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Case Report

Internal Medicine Section

Polycythemia-linked Cerebral Venous Thrombosis with Right Fronto-Temporo-Parietal Infarct: A Case Report

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ABSTRACT

Cerebral Venous Thrombosis (CVT) is a rare form of stroke, often linked to prothrombotic states, inducing Philadelphia-negative Myeloproliferative Neoplasms (MPN) such as Polycythemia Vera (PV) and Essential Thrombocythemia (ET), which predispose via hyperviscosity and Janus Kinase-2 (JAK2)-mediated platelet activation. We report a unique case of CVT in a 65-year-old hypertensive woman, an atypical demographic compared to the usual population, with chronic headaches developed four days of left lower limb weakness. Neuroimaging revealed a superior sagittal and right transverse sinus thrombosis with a non-haemorrhagic fronto-temporo-parietal infarct. Laboratory evaluation demonstrated leukocytosis (14×103/μL), thrombocytosis (739×103/μL), alongside JAK2-V617F positivity. This constellation of findings places her phenotype between ET and PV. Reflex JAK2 testing enabled early provisional diagnosis of MPN-associated CVT despite the absence of marrow biopsy. Prompt anticoagulation with Low-Molecular-Weight Heparin (LMWH) was transitioned to apixaban 5 mg twice daily, and achieved marked neurological improvement by discharge. This case highlights the rarity of CVT in an older hypertensive patient with JAK2-positive MPN but no erythrocytosis, and underscores the importance of reflex mutation screening and timely anticoagulation in achieving favourable outcomes.

Keywords: Anticoagulants, JAK2 protein, Myeloproliferative neoplasms, Thrombocytosis

CASE REPORT

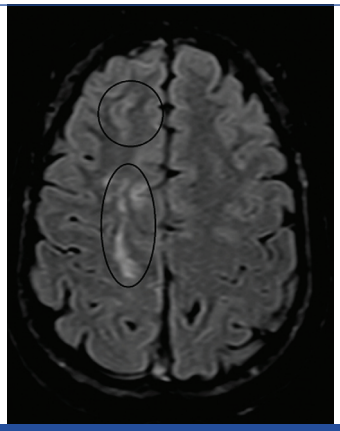
A 65-year-old female presented to the emergency department with four days of sudden, non-progressive left lower limb weakness, associated with slippage of footwear and difficulty in ambulation. The patient had a history of systemic hypertension for one year, which was well-controlled with Telmisartan 20 mg once daily. She did not report any neurological or systemic symptoms. She also reported intermittent dull occipital headaches for the past year, and relieved by over-the-counter analgesics (Tab. Aceclofenac and Paracetamol). She denied any history of fever, recent infections, gastrointestinal or urinary complaints, or respiratory or cardiovascular symptoms. On arrival, she was conscious and oriented. On examination, her Glasgow Coma Scale score of 15/15 (E4V5M6) and with stable vitals (pulse rate 80 beats per minute (pm), blood pressure 130/60 mmHg, respiratory rate 20 breaths pm, SpO_2 98% on room air, and glucose 89 mg/dL).

General and systemic examinations were unremarkable. Muscle tone and power were normal in all limbs, except that power was 4/5 and 3/5 in the left proximal and distal lower limbs. The plantar response was flexor and extensor on the right and left. Deep tendon reflexes were exaggerated on the left (3+ at the knee and ankle). Sensory and cerebellar examinations were within normal limits. Laboratory investigations revealed leukocytosis (14,000 cells/mm^3) and marked thrombocytosis (739,000/mm^3) [Table/Fig-1]. Other biochemical parameters were within normal limits. The Electrocardiogram showed a normal sinus rhythm with no ischaemic changes. Chest X-ray was unremarkable. MRI brain revealed CVT involving the superior sagittal and right transverse sinuses, with acute non-haemorrhagic infarction in the right high fronto-temporo-parietal region, including the centrum semiovale [Table/Fig-2]. MRV resulted in interrupted flow in a superior sagittal sinus [Table/Fig-3] and right transverse sinus [Table/Fig-4].

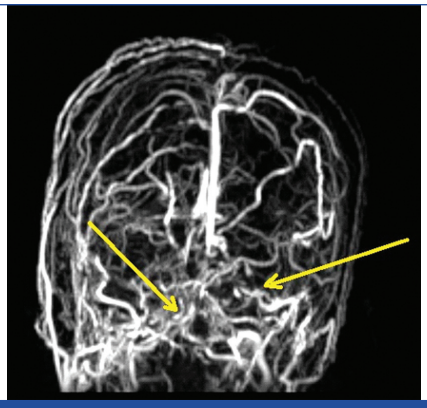
A provisional diagnosis of right CVT with an acute non-haemorrhagic infarct was made. Treatment was initiated with therapeutic anticoagulation (Inj. Enoxparin 40 mg subcutaneously for four days), statins (Atorvastatin 20 mg), antihypertensives (Telmisartan 20

Investigations	Results	Normal range
Total count (cells/mm^3)	14000	4000 - 10000
Neutrophils (%)	73.5	40 - 80
Lymphocytes (%)	12.8	20 - 40
Eosinophils (%)	7.4	1 - 6
Basophils (%)	0.2	~2
Monocytes (%)	6.1	2 - 10
Band cells (%)	0	-5
Total red blood cell count (million/mm$_3$)	7.22	3.8 - 4.8
Packed cell volume (%)	48.9	36 - 46
Mean corpuscular volume (fL)	67.7	83 - 101
Mean corpuscular haemoglobin (pg)	20.3	27 - 32
Mean corpuscular haemoglobin concentration (%)	30	31.5 - 34.5
Platelet count (cells/mm$_3$)	739000	150000 - 450000
Haemoglobin (g/dL)	14.7	12 - 15
Red cell distribution width (%)	24.5	11.6 - 14

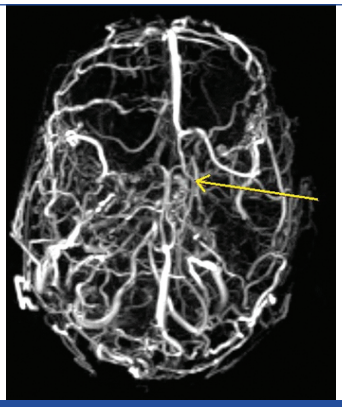
[Table/Fig-1]: Laboratory investigations of the patient.



[Table/Fig-2]: Multifocal Fluid-Attenuated Inversion Recovery (FLAIR) hyperintensities in the right fronto-parietal lobe (black circle) p/o infarct.



[Table/Fig-3]: Magnetic Resonance Venography (MRV) Maximum Intensity Projection (MIP) images showing interrupted flow in the superior sagittal sinus (yellow arrows).



[Table/Fig-4]: Magnetic Resonance Venography (MRV) Maximum Intensity Projection (MIP) images showing interrupted flow in the right transverse sinus (red dotted arrows).

mg), and supportive measures (oxygen, Paracetamol 650 mg and Pantoprazole 40 mg). Neurology consultation concurred with the management and advised against antiplatelet therapy in the absence of arterial involvement. The patient was monitored in the intensive care unit, where she remained neurologically stable without deterioration in sensorium or emergence of new deficits. She was subsequently shifted to the high-dependency unit for continued observation. JAK2-V617F testing was prompted by persistent thrombocytosis to evaluate MPNs (ET/PV) and was positive, confirming an MPN. Bone marrow biopsy is necessary to distinguish ET from PV, yet it could not be performed due to the financial constraints of the patient. Later, on neurology follow-up for two weeks, the patient was transitioned to oral anticoagulation (lifelong) with apixaban 5 mg twice daily and discharged in a haemodynamically stable condition with significant symptomatic improvement. At 30-day follow-up, she was ambulating independently with near-complete resolution of weakness, corresponding to a modified Rankin Scale (mRS) score of 1. Later, they were referred to a haematologist for follow-up upon request to another hospital due to their convenience.

DISCUSSION

In our patient, a 65-year-old hypertensive woman presenting with acute left lower limb weakness and headache, the imaging confirmed CVT involving the superior sagittal and right transverse sinuses, with a right fronto-temporo-parietal infarct. Marked thrombocytosis prompted the workup for an MPN, and JAK2-V617F mutation analysis ultimately established a diagnosis of MPN. She was initially anticoagulated with LMWH and later transitioned to apixaban, with significant clinical improvement upon discharge.

CVT, defined by thrombosis of cerebral veins and dural sinuses, represents only 0.5-1% of all strokes with an incidence of 1.3-1.6 per 100,000 person-years [1,2]. Although historically seen in younger women linked to pregnancy, puerperium, or oral contraceptives, recent evidence demonstrates a growing proportion of older adults with systemic or haematologic disorders [1,2]. With advances in imaging and management, mortality has fallen from 10-15% to <10% [1,2].

Philadelphia-negative MPN, such as ET or PV, are now recognised causes of CVT in patients with unexplained thrombocytosis or erythrocytosis and JAK2, CALR or MPL mutations [3-5]. They induce hypercoagulability through hyperviscosity, platelet dysfunction, JAK2-V617F-mediated endothelial activation, which contributes to thrombotic complications, including CVT [5-7]. Clinically, JAK2-V617F mutation has been identified in 65-97% of PV specimens, 23-57% of ET and 35-57% of myelofibrosis specimens, aiding distinction from reactive disorders [5,6]. Magnetic Resonance Imaging or Venography (MRI/MRV) is crucial for rapid diagnosis, and early anticoagulants, historically heparin to Vitamin K Antagonists (VKA), but Direct Oral Anticoagulants (DOACs) like apixaban remain the cornerstone of CVT treatment, even with haemorrhagic transformation [3,4,8-10].

The MPNs encompass a spectrum of clonal haematopoietic stem cell disorders, most commonly ET and PV, both frequently driven by JAK2-V617F-driven clonal disorders that cause sustained thrombocytosis in ET or erythrocytosis in PV [9]. Both entities share a predisposition to thrombotic complications due to multiple interrelated mechanisms, including elevated cell counts, blood hyperviscosity, leukocyte-mediated inflammation, and JAK2-mediated endothelial dysfunction and platelet activation [5-7,9].

In our patient, leukocytosis and marked thrombocytosis, the hyperviscosity and prothrombotic milieu were evident, although her Hb was within normal limits (as no documented erythrocytosis), combined with hypertension. All these reports mimic the laboratory profile reported by Gangat N et al., who found that ET and PV patients presenting with CVT often exhibited leukocyte counts >12000/mm^3 and platelet count >600,000/mm^3 at diagnosis [3]. In their multicentre series of 74 MPN-associated CVT cases, 42% were attributable to ET and 35% to PV, where the mean leukocyte count among PV-CVT patients was 13,500/mm^3, like our patient's 14,000/mm^3, suggesting that leukocytosis is a strong predictor of thrombosis irrespective of whether ET or PV is ultimately diagnosed [3]. This similarity underscores leukocytosis as a strong predictor of thrombosis, irrespective of whether ET or PV is ultimately diagnosed.

Jianu DC et al., described 18 PV-related CVT cases in which the mean haematocrit was 56% and mean platelet count 820,000/mm3 and all were JAK2-V617F positive [4]. By contrast, ET-related CVT patients in Song J et al., review demonstrated mean platelet counts of 950,000/mm^3 but haematocrit within normal range [5]. Our patient's lack of documented erythrocytosis but pronounced thrombocytosis and leukocytosis places her phenotype between the typical ET and PV presentations, underscoring the overlapping pathophysiology of JAK2-positive MPNs in CVT.

When marrow biopsy is unattainable, as in our patient, due to financial constraints, MPN subtype must be inferred from clinical and laboratory surrogates (Hb, haematocrit, leukocyte and platelet count) plus JAK2-V617F allele burden [9]. Tefferi A et al., recommend labelling such cases as "MPN-unclassified" when JAK2 positivity coexists with thrombocytosis and leukocytosis but normal haematocrit, this provisional diagnosis guides management since both ET and PV carry high thrombotic risk [9].

Our provisional MPN (rather than definitively ET or PV) diagnosis rested on JAK2-V617F positivity and cytoses, without marrow confirmation. Song J et al., recommend JAK2 screening for CVT patients with >600,000/mm^3 or haematocrit >50%, observing that 90% of their reviewed MPN-CVT cases were identified via JAK2 testing before marrow confirmation [5]. Gangat N et al., reported a median three-day interval to JAK2 testing versus eight days to marrow, delaying cytoreduction [3]. In our patient, JAK2 testing was performed on the day of admission and returned positive within 48

hours, allowing prompt classification as MPN-CVT despite missing marrow data. Comparatively, Jianu DC et al., noted that deferring biopsy until after JAK2 positivity prolonged therapy initiation by up to five days [4]. By contrast, our institution's protocol for reflex JAK2 testing in unexplained thrombocytosis enabled a more rapid provisional diagnosis, allowing earlier intervention. Absent marrow morphology, as an approach consistent with the European Leukaemia Net guidance using serial Hb/haematocrit and clinical parameters to distinguish ET from PV and tailor phlebotomy, hydroxyurea or aspirin therapy [9].

Treatment of MPN-CVT necessitates a dual approach: anticoagulation to address the acute thrombotic event and cytoreductive measures to control the underlying haematologic disorder. Immediate LMWH upon CVT diagnosis, reduce mortality and improve functional outcomes [10]. Once stable and with no contraindications, transition to a DOAC is appropriate, with meta-analyses demonstrating efficacy and safety on par with VKA in MPN-CVT [8,10]. Cytoreduction should be phenotype-specific, where hydroxyurea for predominant thrombocytosis and phlebotomy to maintain hematocrit <45% if erythrocytosis present and low-dose aspirin may be added if bleeding risk is low [9]. Close blood counts monitoring is essential, with interferon or alternative cytoreductive agents considered, if hydroxyurea fails to normalise leukocyte or platelet levels [9].

Prognosis in MPN-CVT depends on timely recanalisation, sustained haematologic control, and thrombosis prevention. MPN-CVT carries a 20% five-year recurrence risk versus 8% in non-MPN-CVT maintaining haematocrit <45% and normal platelet counts significantly lowers the risk [1,4]. With prompt anticoagulation plus cytoreduction, over 80% of MPN-CVT patients achieve an mRS of 0-1 at six months [4,9], mirroring outcomes in our patient who improved from a left lower limb weakness to ambulating independently. Persistent JAK2-V617F positivity and hypertension portend ongoing thrombotic risk, warranting lifelong anticoagulation, serial neuroimaging at six and 12 months, and quarterly hematologic follow-up to adjust therapy as needed [2,9]. In our patient, functional recovery was quantified at 30-day follow-up with an mRS score of 1, consistent with favorable outcomes reported in MPN-associated CVT when anticoagulation is initiated promptly. This outcome parallels a meta-analysis showing DOACs are as effective and safe as VKAs in CVT. Kang W et al., further demonstrated favourable outcomes with LMWH transition to DOACs in cancer-associated thrombosis, supporting our management approach in MPN-associated CVT [10].

CONCLUSION(S)

This case highlights the rare occurrence of CVT in an older hypertensive patient with JAK2-V617F positive for MPN but without erythrocytosis, placing her phenotype between ET or PV. Reflex JAK2 testing enables early provisional diagnosis despite the absence of marrow biopsy, allowing timely initiation of anticoagulation. The patient achieved significant neurological recovery (mRS 1 at 30 days), underscoring the importance of rapid neuroimaging, mutation screening, and integrated haematology-neurology management. Long-term care requires indefinite anticoagulation, cytoreduction therapy as indicated, serial neuroimaging to prevent recurrence.

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