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Case Report

Recurrent unilateral dactylitis as an unusual presenting manifestation of polycythemia vera: a case report

Prashanth Gopathi*, Alekya Katti, Praveen Kumar Jangapalli

Department of Medicine, CMR Institute of Medical Sciences, Medchal, Telangana, India

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***Correspondence:**
Dr. Prashanth Gopathi,
E-mail: gopathiprashanth@gmail.com

ABSTRACT

Polycythemia Vera is a chronic myeloproliferative neoplasm characterised by clonal proliferation of erythroid, myeloid, and megakaryocytic cell lines. Typical clinical manifestations include headache, aquagenic pruritus, erythromelalgia, thrombotic events, and splenomegaly. However, atypical presentations may delay diagnosis. We report a rare case of recurrent unilateral dactylitis-like swelling as the presenting manifestation of polycythemia vera in a middle-aged woman. The patient was initially treated for presumed cellulitis because leucocytosis was interpreted in isolation, while the accompanying erythrocytosis and thrombocytosis were overlooked. Recurrent episodes prompted further evaluation, which demonstrated hypercellular bone marrow with panmyelosis and a positive JAK2 V617F mutation, confirming the diagnosis of polycythemia vera". This case highlights the importance of careful interpretation of complete blood counts, detailed clinical history taking, and maintaining suspicion for haematological disorders in patients presenting with recurrent inflammatory digit swelling.

Keywords: Polycythemia vera, Dactylitis, Myeloproliferative neoplasm, JAK2 V617F mutation, Thrombosis

INTRODUCTION

Myeloproliferative neoplasms (MPN) are clonal haematological disorders characterised by proliferation of myeloid lineages in the bone marrow.[1] The current WHO classification acknowledges four main subgroups of MPNs: Chronic myeloid leukaemia (CML), classical Philadelphia chromosome-negative MPNs (Polycythemia vera (PV), Essential thrombocytosis (ET), Primary myelofibrosis (PMF), CEL and MPN-Unclassified.[2]

In PV, ET, and PMF, erythroid and megakaryocytic hyperplasia predominate, and they are characterised by driver mutations that directly or indirectly activate the JAK-2 signalling network essential for the functioning of the erythropoietin and thrombopoietin receptors and also utilised by the granulocyte colony-stimulating factor (G-CSF) receptor.[3]

Although PV is rare disease, it is the most common classical Philadelphia chromosome-negative MPNs, and its global annual incidence is approximately 0.8-2.8 per 100,000 population, with a median age at diagnosis of around 60-64 years and a slight male preponderance 4. Common clinical features include fatigue (91%), insomnia (68%), aquagenic pruritus (pruritus on contact with water), splenomegaly (36%), erythromelalgia (29%), arterial thrombosis (16%), and venous thrombosis (9.4%).[5-7]

On the other hand, dactylitis is one of the hallmark manifestations in psoriatic arthritis8, sickle cell disease, acute lymphoblastic leukaemia, and infections.[9] Unilateral dactylitis as the presenting manifestation of a myeloproliferative neoplasm is exceedingly rare.

Although erythromelalgia has been described in polycythemia vera, isolated unilateral digit swelling may lead clinicians towards an infectious or inflammatory

diagnosis, thereby delaying definitive diagnosis. We present a case of polycythemia vera presenting unusually as recurrent unilateral dactylitis-like swelling, emphasising the importance of comprehensive haematological assessment in recurrent unexplained inflammatory presentations

CASE REPORT

A middle-aged female patient presented with recurrent painful swelling involving the digits of one hand. The first episode began suddenly and was initially presumed by the patient to be secondary to an insect bite. She did not seek medical attention because the swelling resolved spontaneously over four days.

Two weeks later, she experienced a second similar episode associated with pain and swelling of the same digits. She consulted a local practitioner, where cellulitis was suspected clinically. A complete blood count revealed an elevated total leukocyte count. However, the associated elevation of haemoglobin and platelet counts was not given significant consideration at that time.

The patient was treated with antibiotics and anti-inflammatory medications, following which the swelling resolved. After approximately two months, the patient experienced a third episode of painful unilateral digital swelling, initially suspecting an underlying metabolic disorder such as diabetes mellitus.

On evaluation, complete blood count demonstrated persistent trilineage haematological elevation, including erythrocytosis Hb-17gm/dl, (haematocrit-58.2%) leucocytosis (27300 cells/cubic mm) and thrombocytosis (7.06lakh/cubic mm).

The persistent elevation across all three cell lines raised suspicion of an underlying myeloproliferative disorder rather than a simple infectious process. Peripheral smear examination revealed increased red blood cell mass, leucocytosis and thrombocytosis, features suggestive of hyperactive marrow activity.

A Doppler study was performed to evaluate for vascular abnormalities and thrombotic phenomena, which revealed a chronic thrombus in intrahepatic portal vein branches. Ultrasound showed splenomegaly which further supported the suspicion of MPN.

Further haematological workup included bone marrow aspiration and biopsy, which demonstrated hypercellular marrow with pan myelosis, findings suggestive of a myeloproliferative neoplasm, findings consistent with polycythemia vera. Erythropoietin levels were sent and were found to be normal. MPN mutation panel including JAK2 mutation analysis was sent. “Molecular testing demonstrated the presence of the JAK2 V617F mutation, fulfilling the third major diagnostic criterion for polycythemia vera.”

The bone marrow aspirate was particulate and hypercellular, with a myeloid-to-erythroid (M:E) ratio of 1.4:1. Erythropoiesis was increased in number, showing normoblastic maturation. The myeloid series showed all stages of maturation. Megakaryocytes were increased in number, with hyperlobulated forms noted.

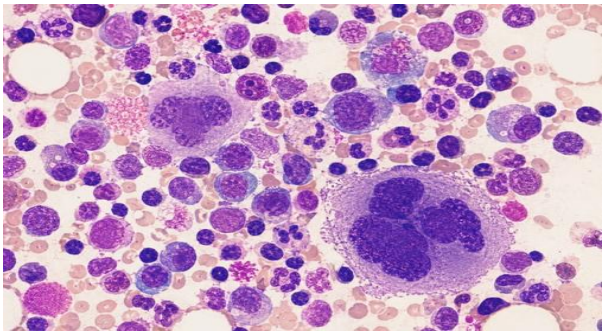

Figure 1: Hyper cellular marrow with pan myelosis, consistent with myeloproliferative neoplasm-morphology favouring polycythemia vera.

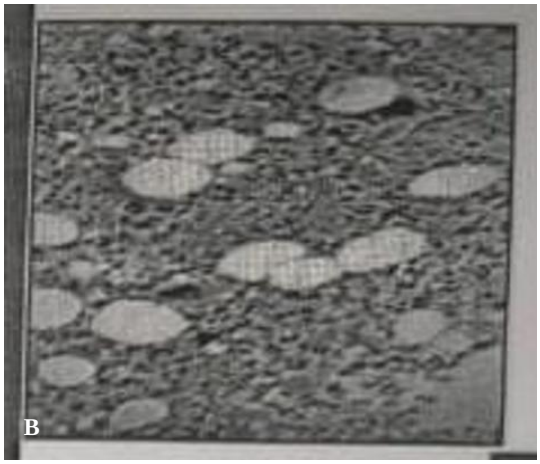

Figure 2 (A and B): The patient was counselled regarding the probable haematological aetiology of her recurrent symptoms and advised close haematology follow-up.

DISCUSSION

Myeloproliferative neoplasms are a heterogeneous group of clonal hematologic disorders characterized by proliferation of one or more myeloid cell lineages. They include

CML, PV, ET and PMF

Patients with PV most commonly present with headache, pruritis, fatigue: Most often recognized by the incidental discovery of a high Hb or Hct. Splenomegaly in 30-50% of cases may be the initial presenting sign in PV. Polycythemia vera commonly presents with symptoms related to hyperviscosity, thrombosis, or constitutional manifestations.[10] However, our patient presented with recurrent unilateral dactylitis-like digital swelling, an unusual and misleading clinical presentation that initially diverted attention from the underlying myeloproliferative disorder. Serum erythropoietin was within the normal range and therefore did not fulfil the minor criterion." During previous evaluations, the complete blood count was primarily interpreted in the context of leucocytosis, while the concomitant elevation of haemoglobin and platelet counts was overlooked. The recurrence of symptoms prompted a more comprehensive review of the haematological parameters, raising suspicion for an underlying myeloproliferative neoplasm. Subsequent investigations were favouring polycythemia vera.

The criteria for the diagnosis of PV have been updated in the WHO 2016[2]

Major criteria

Elevated haemoglobin/haematocrit or increased red cell mass Haemoglobin >16.5 g/dL in men or >16.0 g/dL in women or Haematocrit >49% in men or >48% in women or increased red cell mass. Bone marrow biopsy showing hypercellularity with trilineage proliferation (pan myelosis). Increased erythroid, granulocytic, and megakaryocytic proliferation. Pleomorphic mature megakaryocytes. Presence of a JAK2 mutation, JAK2 V617F mutation or JAK2 exon 12 mutation

Minor criterion

Subnormal serum erythropoietin (EPO) level.

Table 1: Clinical timeline.

Time	Event
Initial presentation	Sudden unilateral painful digital swelling; spontaneous resolution after 4 days
2 weeks later	Recurrence; diagnosed as cellulitis; antibiotics/NSAIDs given
~2 months later	Third episode; persistent/recurrent unilateral swelling
Third presentation	Hb 17 g/dL, Hct 58.2%, WBC 27,300/mm³, platelets 7.06 lakh/mm³
Further evaluation	Splenomegaly + portal venous thrombosis
Bone marrow	Hypercellular marrow with pan myelosis
Molecular testing	JAK2 V617F positive.
Final diagnosis	**Polycythemia vera**

Table 2: Diagnostic evaluation of polycythemia vera.

Investigation	Patient finding	Significance
Haemoglobin	17 g/dl	Elevated
Haematocrit	58.2%	Elevated
WBC	27,300/mm³	Leucocytosis
Platelets	7.06 lakh/mm³	Thrombocytosis
Peripheral smear	Erythrocytosis + leucocytosis + thrombocytosis	Trilineage elevation
Bone marrow	Hypercellular with pan myelosis	Major PV criterion
EPO	Normal	Minor criterion not fulfilled
JAK2	Positive	Major PV criterion
Spleen	Splenomegaly	Supports MPN
Portal vein	Chronic thrombosis	Thrombotic complication

Diagnosis of Polycythemia vera

Diagnosis requires all 3 major criteria, OR First 2 major criteria + the minor criterion. "The patient fulfilled all three major diagnostic criteria for polycythemia vera: elevated haemoglobin/haematocrit, bone marrow biopsy demonstrating pan myelosis, and detection of the JAK2 V617F mutation. Serum erythropoietin was within the normal range and therefore did not fulfil the minor criterion. The diagnosis of polycythemia vera was established based on the three major criteria."

Treatment of polycythemia vera (PV)

Modern management of PV includes phlebotomy, hydroxyurea, JAK2 inhibitors, aspirin, and supportive care. Phlebotomy is performed to maintain haematocrit below 45% and reduce thrombotic risk. High-risk patients should receive hydroxyurea (starting at 500 mg once or twice daily) along with aspirin and phlebotomy. Ruxolitinib (a JAK1/JAK2 inhibitor) is used in patients intolerant of or resistant to hydroxyurea and improves haematocrit control and spleen size.[11] The patient underwent two cycles of phlebotomy and started an aspirin 75mg once daily. The main goal of treatment is to control myeloproliferative activity and prevent thrombotic and haemorrhagic complications

This case highlights the importance of systematic interpretation of all components of the complete blood count rather than focusing on a single abnormality. Although CBC is a simple, inexpensive, and widely available investigation, it can provide crucial diagnostic clues when interpreted in conjunction with the clinical presentation. Failure to recognize subtle but persistent abnormalities may delay the diagnosis of potentially serious haematological disorders. Our case emphasizes that careful clinicopathological correlation remains fundamental to accurate diagnosis and timely management

CONCLUSION

This case highlights an unusual dactylitis-like presentation of polycythemia vera and emphasizes the importance of comprehensive interpretation of the complete blood count in patients with recurrent unexplained digital swelling. Persistent erythrocytosis, leucocytosis and thrombocytosis should prompt evaluation for an underlying myeloproliferative neoplasm. Early recognition of JAK2 V617F-positive polycythemia vera is particularly important because of the associated risk of thrombotic complications.

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