JACC: CASE REPORTS
PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION.

VOL. 30, NO. 5, 2025

HEART FAILURE AND CARDIOMYOPATHIES

CLINICAL CASE

Regression of Left Ventricular Hypertrophy After Polycythemia Vera Treatment With Incidental Finding of Unicuspid Aortic Valve

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ABSTRACT

We present a unique case of a 38-year-old man with 2 rare conditions: unicuspid aortic valve and polycythemia vera. The patient developed left ventricular hypertrophy, which showed regression after polycythemia vera treatment. This is the first reported case of left ventricular hypertrophy regression after polycythemia vera treatment. (JACC Case Rep. 2025;30:103176) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 38-year-old man presented to the cardiology clinic to establish care for hypertension (HTN) after being hospitalized for a hypertensive emergency when he had chest pain, shortness of breath, hemoptysis, night sweats, blurry vision, and numbness and tingling of his fingers and lower extremities. Physical examination was significant for 2/6, mid peaking systolic, crescendo-decrescendo murmur appreciated throughout the precordium but best heard at the right upper sternal border. Electrocardiograph met criteria for left ventricular hypertrophy (LVH) (**Figure 1A**). Transthoracic echocardiogram revealed severe LVH with an ejection fraction of >65% (**Figure 2A**, Video 1) and a bicuspid aortic valve with mild to moderate aortic stenosis (mean gradient 20) and moderate aortic regurgitation. He was managed with oral antihypertensive medications and discharged with outpatient follow-up.

TAKE-HOME MESSAGES

- PV is a myeloproliferative neoplasm that causes neoplastic proliferation of hematopoietic progenitor cells.
- It is associated with LVH which can regress after PV treatment.

PAST MEDICAL HISTORY

Past medical history included bicuspid aortic valve (later found to be unicuspid during surgery), severe aortic regurgitation secondary to abnormal aortic valve, polycythemia vera (PV), HTN, and anxiety. Initial laboratory findings showed elevated hemoglobin (21.5 g/dL) and hematocrit (65.3%). JAK2 V617F mutation was positive which is consistent with PV. This finding was confirmed by bone marrow biopsy.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received July 16, 2024; revised manuscript received October 23, 2024, accepted November 18, 2024.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2024.103176

ABBREVIATIONS AND ACRONYMS

HTN = hypertension

LVH = left ventricular hypertrophy

PV = polycythemia vera

DIFFERENTIAL DIAGNOSES

Differential etiologies for his LVH include uncontrolled HTN and PV.

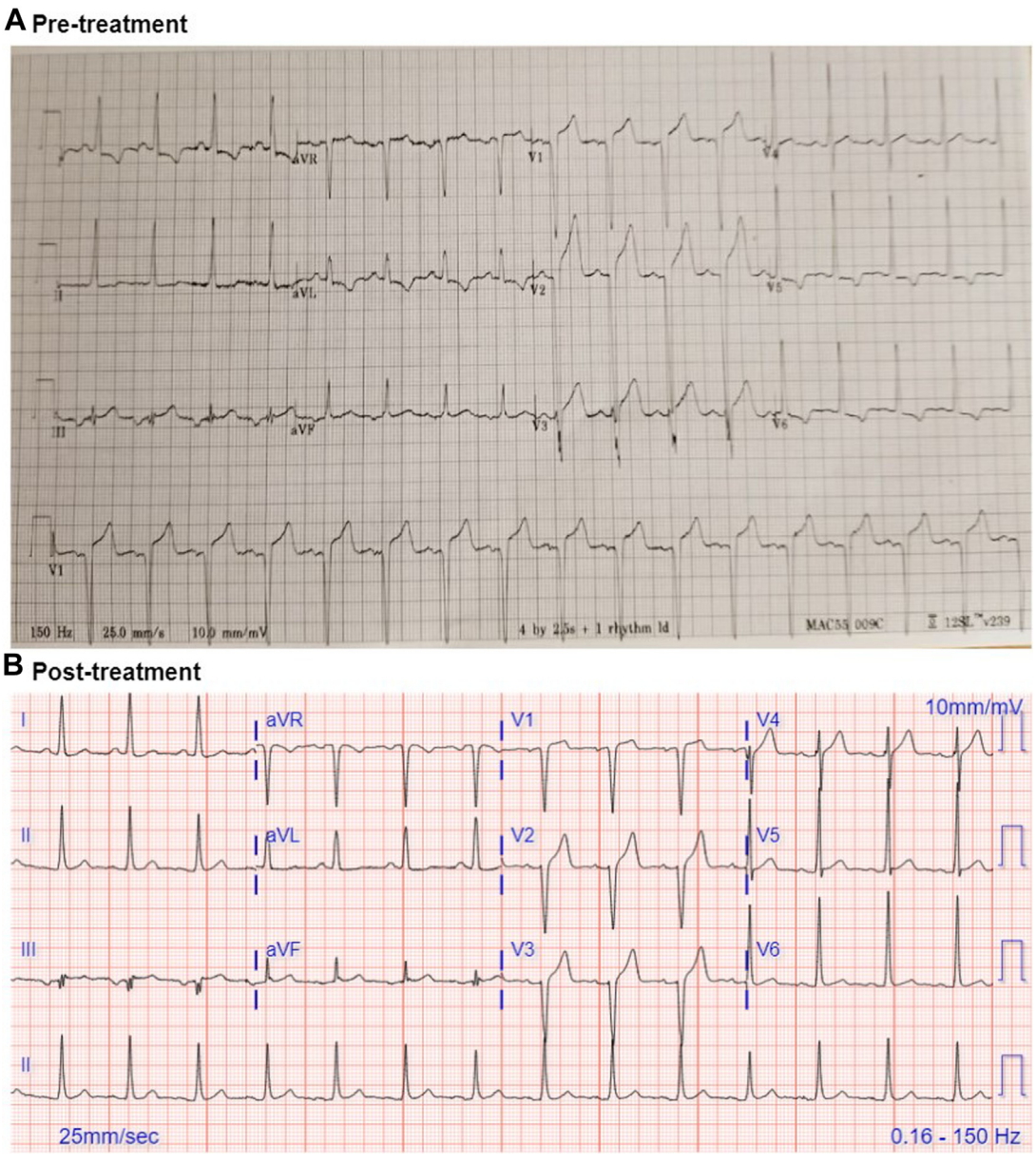
INVESTIGATIONS

Cardiac magnetic resonance showed a severe concentric LVH with maximum wall thickness of 2.4 cm, a bicuspid aortic valve, and findings concerning for an infiltrative cardiomyopathy (Video 3). The patient underwent an extensive infiltrative evaluation, including an endomyocardial biopsy, which were all unremarkable.

MANAGEMENT

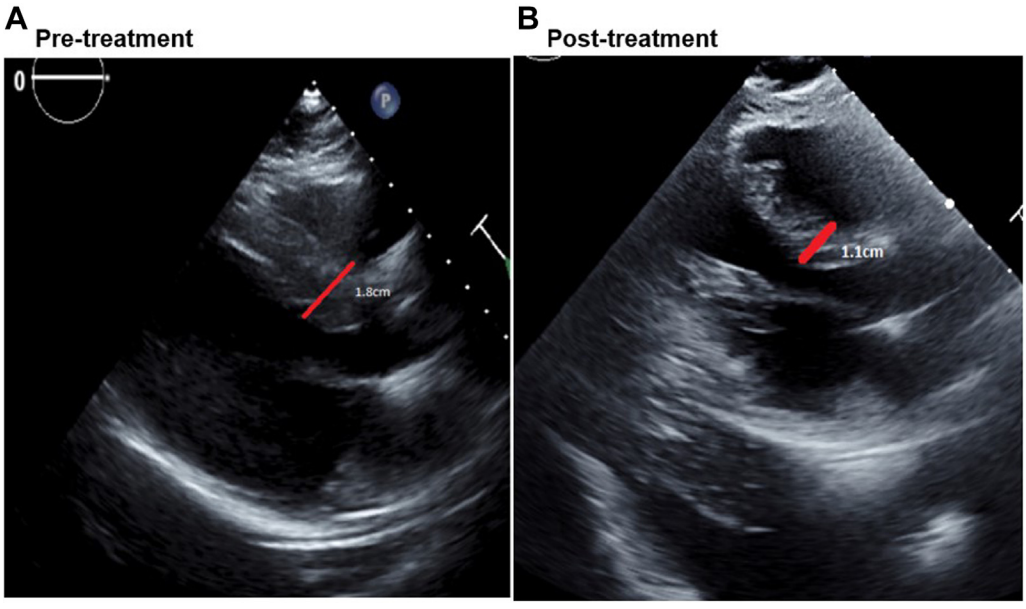
The patient's LVH with mid cavitary obstruction was thought to be likely secondary to uncontrolled HTN, which was compounded by PV. The patient was

FIGURE 1 Electrocardiogram



Left ventricular hypertrophy before polycythemia vera treatment (A) and after treatment (B).

FIGURE 2 Transthoracic Echocardiogram



Severe left ventricular hypertrophy before polycythemia vera treatment (A) and after treatment (B).

treated with phlebotomies every other week and started on aspirin and hydroxyurea, which resulted in improvement of the majority of his symptoms. Repeat echocardiogram 2 years later showed that his LVH had regressed with thickness decreased from 1.8 cm (**Figure 2A**, Video 1) to 1.1 cm (**Figure 2B**, Video 2). Repeat electrocardiograph showed improvement in LVH as well (**Figures 1A and 1B**). Hemoglobin was 21.5 g/dL before PV treatment and 16.6 g/dL after treatment, which correlated with LVH regression timing.

However, the patient developed significantly worsening shortness of breath. Repeat transesophageal echocardiogram showed a prolapsed aortic valve and severe aortic regurgitation. Aortic valve mean gradient was 27 mm Hg, peak velocity 3.2 m/s, aortic valve area 1.3 cm^2, and peak left ventricular outflow tract obstruction gradient 15 mm Hg with Valsalva. Therefore, the patient underwent aortic valve replacement (27-mm Inspiris) with root enlargement (Nicks procedure using a Dacron patch). Intraoperatively, he was found to have a unicuspid aortic valve intraoperatively (**Figure 3**).

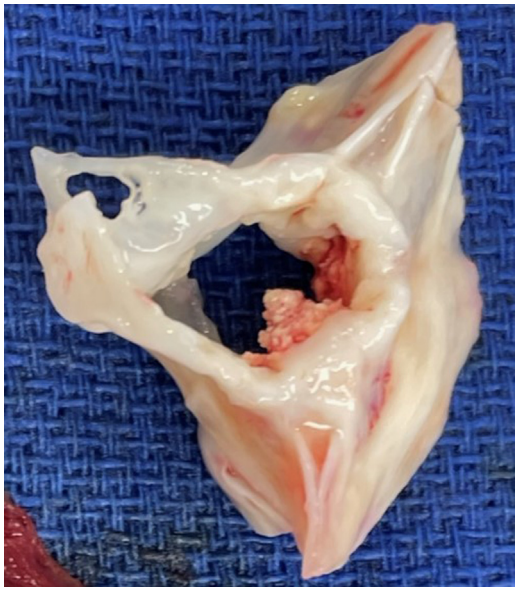
OUTCOME AND FOLLOW-UP

In follow-up, his functional symptoms have improved significantly. Postoperative transthoracic echocardiogram showed aortic valve mean gradient of 5 mm Hg, peak velocity 1.49 m/s, calculated aortic valve area 2.6 cm^2, and left ventricular outflow tract obstruction gradient of 4.7 mm Hg. There was no evidence of aortic valve stenosis or regurgitation.

DISCUSSION

Primary PV is a myeloproliferative neoplasm associated with JAK2 mutation. It can lead to elevated hemoglobin, hypercoagulability, and cardiac dysfunction as a consequence of increased blood viscosity.[1] High blood viscosity in PV affects the afterload of the heart and increases peripheral resistance resulting in an increased left ventricular wall strain leading to LVH. In addition, the activation of the JAK-STAT pathway is associated with cardiac fibrosis and cardiac remodeling in experimental studies with mice.[2] There is a >2-fold increase risk of heart failure in patients with myeloproliferative neoplasm compared to those without myeloproliferative neoplasm, with an incidence of 9.27 vs 3.7 per 1,000 person-years.[1] Overproduction of red blood cells and high hematocrit levels associated with PV can contribute to systemic HTN. Studies have shown that a concentric pattern of LVH is more prevalent in patients with high hematocrit levels than in those

FIGURE 3 Unicuspid Aortic Valve



with intermediate hematocrit levels.[3] It is important to note that this is the first reported case of LVH regression after PV treatment.

CONCLUSIONS

We present a unique case of a patient with 2 rare conditions: unicuspid aortic valve and PV. The patient developed LVH with mid cavitary obstruction secondary to uncontrolled HTN and PV. Notably, his LVH showed regression after PV treatment. This case demonstrates the benefits of early diagnosis and treatment of PV.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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REFERENCES

1. Leiva O, Hobbs G, Ravid K, Libby P. Cardiovascular disease in myeloproliferative neoplasms. *JACC CardioOncol*. 2022;4:166-182. https://doi.org/10.1016/j.jaccao.2022.04.002

2. Wang W, Liu W, Fidler T, et al. Macrophage inflammation, erythrophagocytosis, and accelerated atherosclerosis in Jak2 V617F mice. *Circ Res*. 2018;123:e35-e47. https://doi.org/10.1161/CIRCRESAHA.118.313283

3. Stritzke J, Mayer B, Lieb W, et al. Haematocrit levels and left ventricular geometry: results of the MONICA Augsburg Echocardiographic Substudy. *J Hypertens*. 2007;25:1301-1309. https://doi.org/10.1097/HJH.0b013e3280f9df97

KEY WORDS aortic valve, cancer, chronic heart failure, hypertension, valve repair

APPENDIX For supplemental videos, please see the online version of this paper.