Radiology Case Reports 19 (2024) 3386–3389

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Case Report

Acute myocardial infarction and polycythemia rubra vera: The double effect of treatment with hydroxyurea ☆,☆☆,★

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ARTICLE INFO

Article history:
Received 15 April 2024
Revised 1 May 2024
Accepted 3 May 2024

Keywords:
Acute myocardial infarction
Polycythemia rubra vera
Hydroxyurea

ABSTRACT

We report a case of acute myocardial infarction in a patient with polycythemia rubra vera, who has been treated with hydroxyurea. The patient presented with chest pain extending to both arms accompanied by nausea and sweating. Hemoglobin was 18.1 mg/dL, hematocrit 53.2%, white blood cells 9600/mm^3, and platelets 745,000/mm^3. The levels of specific cardiac injury markers were increased, troponin I increased to 110 ng/mL and creatine kinase-MB to 361 U/l, respectively. Electrocardiography showed sinus rhythm with ST-segment elevation in leads V2-6, D1, and aVL as well as ST depression in D2, D3 and aVF. Echocardiography demonstrated hypokinesis of the interventricular septum and lateral wall with mildly reduced left ventricle (LV) ejection fraction (EF≈45%). Coronary angiography revealed proximal-LAD subtotal occlusion and 80% mid-LAD stenosis with distal-LAD vasospasm. Percutaneous coronary intervention was performed with a drug-eluting stent in mid- and proximal-LAD. Hypercoagulable state of polycythemia rubra vera may be complicated with acute myocardial infarction, in addition to the vasospastic effect and endothelium lesions of hydroxyurea regardless its favorable effect as a standard therapy.

☆ Authorship declaration: The authorship is based on the latest guidelines of the International Committee of Medical Journal Editors. All authors listed are in agreement regarding the manuscript.

☆☆ Acknowledgments: This manuscript received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

★ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2024.05.014

Introduction

Polycythemia rubra vera (PRV) is a myeloproliferative disease characterized with an increased number of blood cells causing hyperviscosity. Acute coronary disease occurs in 11.4% of patients with polycythemia vera, more commonly in patients over 40 years of age. Most (>80%) of polycythemia rubra vera patients have a clonal and recurrent mutation in the JH2 pseudo-kinase domain of the Janus kinase 2 (JAK2) gene [1].

Hydroxyurea is the treatment of choice in high-risk subjects with this myeloproliferative disease and absolutely recommended in very high-risk subjects of any age. Additionally, there are data suggesting that this treatment could be used in relatively low-risk subjects [2–4]. Although is used as first-line therapy in PRV, thromboembolic complications (e.g. myocardial infarction (MI), ischemic stroke, peripheral artery thrombosis, deep venous thrombosis, etc.) are present in such patients treated with hydroxyurea, knowing its double effect [5].

We report a case of ST-segment elevation myocardial infarction in a patient with polycythemia rubra vera, who is treated with hydroxyurea.

Case presentation

A 35-year-old female patient presented to our emergency center with chest pain extending to both arms accompanied by nausea and sweating. Her history was negative for diabetes mellitus, hypertension, hyperlipidemia, smoking habit, and family history. Blood pressure was 125/85mmHg and heart rate was 105/min. Auscultation of the chest revealed normal lung sounds without rales and rhythmic heart sounds with no added sounds or murmurs. Abdominal examination did not show hepatosplenomegaly and Traube's space was not obliterated. Peripheral arterial pulses were palpable in all distal extremities.

Complete blood count analysis was as follows: hemoglobin 18.1 mg/dL, hematocrit 53.2%, white blood cells 9,600/mm^3, and platelets 745,000/mm^3. Serum concentrations of urea, creatinine, electrolytes, and hepatic functions were within normal range. She was also tested for

Janus kinase 2 (JAK2) V617F resulting positive. The levels of cardiac troponin I and CK-MB increased to 110 ng/ml and 361 U/l, respectively. Arterial blood gas analysis showed a PO2 of 95.6 mmHg.

Electrocardiography showed sinus rhythm, 100 bpm, left axis deviation, ST-segment elevation and inverted T waves in leads V2-6, D1, and aVL as well as ST depression in D2, D3, and aVF (Fig. 1). Echocardiography demonstrated hypokinesis of the interventricular septum and lateral wall with mildly reduced left ventricle (LV) ejection fraction (EF≈45%). Abdominal ultrasonography revealed that the liver size was within normal limits, but there was craniocaudal splenomegaly of 132 mm.

Coronary angiography revealed proximal-LAD subtotal occlusion, 80% mid-LAD stenosis and distal-LAD vasospasm (Left anterior oblique (LAO) cranial view) (Fig. 2). Percutaneous coronary intervention was performed with a drug-eluting stent in mid- and proximal-LAD (Fig. 3), with small thrombotic mass after stenting in mid-LAD (Fig. 3B). Following the PCI, we continued heparin administration for 2 days to maintain activated partial thromboplastin time at 60–80 s. We administered oral aspirin (100 mg) and prasugrel (10 mg) for 6 months and then aspirin monotherapy.

Discussion

There have been reported cases of ST-segment elevation myocardial infarction during hydroxyurea treatment in patients without prior heart disease [6,7]. Multiple factors are thought to be implicated in the hypercoagulability associated with polycytemia rubra vera including: increased hematocrit and

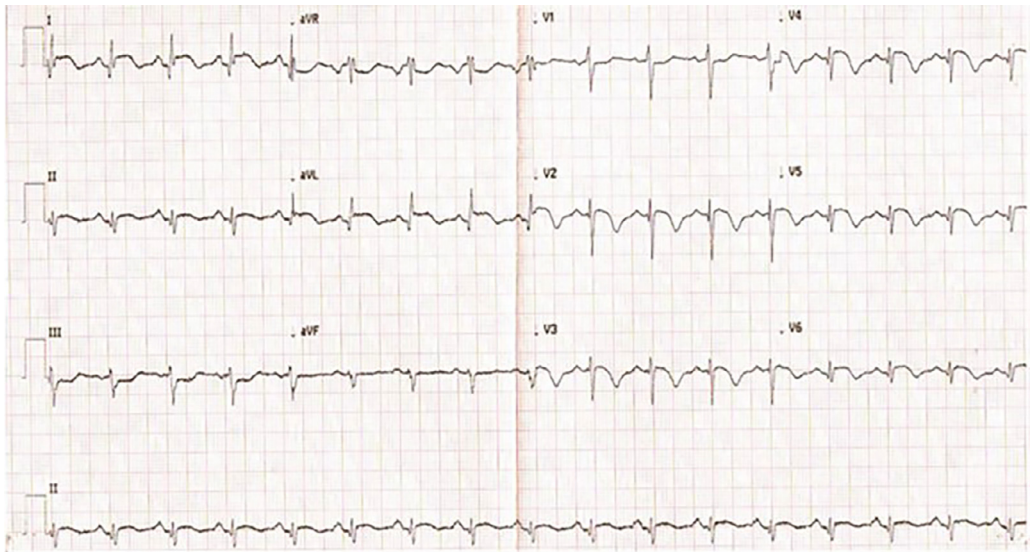

Fig. 1 – Twelve-lead ECG showed ST-segment elevation and inverted T waves in leads V2-6, D1 and aVL.

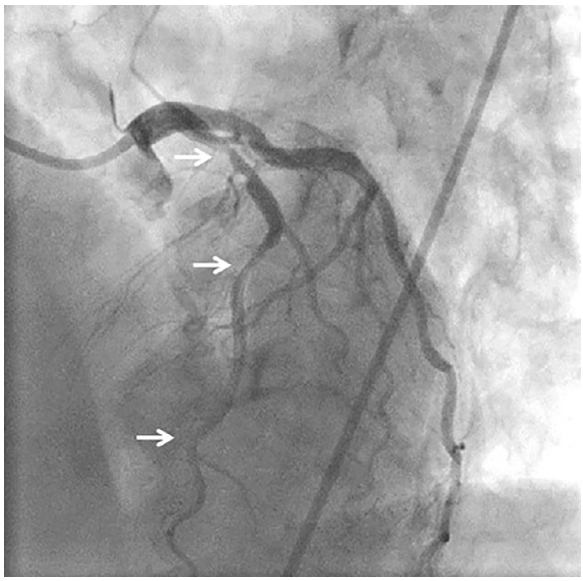

Fig. 2 – Proximal-LAD subtotal occlusion, 80% mid-LAD stenosis and distal-LAD vasospasm (arrow) (LAO/cranial view).

hyperviscosity, red blood cell effect on platelet interaction with the vessel wall and platelet dysfunction, iron deficiency, and coagulation system abnormalities [8,9].

On the other hand the JAK2V617F mutation is related to the quantity of immature platelets fraction (IPF) in patients with myeloproliferative neoplasms (MPN) [10,11]. This might be a contributing factor to the prothrombotic state of these patients. Thus, JAK2-mutated patients are more likely to suffer from a major thrombotic events [12].

It is evident that hydroxyurea treatment has favorable effect on MPN, but it also carries the risk of coronary vasospasm. In addition to vasospastic effects, the chemotherapy regimen also was thought to cause endothelium lesions [13–16].

The presence of IPF increases the risk for coronary events in term of existence of thrombotic risk. To now is known that cytoreductive treatment decreases vascular events, in particular at high-risk patients, but there doesn't exist any particular threshold of platelet counts showing safety toward free thrombosis events [17]. Thus, an increased tendency of blood clotting in PVR and the double effect of treatment with hydroxyurea may be followed with acute myocardial infarction.

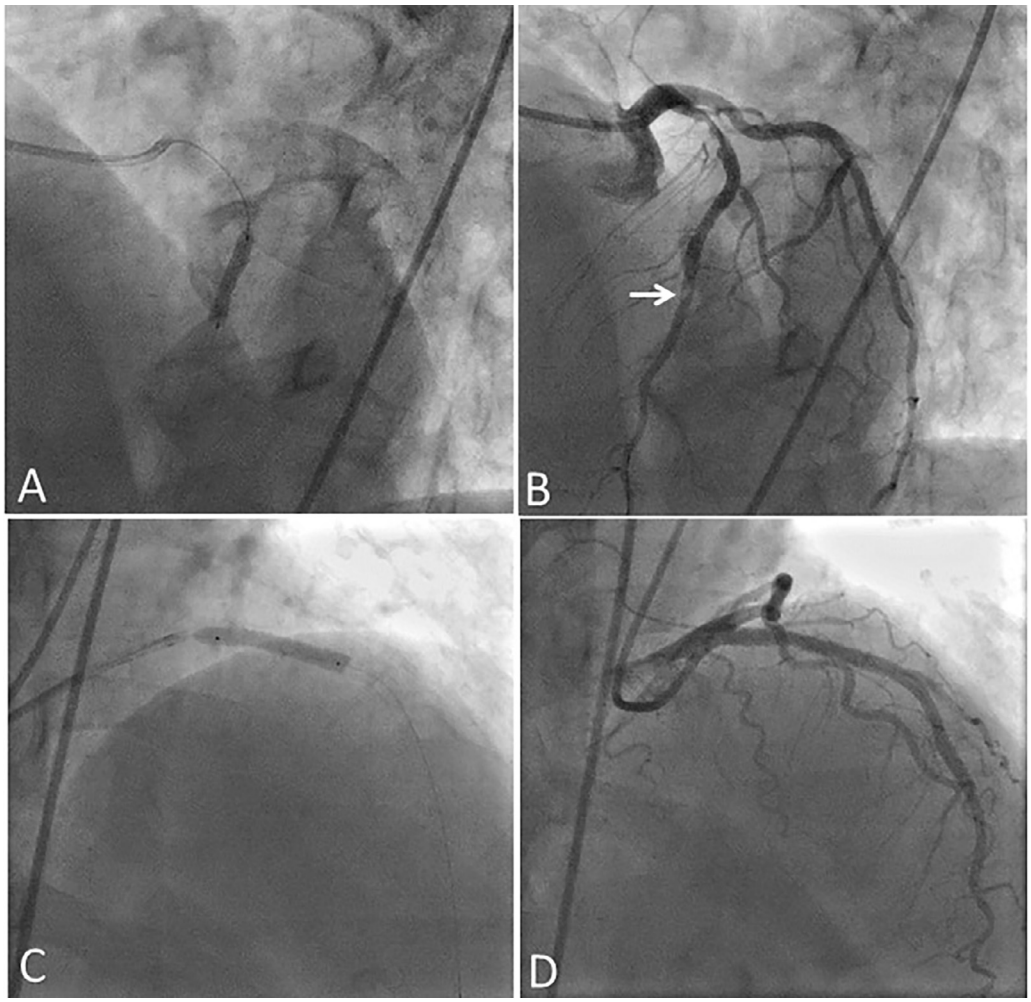

Fig. 3 – Percutaneous coronary intervention was performed with a drug-eluting stent in mid- and proximal-LAD, with small thrombotic mass after stenting in mid-LAD (arrow) (B).

Conclusion

We report a patient with polycythemia rubra vera who developed ST-segment elevation myocardial infarction while she was being treated with hydroxyurea. Hypercoagulable state of polycythemia rubra vera may be complicated with acute myocardial infarction, in addition to the vasospastic effect and endothelium lesions of hydroxyurea regardless its favorable effect as a standard therapy.

Author's contribution

XT analyzed and interpreted the patient data and was a major contributor in writing the manuscript. AB analyzed and interpreted the patient data and contributed in writing the manuscript. XK analyzed and interpreted the patient data. HÇ analyzed and interpreted the patient data. FK analyzed and interpreted the patient data. DK analyzed the data and contributed in interpreting them. All authors read and approved the final manuscript.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying image. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

REFERENCES

[1] James C, Ugo V, Le Couédic JP, Staerk J, Delhommeau F, Lacout C, et al. A unique clonal JAK2 mutation leading to constitutive signalling causes polycythaemia vera. Nature 2005;434(7037):1144–8.

[2] De Stefano V, Za T, Rossi E, Vannucchi A, Ruggeri M, Elli E, et al. Recurrent thrombosis in patients with polycythemia vera and essential thrombocythemia: incidence, risk factors, and effect of treatments. Haematologica 2008;93(3):372–80.

[3] Landolfi R, Di Gennaro L. Prevention of thrombosis in polycythemia vera and essential thrombocythemia. Hematologica 2008;93(3):331–5.

[4] Landolfi R, Di Gennaro L, Barbui T, De Stefano V, Finazzi G, Marfisi R, et al. Leukocytosis as a major thrombotic risk factor in patients with polycythemia vera. Blood 2007;109(6):2446–52.

[5] Okabe H, Sonoda S, Abe K, Doi H, Matsumura T, Otsuji Y. Acute myocardial infarction following sequential multi-vessel occlusion in a case of polycythemia vera. J Cardiol Cases 2019;20(4):111–14.

[6] Kälsch H, Wieneke H, Erbel R. Acute myocardial infarction in a patient with chronic myelocytic leukemia during chemotherapy with hydroxyurea. Herz 2010;35(6):420–2.

[7] Fattori A, de Souza RA, Saad ST, Costa FF. Acute myocardial infarction in sickle cell disease: a possible complication of hydroxyurea treatment. Hematol J 2005;5(7):589–90.

[8] Bahbahani H, Aljenaee K, Bella A. Polycythemia vera presenting as acute myocardial infarction: an unusual presentation. J Saudi Heart Assoc 2015;27(1):57–60.

[9] Kazma HK, Fakih M, Danash B, Mohammed M, Kazma H. ST-elevation myocardial infarction in a 56-year-old man with acute myeloid leukemia: a case report. Cureus 2023;15(12):e50230.

[10] Panova-Noeva M, Marchetti M, Buoro S, Russo L, Leuzzi A, Finazzi G, et al. JAK2V617F mutation and hydroxyurea treatment as determinants of immature platelet parameters in essential thrombocythemia and polycythemia vera patients. Blood 2011;118:2599–601.

[11] Sharma P, Gupta S, Patel P, Zhang Y, Peles S. Acute ST-segment elevation myocardial infarction as the first manifestation of essential thrombocytosis. Cureus 2019;11(2):e4032.

[12] Wang L, Mao X, Zheng J, Gu X, Yang J. A case of simultaneous acute cardio-cerebral infarction in a woman with essential thrombocythemia. J Int Med Res 2019;47(9):4557–61.

[13] Cheng CW, Hung MJ. Coronary spasm-related acute myocardial infarction in a patient with essential thrombocythemia. World J Cardiol 2011;3(8):278–80.

[14] Kälsch H, Wieneke H, Erbel R. Akuter Myokardinfarkt bei einem Patienten mit chronischer myeloischer Leukämie während Chemotherapie mit Hydroxyurea. Herz 2010;35:420–2.

[15] Borek G, Gebel F, Jenzer HR. Acute myocardial infarct following chemotherapy for testicular carcinoma: on the coronary toxicity of cytostatic agents. Schweiz Med Wochenschr 1991;121(11):385–9.

[16] Gorgulu S, Celik S, Tezel T. A case of coro- nary spasm induced by 5-fluorouracil. Acta Cardiol 2002;57(5):381–3.

[17] Carlos Hernandez-Boluda J, Gomez M. Target hematologic values in the management of essential thrombocythemia and polycythemia vera. Eur J Haematol 2015;94(1):4–11.