Polycythemia Vera: A Rare Ethiology of Heart Failure

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Introduction

The prevalence of heart failure (HF) with systolic dysfunction exceeds 20 million patients worldwide, which corresponds to 2% of the Western population.¹ Over the past two decades, there have been major advances in the outpatient treatment of individuals with HF, including the use of not only pharmacotherapy (use of beta-blockers and aldosterone receptor blockers),² but also implantable devices (resynchronization devices and implantable defibrillators).³ Despite all these advances, the mortality of patients with HF remains high, exceeding, for example, many oncological diseases. Another major treatment challenge in this area refers to the outcomes considered as secondary but also important, such as the improvement in quality of life and the reduction in the number of hospitalizations in these patients.¹

With regard to the etiology, there are different causes of HF, and their recognition can be crucial for treatment optimization. Ventricular dysfunction secondary to myocardial ischemia, arrhythmias, and hypothyroidism, among others, should be treated with a focus on the underlying disease.² Thus, the identification of the HF cause must be pursued until possibilities are exhausted, so that its treatment can be individualized. In fact, the so-called cardiomyopathies of idiopathic origin have a worse prognosis when compared with those whose etiology is known.⁴

The objective of this study was to report a case of HF secondary to polycythemia vera, a rare hematologic disease.

Keywords

Heart Failure / etiology; Polycythemia Vera / etiology; Microcirculation; Hematologic Diseases / physiopathology.

Case report

The patient is a 47-year-old dark-skinned man, born in Belo Horizonte (MG) and currently working as a gardener in the state of Rio de Janeiro. He sought the emergency service reporting dyspnea for 2 months, initially to moderate effort, which had progressed to dyspnea at rest in the previous week. Associated with the presentation, he reported orthopnea and paroxysmal nocturnal dyspnea. He denied chest pain, fever, weight loss, or recent infections. In his past medical history, he had no reports of comorbidities, hospitalizations, previous diseases, or surgeries. He denied allergies, smoking, and alcoholism. Despite working as a gardener, the patient was a retired military and had worked with ship boilers for several years. He was unaware of the cause of death of his parents and had two sisters who were alive and apparently healthy. There were no other relevant data in his medical history.

On physical examination, the patient was eupneic at rest. Had no pallor of conjunctiva or mucous membranes, was anicteric, acyanotic, hydrated, and presented lower extremities edema. He presented jugular venous distension at 45°, a two-paced regular cardiac rhythm, normal cardiac sounds, a heart rate of 92 bpm, and blood pressure of 110 x 60 mmHg. There were no murmurs or pericardial friction.

On pulmonary auscultation, he presented decreased breath sounds at both lung bases, with crackles in the lower two-thirds of both lungs. His abdomen was soft, tympanic, with peristaltic movements, and painful on deep palpation in the right hypochondrium. His liver had normal measurements and a blunt edge, and was painful to palpation. His Traube’s space was occupied, but his spleen was not palpable. There was no peripheral edema.

Considering a diagnosis of HF, treatment with vasodilators (angiotensin converting enzyme inhibitor)
and a diuretic (furosemide) was initiated and tests were requested. Since the patient lived far from the hospital, it was decided to admit him to better evaluate and stabilize his clinical condition.

His tests revealed polycythemia (with a hematocrit of 50.7% and a hemoglobin level of 17.1 g%) with normocitosis. Leukogram, biochemistry, and liver and thyroid function tests were normal. Transthoracic echocardiography revealed dilated cardiomyopathy with a left ventricular ejection fraction of 24%.

Despite the patient’s clinical improvement, his medical history precluded an identification with a certainty of the cause of his severe ventricular dysfunction, considering that he was young and had no classic risk factors for HF. To search for his condition's etiology, myocardial scintigraphy with dipyridamole was performed, blood cultures were collected, and tuberculosis, HIV, Chagas disease, and collagen diseases were investigated. All these tests were negative. Considering the increased hematocrit, we chose to perform a pulmonary evaluation with spirometry at rest, which was also normal.

Finally, his increased hematocrit in the absence of smoking or lung disease led to an investigation of polycythemia vera, which was confirmed. The patient was offered the option to undergo endomyocardial biopsy but refused it. Cardiac magnetic resonance was not available; therefore it was not performed.

With the diagnosis of polycythemia vera, aspirin was added to the patient’s treatment. After hospital discharge, the patient was referred to a cardiology and hematology service at his city for follow-up.

**Discussion**

Polycythemia vera, a myeloproliferative disease, was first described by Vasquez in 1892 and, subsequently, by Osler in 1903, and is also known as Osler-Vaquez disease. It is a rare disease (4–16 cases/million), characterized by increased hematocrit and, consequently, blood viscosity.

The relationship of ventricular function seems to show a paradoxical behavior in respect to polycythemia vera. A study conducted in the 1960s examined invasively examined the hemodynamic profile of 10 patients with polycythemia vera and compared them with those of healthy individuals, concluding that patients with polycythemia vera exhibited hypervolemia, which translated into an increased cardiac output and oxygen consumption. Taking into account the pathophysiological mechanisms, the occurrence of hypervolemia and the increase in hematocrit may justify the higher cardiac output and aerobic capacity. However, this statement is true up to certain limits.

In fact, extreme increases in hematocrit may increase the blood viscosity to levels that lead to higher blood viscosity levels to hypercoagulability. However, the thrombophilic mechanism in patients with polycythemia vera is, in fact, even more complex. It includes not only the increased hematocrit and platelet count rates, but also the interactions among platelets, leukocytes, and cellular products and a reduction in endogenous anticoagulants. Most of the few cases found in the literature in which polycythemia vera has led to HF occurred due to hypercoagulability. These cases have reported massive intraventricular thrombi causing obstructive HF, with no direct damage to the mechanisms of ventricular contraction.

In the international literature, we found only one case of polycythemia vera that evolved to ventricular dysfunction due to cardiomyocyte necrosis and slow blood flow in the coronary microcirculation, probably due to hyperviscosity. In spite of our patient having refused to undergo endomyocardial biopsy, the absence of intracardiac thrombi on echocardiography makes the hypothesis of microinfarcts the only possible pathophysiological mechanism for his ventricular dysfunction. It is worth noting that the negative outcome for myocardial ischemia on myocardial scintigraphy does not exclude our pathophysiological hypothesis since this method does not have adequate sensitivity to identify microinfarcts.

As illustrated in this case, the certainty of the etiological association in cases of HF is not always simple and straightforward. In fact, the reasonable pathophysiological implication and the absence of other possible etiologies are the main factors that lead to the etiological definition (or strong suspicion) in most cases of nonischemic HF in clinical practice.

In the present case, once the main HF causes among us were excluded, and due to the refusal of the patient to undergo endomyocardial biopsy, only two options would remain for the diagnostic conclusion: idiopathic nonischemic HF in a patient with polycythemia vera or HF secondary to polycythemia vera.

Considering the rarity of polycythemia vera, the existence of previous reports pointing to the possible development of HF in patients with this hematological disease, the absence of other causes for the HF in this patient, and the medical reasoning of always trying to explain a patient’s complaints with the lowest
possible number of diagnoses, we believe that the most reasonable option to consider is that of polycythemia vera as the cause of this patient’s HF syndrome, and not as an associated diagnosis in this case.

Conclusion

We present a case in which a causal association between polycythemia vera and HF was made by exclusion of other diagnoses. The etiology of HF must be sought in all new cases, because its recognition can direct the therapy and influence the prognosis of these patients.

Author contributions

Conception and design of the research: Castro RRT. Acquisition of data: Cardoso MB, Albuquerque MF, Pais RRS, Castro RRT. Analysis and interpretation of the data: Cardoso MB, Albuquerque MF, Pais RRS, Castro RRT. Writing of the manuscript: Cardoso MB, Castro RRT. Critical revision of the manuscript for intellectual content: Albuquerque MF, Pais RRS, Castro RRT.

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Study Association

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References