INTRODUCTION

Left ventricular pseudoaneurysm (LVPA) is a rare and life-threatening complication that results from cardiac rupture contained by the pericardium and, unlike a true aneurysm, it is characterized by the absence of myocardial tissue in its wall [1,2]. Although LVPA may occur after cardiac surgery, trauma, or infective endocarditis, the most common cause is myocardial infarction (MI) [1]. The incidence of LVPA following acute MI is 0.2% to 0.3% [3]. Polycythemia vera (PV) is a well-known cause of acute MI. However, the association of LVPA formation to PV is unknown and, to our knowledge, has never been reported. Here we describe the case of a 58-year-old man with PV who was admitted for acute dyspnea following an inferior myocardial infarction that occurred seven weeks prior to presentation. Multimodality imaging disclosed the presence of an LVPA. The patient refused surgical treatment and was readmitted three months later with acute decompensation. Follow-up imaging revealed increased LVPA. Review of the literature however showed no report of PV presenting as an LVPA that worsened in a follow-up admission. This rare association prompted a definitive diagnosis. In this case, multimodality imaging was crucial to establishing a definitive diagnosis and guiding therapy.

Key words · Polycythemia vera · Pseudoaneurysm · Myocardial infarction · Echocardiography · Computed tomography, x-ray · Coronary angiography.

CASE REPORT

A 58-year-old man who had experienced inferior MI seven weeks prior to presentation and had been non-compliant to his treatment, was admitted for acute dyspnea. His medical history included PV that was diagnosed two years ago but was poorly managed with irregular phlebotomies.

On presentation, he had tachypnea (respiratory rate of 24 breaths per minute), tachycardia (heart rate of 108 beats per minute), oxygen saturation of 93% on ambient air, and a normal blood pressure of 110/70 mm Hg. Chest auscultation revealed right basal crackles and decreased air entry on the left middle and lower lung fields. Cardiac auscultation revealed regular tachycardia without heard murmurs but with a friction rub best heard at the apex. A chest radiograph showed cardiomegaly and a large left pleural effusion. An electrocardiogram showed sinus tachycardia with Q waves in the inferior leads. Laboratory work-up was unremarkable except for a hemoglobin level of 17 g/dL.

Two-dimensional transthoracic echocardiography (2D-TTE) showed a severely hypokinetic inferolateral wall with an outpouching formation contained by pericardial fluid and thrombus (Fig. 1A, Supplementary Video 1 in the online-only Data.
Supplement). A CT scan of the chest after administration of intravenous contrast showed a hyperdensity arising from the lateral border of the LV with an irregular contour, in addition to a layered hyperdensity in the adjacent pericardium, suggestive of chronic/old leakage (Fig. 1B, Supplementary Video 2 in the online-only Data Supplement). The hemopericardium had reached a thickness of 2.4 cm (Fig. 1B, Supplementary Video 2 in the online-only Data Supplement). Three-dimensional reconstruction cardiac CT scan showed the presence of the same outpouching (Fig. 1C). The findings were suggestive of an LVPA protruding from the inferolateral wall. The patient was admitted to the coronary care unit for stabilization and further treatment. Invasive coronary angiography showed total occlusion of the proximal left circumflex artery, with contrast layering and delayed contrast clearance suggestive of thrombus formation (Fig. 1D, Supplementary Video 3 in the online-only Data Supplement). The remaining coronary arteries were normal.

The patient was referred for cardiac surgery but refused treatment and was lost to follow-up. Three months later, he presented to the emergency department with severe dyspnea. 2D-TTE showed a large, thin outpouching protruding from the LV with a “to-and-fro” color Doppler signal (Supplementary Video 4 in the online-only Data Supplement). A CT scan of the chest revealed a large LVPA (Fig. 2). The patient was transferred to another hospital for emergent surgery.

DISCUSSION

LVPA occurs when rupture of the free wall is contained by overlying adherent pericardium and organizing thrombus. The pathogenesis begins with a small endocardial rupture within one day of the onset of MI or becomes associated with an intramural hemorrhage dissecting the myocardium and ruptures several days later. Distinguishing true from false aneurysms is challenging but crucial as false aneurysms have a higher propensity for rupture. When the pericardium is adherent at the site of rupture, an LVPA is formed. Patients usually present with symptoms of heart failure such as dyspnea or angina [3]. Inferior infarcts occur at approximately twice the incidence rate of anterior infarcts [4]. LVPA is usually an incidental echocardiographic find-
Color-flow imaging by 2D-TTE is of significant value to establish the diagnosis, allowing detection of flow in and out of the pseudoaneurysm and within the pericardial space, even if pulsed-wave and continuous-wave Doppler were unhelpful [5]. Despite this, CT scan seems more sensitive for identification of the anatomy, especially when three-dimensional reconstruction is used [6], as in our case.

On the other hand, PV is a primary disorder of bone marrow stem cells, resulting in overproduction of red cells and, to a lesser extent, neutrophils and platelets. The incidence rate is 2.3 per 100,000 persons per year [7]. Patients with PV are at particular risk for arterial and venous thrombosis, and its significant associated morbidity and mortality. Thrombotic events are present in 20% to 50% of patients at the time of diagnosis and involve major vessels and the microcirculation [8]. Although LVPA is a rare complication of MI, more evidence is needed to demonstrate a direct association between PV and LVPA.

Coronary events are not uncommon during the course of PV. In a study on 149 patients diagnosed with PV who were followed for ten years, Rossi et al. [9] found that 11.4% had MI. Despite the association of PV with coronary artery disease, its presentation as acute MI is rare [10-12]. Nevertheless, patients with PV have never presented with a rare complication such as LVPA. The diagnosis described in this report was confirmed by multimodality imaging since, to our knowledge, there have been no reports describing the current association. In addition, despite the urgent surgical need for intervention, the patient survived this complication for five months from his initial presentation.

In conclusion, PV has frequently been linked to MI but the association with LVPA has never been described. The natural course of LVPA warrants more evidence to demonstrate a direct association with PV complicated by MI. This finding prompted further investigation to confirm the diagnosis. In this case, the worsening size of the LVPA was demonstrated with multimodality imaging, which was crucial to establishing a definitive diagnosis and guiding therapy.

Supplementary Movie Legends

Video 1. 2D-TTE subcostal view showing color Doppler in the LVPA and the pericardium, suggestive of leakage (yellow arrow).

Video 2. Cine-loop CT scan of the chest showing the LVPA (yellow arrow) and the surrounding thrombus formation, pericardial fluid, and leakage (yellow asterisk). Note the presence of hemothorax.

Video 3. Cine-loop coronary angiography showing a totally occluded left circumflex artery with delayed contrast clearance, suggestive of thrombus formation (yellow arrow).

Video 4. 2D-TTE showing a “to-and-fro” color Doppler signal moving through the neck of the LVPA.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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