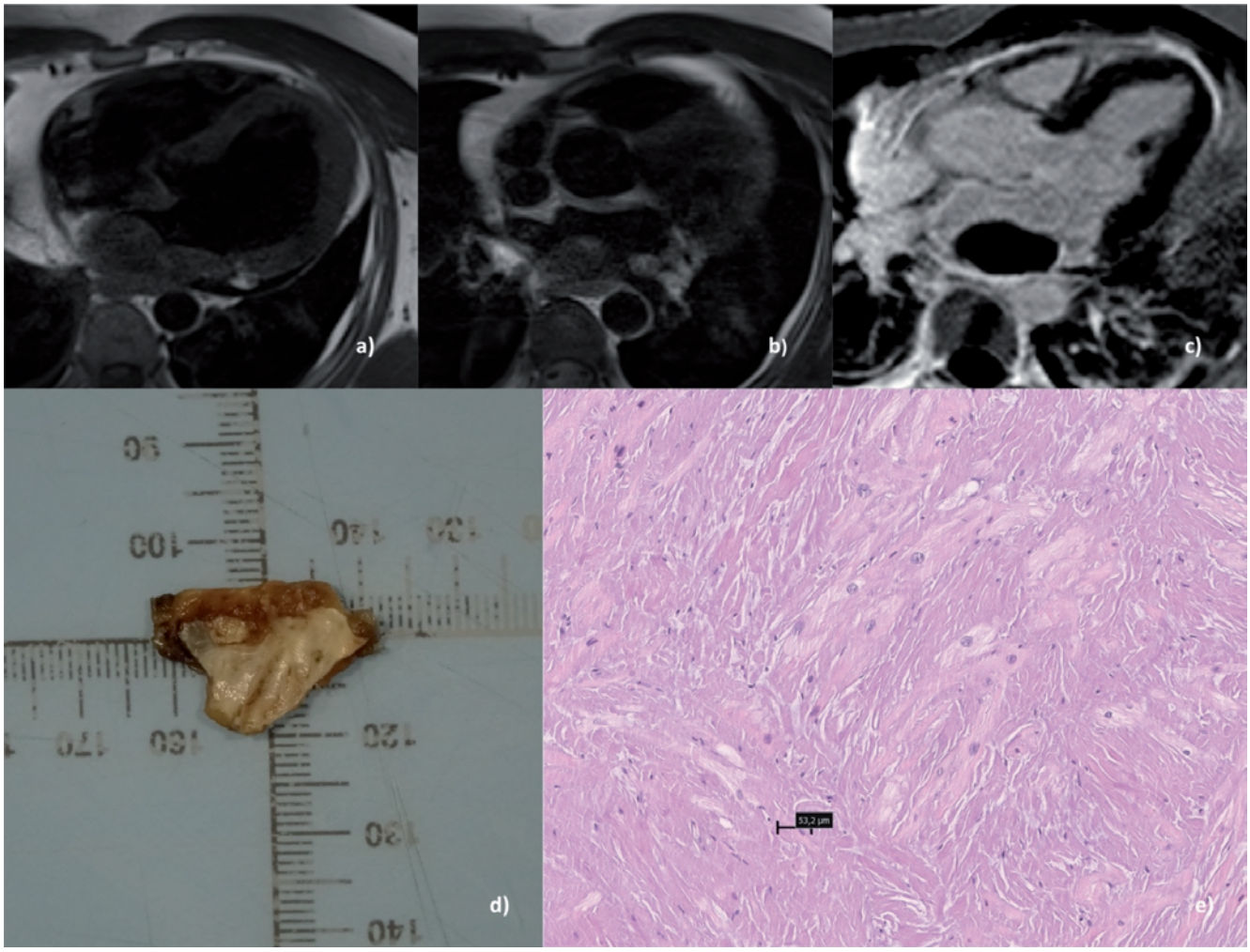

Figure 2. Cardiac magnetic resonance and histopathologic characterization of the mass. a) Four-chamber T1-weighted image; b) four-chamber T2-weighted image; c) late gadolinium enhancement image; d) macroscopic view; e) histological view.

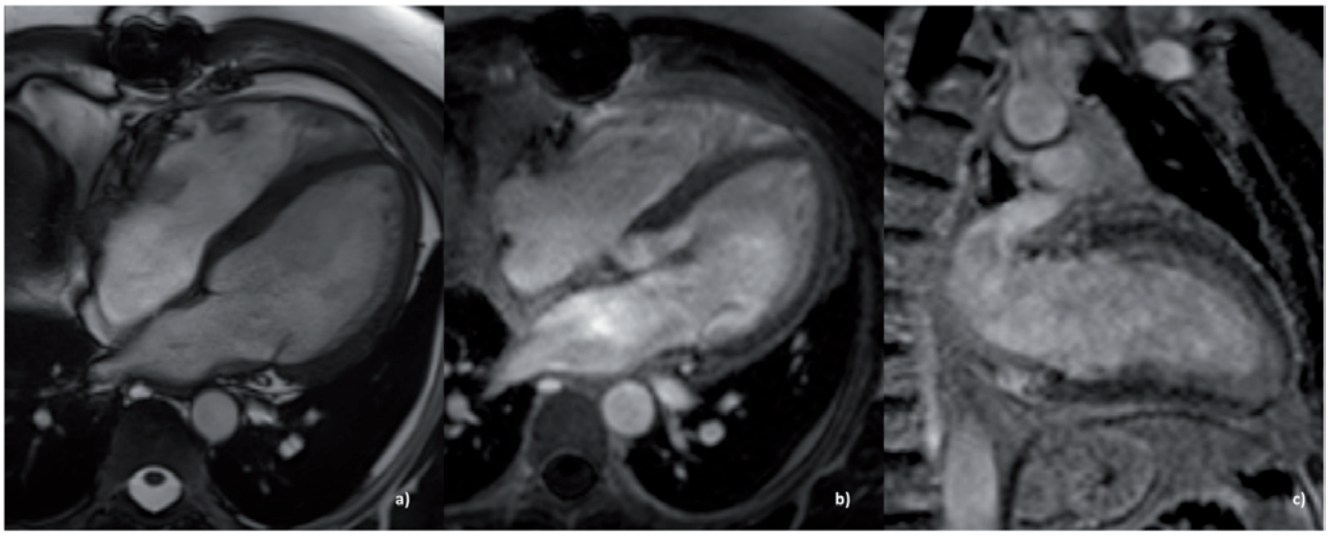

Figure 3. Cardiac magnetic resonance reevaluation at the 1-year follow-up. a) Four-chamber cine image; b) four-chamber late gadolinium enhancement image; c) two-chamber late gadolinium enhancement image.

This is a very rare entity and is considered a tumor of intermediate or uncertain behavior [2-5]. Most reported cases occurred in young people and at an atrial location [2,5]. Pathophysiological mechanisms are currently unknown [5]. Multi-imaging and histopathological assessment are essential for diagnosis [1,4], which is usually difficult and complex [5,6]. Main DD comprise benign (fibroma or myxoma), as well as malignant tumors, namely sarcoma [5,7]. Surgical resection is the recommended therapeutic approach, considering IMT has a high risk of complications [2,5,6]. Tumor recurrence has been documented, though its rate has not been properly evaluated due to IMT's rarity [2-6].

In accordance with the paucity of data, maintenance of a (likely) lifelong follow-up was decided and explained to the patient, with annual imaging and clinical reassessment.

Discussion and Conclusions

This case presents a rare diagnosis after an unexpected finding in a young male presenting with chest pain. Initial MI/myopericarditis DD would never predict the complex diagnostic work-up ahead. Chest pain nature (pleuritic), elevated inflammatory markers, residual increase of troponin values, as well as echocardiographic documentation of a small circumferential pericardial effusion, and the unexpected finding of a prominent and infiltrative LA mass, made myopericarditis, in the context of the inherent inflammation related to the mass, the most likely diagnosis. Focusing on the LA mass etiological study, the first imaging findings (TTE, TEE, and CTA) pointed to a hypovascular tumor as the likely diagnosis. CMR shed additional light, as sarcoma came out as the main DD hypothesis. After exclusion of extra-cardiac lesions, the final diagnostic step comprised a histopathological study of the mass. Unpredictably, but fortunately, both histopathological studies showed non-malignant inflammatory tissue, with identifiable cardiac muscle tissue and fibrosis. The patient underwent partial mass resection. IMT stood out as the most likely diagnosis. This is a rare tumor with uncertain behavior, mainly occurring in young people and atrial location [1-4]. Clinical presentation and outcomes are variable, according to mass location and size [4]. Both multi-imaging and histopathological evaluation are essential for diagnosis, which is commonly challenging due to IMT's rarity and scarce knowledge about its characteristic findings. Sarcoma is a frequently strong DD, as observed in this case. IMT histological reports usually show the presence of fibroblasts and myofibroblasts with little/no atypia or mitoses (different from sarcoma), and inflammatory cells [1,4]. Hypocellular, scar-like patterns, resembling our histological findings, have also been reported [1]. Surgical resection is recommended given the risk of potentially fatal outcomes (through mass prolapse/embolization) and the possibility of recurrence [1-5].

This is a unique case by the unexpected findings at presentation, as well as the complex diagnostic work-up and the surprisingly unusual final diagnostic hypothesis. It also highlights the increasing importance of a multimodality imaging approach, as well as the critical role of multidisciplinary discussion in optimizing patient management in such complex cases.

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