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Case Report

Incidental cardiac lymphangioma misdiagnosed as atrial thrombus: A case report ☆,☆☆

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ABSTRACT

Cardiac lymphangioma is a characteristically benign primary neoplasm of the heart, previously reported only in a handful of cases. A right atrial lesion was found of a 56-years old healthy male patient. The lesion was surgically excised and identified as cardiac lymphangioma in postoperative pathological analysis. While open surgical tumor resection is preferred in patients with cardiac lymphangioma, preoperative characterization of suspected lesions may warrant conservative management in selected cases.

Introduction

Cardiac lymphangioma, benign in nature, is an exceptionally rare primary neoplasm of the heart, with only a handful of cases reported in the literature [1]. While it may be clinically asymptomatic, various symptoms, ranging from mere arrhythmia to chest pain, valvulopathy, or heart failure, have been described [2–5]. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are commonly utilized to discern and outline the suspected pathology [1,2]. Open surgical tumor resection is the preferred mode of treatment and has been reported to yield favorable long-term outcomes [1].

We report a case of cardiac lymphangioma, incidentally discovered as a right atrial mass (RA) through preoperative workup of a 56-year-old male victim of traumatic falling, which was initially misdiagnosed as RA thrombus.

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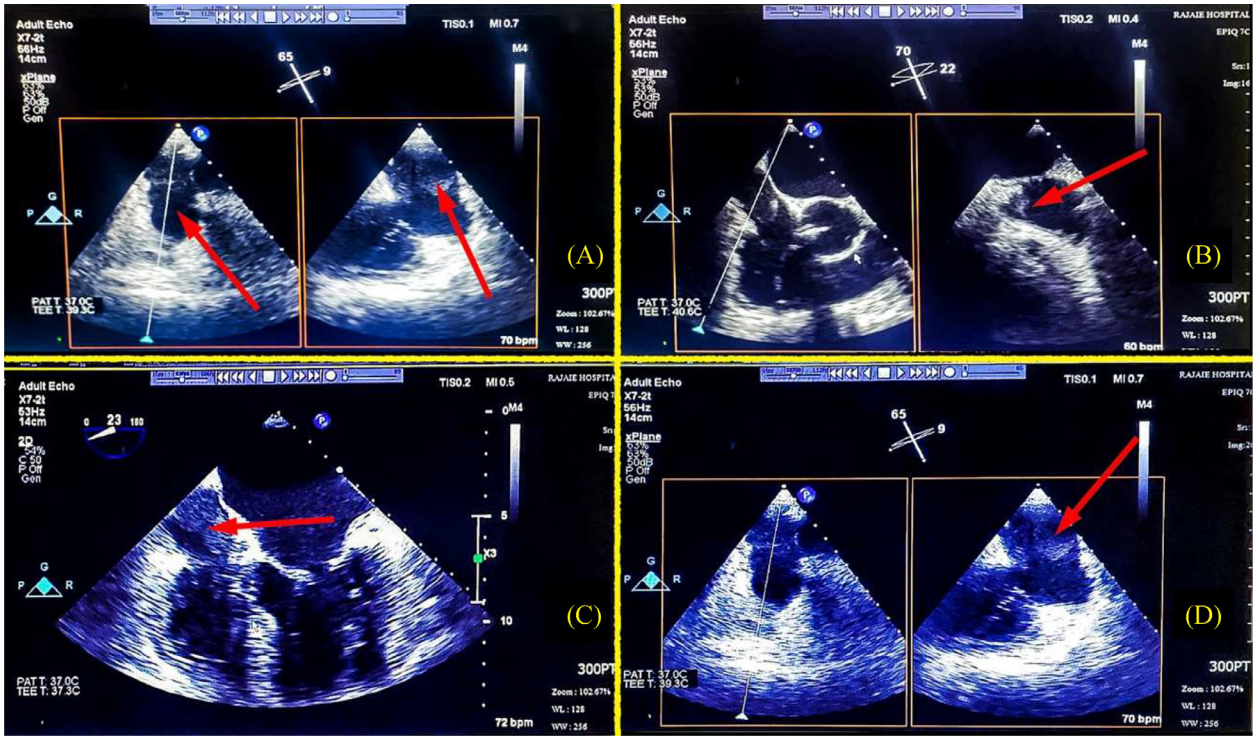

Fig 1 – Echocardiography of cardiac lymphangioma. A large, semimobile, well-defined multilobulated, hypoecho mass in RA cavity causing turbulency but no significant gradient at the site of IVC entrance to RA.

Case presentation

A 56-year-old previously healthy man presented to the emergency department following left shoulder trauma secondary to isolevel falling. Radiologic workup revealed a rotator cuff ligament tear, and the patient was referred to anesthesiology clinic for preoperative evaluations. Standard 12-lead electrocardiogram revealed dysrhythmia, and transthoracic echocardiography exhibited moderate mitral regurgitation, severe mitral stenosis, moderate aortic insufficiency, and moderate tricuspid regurgitation, prompting further cardiac workup. While confirming previous findings, transesophageal echocardiography (TEE) detected moderate aortic stenosis, severe left atrial enlargement, and spontaneous echo contrasts ("smokey" pattern) in the left atrium and left atrial appendage (Figs. 1 and 2). A semi-mobile, well-defined bilobed hypoechoic mass (4 × 2 cm and 4 × 8 cm) displaying abundant smokey patterns situated in the right atrium adjacent to the inferior vena cava (IVC) opening was observed to subject the IVC inflow tract to turbulence. Ejection fraction was calculated to be 55%, and the right atrial size was deemed as normal.

The right atrial mass was interpreted as a freshly formed thrombus, and the patient was accordingly prepared for emergency open surgical evacuation. Cardiac exploration under general anesthesia and cardiopulmonary bypass revealed the previously presumed right atrial thrombus to be a vascular mass. Total excision of the vascular mass was followed by aortic and mitral valve replacement, and tricuspid valve repair. A left atrial thrombus was intraoperatively detected and evacuated. The total durations of aortic clamp and cardiopulmonary bypass were 60 and 110 minutes, respectively. The patient was cautiously monitored in the cardiac intensive care unit (cICU) for 48 hours post-operatively to ensure hemodynamic stability, and then transferred to the post-cICU ward. Three days later, the patient was discharged in good condition.

Pathological analysis uncovered lymphoid tissue within the resected vascular mass, and immunohistochemical staining characterized it as cardiac lymphangioma.

Discussion

Cardiac tumors, while first described in 1911, were not identified in a living patient until 1934 [2,4]. Myxomas, lipomas, papillary fibroelastoma, and rhabdomyomas constitute the majority of benign primary cardiac tumors, while malignant primary cardiac tumors are more commonly identified as sarcoma subtypes, including angiosarcoma, undifferentiated sarcoma, malignant fibrous histiocytoma, leiomyosarcoma, and osteosarcoma [6,7]. Ever since the first description of cardiac tumors, only a handful of cardiac lymphangiomas have been reported in the literature [1]. Understandably, the frequency of cardiac lymphangiomas has never been described; however, the total incidence of primary cardiac tumors is reported to fall within the range of 0.001% to 0.3% in autopsy series [8].

The clinical presentation of cardiac lymphangiomas may vary from one patient to another. In symptomatic cases, clin-

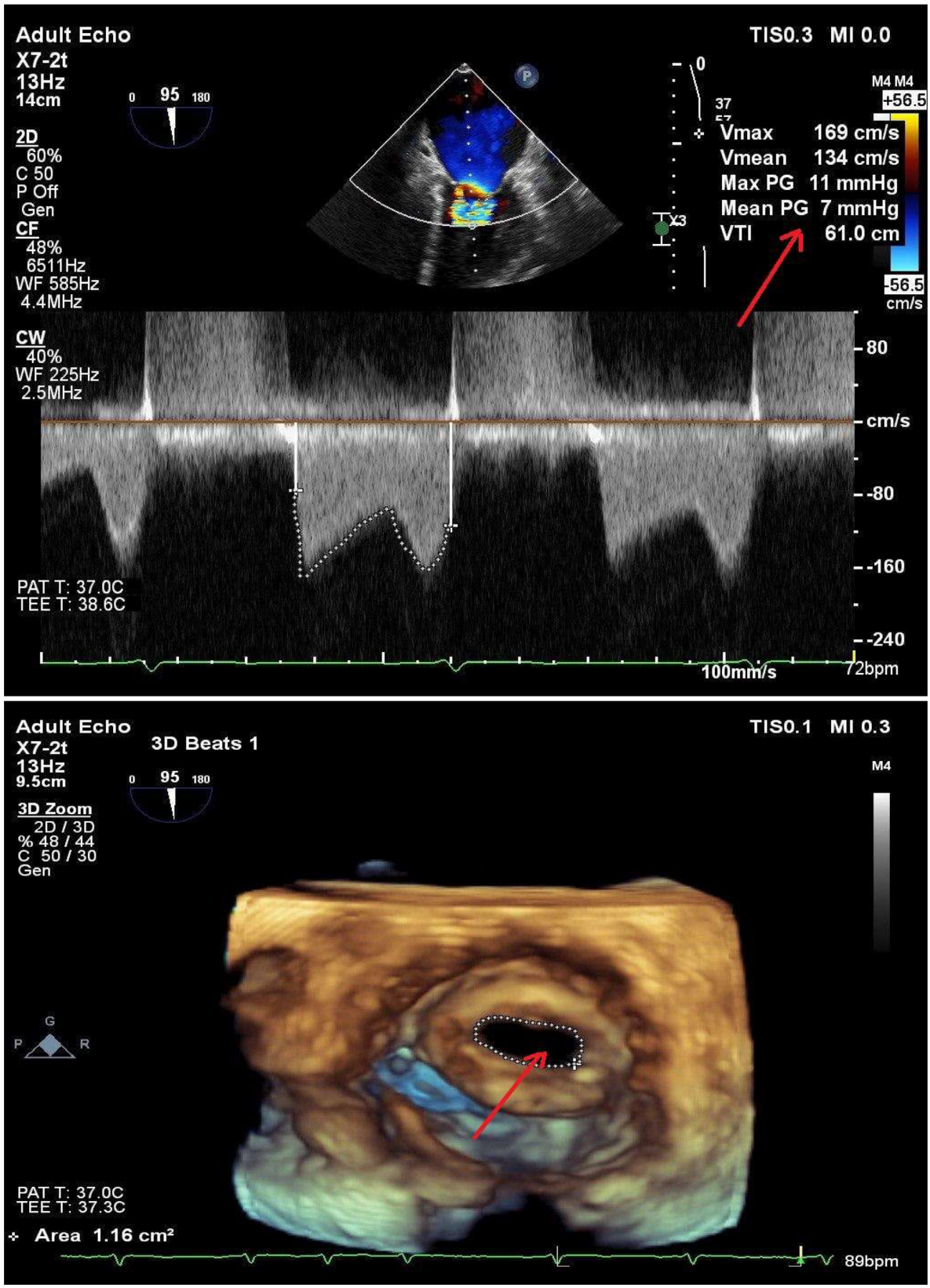

Fig 2 – Manifestation of cardiac lymphangioma. A severe rheumatismal mitral stenosis (MVA by 3D planimetry = 1.16 cm^2, MG = 7 mmHg).

ical manifestation largely depends on the anatomical location of the lesion and its exerted mass effect, and include arrhythmia, palpitation, chest pain, valvulopathy, and heart failure [1–5,9]. Asymptomatic lymphangiomas, however, are not uncommon and are therefore found only incidentally [2,3]. Association of cardiac lymphangiomas with other lesions, including breast cancer, anatomically distinct lymphangioma, and cardiac angiosarcoma, is well-described, and may facilitate the detection of asymptomatic cardiac lymphangiomas [1,10]. Conversely, laboratory investigations may display nonspecific disturbances such as leukocytosis, elevated erythrocyte sedimentation rate or C-reactive protein, thrombocytopenia, or hemolytic anemia, which are present in a host of other conditions and might complicate differential diagnoses [11].

While cardiac lymphangiomas may vary in their clinical presentations, almost all of the reported cases have been uniformly treated with open surgical tumor resection [1]. In spite of the benign nature of cardiac lymphangiomas, fear of possible cardiac and hemodynamic complications, or lack of preoperative histopathological characterization of the lesion may justify the clinical inclination for surgical treatment [10,12]. Conversely, treatment of other primary cardiac tumors is based on their symptomatology and histopathological nature, which could mandate unnecessary surgical interventions [6]. Accordingly, authors of this report suggest that conservative management of cardiac lymphangiomas in asymptomatic cases or cases with clinically insignificant symptoms, deserve further consideration by means of a thorough decision-making process. While biopsy is the gold standard of histopathological characterization of the suspected lesion, and novel imaging methods such as contrast echocardiography may lessen the risk of complications associated with cardiac biopsy [2]. Additionally, not only the pretest probability of malignancy in suspicion of a primary cardiac tumor is substantially lower than its benign counterpart, but also a neoplastic mass defined as lymphangioma is characteristically unlikely to go through malignant transformation [9,13]. Else, lymphangiomas of other anatomical sites have been reported to spontaneously resolve or regress, and since the natural history of cardiac lymphangiomas is not comprehensively delineated, this is a point worth noting [13–15].

According to the review of Diao et al., fourteen cases of cardiac lymphangioma had been reported until 2018 [1]. We conducted a PubMed search for cardiac lymphangioma cases being reported between 2018 and 2022 using the MeSH terms “heart” and “lymphangioma.” Only one case of cardiac lymphangioma was reported in the respective timeframe; Ko et al. described a variant of cardiac lymphangioma in the form of hygroma at the superior vena cava opening. A cardiac lesion causing superior vena cava syndrome in a patient with a history of cardiac angiosarcoma, was surgically excised in case of malignant recurrence, which turned out to be hygroma [10].

Conclusions

In conclusion, a case of a 56-year-old man presenting with left shoulder trauma resulting in a rotator cuff ligament tear led to the incidental discovery of a complex cardiac pathology. Initial evaluation revealed multiple valvular abnormalities and a suspected right atrial thrombus, later identified as a cardiac lymphangioma. Prompt surgical intervention, including excision of the mass and valve replacement, resulted in successful management of the condition. This case underscores the importance of comprehensive cardiac assessment in trauma patients and highlights the diagnostic challenges associated with rare cardiac tumors. Early recognition and appropriate management are crucial for optimizing patient outcomes in such cases.

Patient consent

Authors have obtained written informed consent for publication of this article from the patient(s), their guardian(s), or legal representative(s).

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