Case Report

Papilledema with Lateral Rectus Palsy in a young patient with PolycythemiaVera : A case report

Abstract: A 27-year-old man with persistent headache, nausea and double vision for two weeks was found to have right lateral rectus palsy and Grade 4 Papilledema. Further, blood investigations showed Polycythemia Vera and imaging confirmed cerebral venous thrombosis. Patient was managed conservatively and followed up till edema and diplopia subsided. This case highlights the importance of managing neurological symptoms and underlying hematological conditions to prevent complications like venous thrombosis in young adults.

Key-words – Papilledema, Lateral rectus palsy, Polycythemiavera, cerebral venous thrombosis

Introduction: Polycythemiavera (PV) is a myeloproliferative neoplastic condition characterized by unregulated red blood cell production, resulting in high red blood cell mass (RBC). [1]

PV affects all demographics age groups and has no known familial predilection. The median age of diagnosis is about 60 years. While it primarily affects older persons, younger patients may potentially get the condition. There are fewer insights on the ocular manifestations of polycythemia, however, transient visual disturbances [amaurosis fugax], scintillating scotoma, ophthalmic migraine, papilledema, Retinal vein occlusion can occur. [2,3]

Case report: A 27-year-old male presented with continuous, generalised headache associated with nausea and diplopia which was binocular, uncrossed, horizontally separated for 2 weeks.

On ocular examination, best corrected visual acuity was 20/20 in both eyes (BE). The colour vision was normal in BE. In primary gaze,

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BE showed alternating esotropia. Slit lamp biomicroscopy was within the normal limits. The pupils were reacting normally to light and did not show any afferent defect. On fundus examination, BE media were clear and optic disc showed florid disc edema, hyperemic with blurred disc margins with obliteration of cup (Frisen grade 4). Paton lines were present. Dilated tortuous veins were also seen around the disc. Foveal reflex was present in BE. On orthoptic evaluation, Hirschberg test showed 15˚ esotropia in right eye (RE) and limitation in abduction RE (Fig 1). Visual field assessment showed enlargement of blind spot BE. Intraocular pressure was 18 mmHg in BE.

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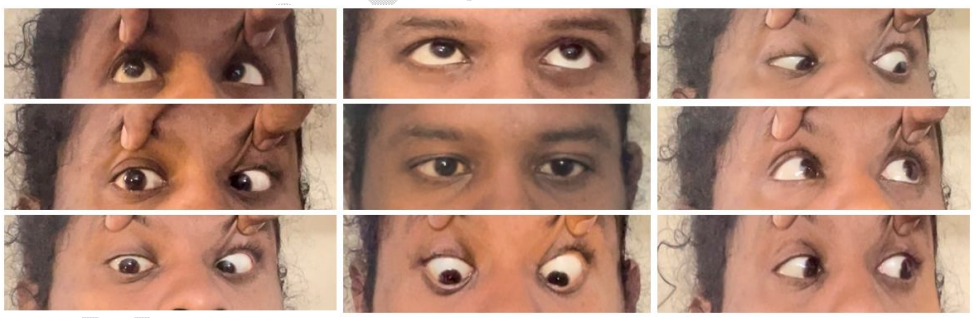
Fig 1 – Limitation of abduction in RE in dextroversion and dextroelevation



Haematological investigations showed raised haemoglobin (18.5 gm%), total count (15.88), hematocrit (54%), PT (30.3), INR (2). Erythropoietin levels were decreased. Peripheral smear showed normocytic normochromic RBCs with severe eosinophilia. The individual had not been exposed to high altitude, was a non-smoker and had no history of medical illness. A diagnosis of Polycythaemia Vera was made. Magnetic Resonance Imaging (MRI) Brain with

Magnetic Resonance venogram (MRV) revealed thrombus in posterior part of the superior sagittal sinus extending to left transverse and sigmoid sinus. (Fig 2)

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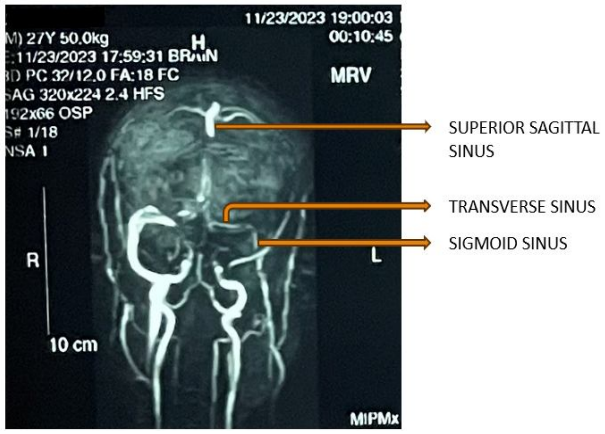

Fig 2 – Thrombus in superior sagittal sinus

Neurology consultation was obtained and patient was initiated on Injection Low Molecular Weight Heparin (LMWH) 60mg subcutaneous stat, Tab. Acetazolamide 250mg twice a day and was overlapped with oral anticoagulants in the form of Tab. Nicoumalone at 2mg per day. Regular follow ups were done. At 3rd week, patient was asymptomatic with resolving papilledema with full EOMs.

Discussion: Swelling of the optic disc due to increased intracranial pressure (ICP) is termed as papilledema. Patients with papilledema may experience headache, nausea and vomiting, transient visual obscuration, diplopia and pulsatile tinnitus. The increased pressure compresses the nerve fibres at the level of the optic disc leading to the disruption of the normal axoplasmic flow thus causing the swelling of the nerve fibres at the optic disc. The most important causes of papilledema can be brain tumours, cerebral edema, meningitis, and cerebral venous thrombosis. [5]

When intracranial pressure is elevated, the sixth cranial nerve is stretched leading to a false localizing sign, indicating abducens nerve palsy presenting as horizontal diplopia. [4,5]

PV is characterised by clonal proliferation of myeloid cells. The excessive production of RBCs increases blood viscosity, leading to complications such as thrombosis including Cerebral venous thrombosis (CVT), elevated risk of stroke, myocardial infarction, and other cardiovascular events. CVT is a rare presentation of PV in clinical practice. Symptoms of PV are often insidious during the onset, and a lack of specific clinical manifestations of CVST may lead to delayed diagnosis. PV can also cause additional symptoms and complications, including headaches, dizziness, aquagenic pruritus, and splenomegaly. [1-6]

CVT is a rare and potentially fatal condition. It is thought to be an underdiagnosed illness that primarily affects young and middle-aged people. Although non-specific, but around 90% of the patients presents with headache. Elevated venous pressure can either directly increase pressure in venules and capillaries or indirectly elevate them by raising ICP resulting in headaches, seizures, focal neurological deficits, and papilledema. [6-9]

MRI with MRV are highly sensitive techniques for detecting CVT as well as for raised ICP which helps in identifying underlying causes and thus preventing severe complications optic atrophy, permanent visual field defects and blindness.[8]

The primary approach is anticoagulation to reduce thrombotic occlusion of venous outflow. Management of papilledema, headaches, or nerve palsies focuses on lowering pressure and preserving vision.[8]

Conclusion: Prompt diagnosis and intervention of papilledema are essential to prevent lasting vision damage and address increased intracranial pressure. Understanding the link between polycythemia and cerebral venous thrombosis is crucial for effective management and risk mitigation and hereby improving patient outcomes.

Patient Consent Declaration:

We have obtained all necessary patient consent forms, authorizing the use of their images and clinical details in the journal. Patients understand that their identities will be protected, with names and initials omitted, though complete confidentiality cannot be guaranteed.

Comment [PNLP23]: You have addressed informed consent.

References

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Comment [PNLP25]: Reference and citation style match.

Reporting Checklist for Case Report Or Case Series.

INSTRUCTIONS TO AUTHORS

Complete this checklist by entering the page numbers from your manuscript where readers will find each of the items listed below.

Your article may not currently address all the items on the checklist. Please modify your text to include the missing information. If you are certain that an item does not apply, please write "n/a" and provide a short explanation.

Upload your completed checklist as an extra file when you submit to a journal.

In your methods section, say that you used the CAREreporting guidelines, and cite them as:

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		Reporting Item	Page Number
Title			
	#1	The area of focus and “case report” should appear in the title	
Keywords			
	#2	Two to five key words that identify topics in this case report	
Abstract			
Introduction	#3a	What is unique and why is it important?	
	#3b	The patient’s main concerns and important clinical findings.	
	#3c	The main diagnoses, interventions, and outcomes.	
Conclusion	#3d	What are one or more “take-away” lessons?	
Introduction			

	#4	Briefly summarize why this case is unique with medical literature references.
Patient information		
	#5a	De-identified demographic and other patient information.
	#5b	Main concerns and symptoms of the patient.
	#5c	Medical, family, and psychosocial history including genetic information.
	#5d	Relevant past interventions and their outcomes.
Clinical findings		
	#6	Relevant physical examination (PE) and other clinical findings.
Timeline		
	#7	Relevant data from this episode of care organized as a timeline (figure or table).
Diagnostic assessment		
	#8a	Diagnostic methods (PE, laboratory testing, imaging, surveys).
	#8b	Diagnostic challenges.
	#8c	Diagnostic reasoning including differential diagnosis
	#8d	Prognostic characteristics when applicable
Therapeutic Intervention		

	#9a	Types of intervention (pharmacologic, surgical, preventive).
	#9b	Administration of intervention (dosage, strength, duration)
	#9c	Changes in the interventions with explanations.
Follow up and outcomes		
	#10a	Clinician and patient-assessed outcomes when appropriate
	#10b	Important follow-up diagnostic and other test results.
	#10c	Intervention adherence and tolerability (how was this assessed)?
	#10d	Adverse and unanticipated events.
Discussion		
	#11a	Strengths and limitations in your approach to this case.
	#11b	Discussion of the relevant medical literature.
	#11c	The rationale for your conclusions.
	#11d	The primary “take-away” lessons from this case report.
Patient perspective		
	#12	The patient can share their perspective on their case
Informed consent		
	#13	The patient should give informed consent.

Subject: Submission of Peer Review for Manuscript Titled "Papilledema with Lateral Rectus Palsy in a Young Patient with Polycythaemia Vera: A Case Report"

Dear Editorial Board,

I hope this letter finds you well. I am writing to submit my peer review of the manuscript titled **"Papilledema with Lateral Rectus Palsy in a Young Patient with Polycythaemia Vera: A Case Report"** for your consideration.

The manuscript presents a compelling case of a 27-year-old male diagnosed with **Polycythaemia Vera (PV)** and its association with **papilledema** and **lateral rectus palsy**. The case report offers valuable insights into managing neurological symptoms and underlying haematological conditions to prevent severe complications such as **cerebral venous thrombosis**. The discussion elaborates well on the mechanisms leading to papilledema and the interventions to manage elevated intracranial pressure.

My Feedback on the Manuscript:

- **Strengths**:
  - The manuscript emphasizes the importance of recognizing neurological symptoms early, especially in younger patients, where conditions like PV may not be a primary diagnostic consideration.
  - It provides clear diagnostic pathways and a step-by-step therapeutic approach, contributing to the understanding of rare presentations of polycythaemia.
- **Areas of Improvement**:
  1. **Abstract**: The conclusion of the abstract could better highlight the broader implications of managing haematological disorders and the importance of multidisciplinary care.
  2. **Timeline**: Adding a timeline of symptoms, diagnosis, and treatment interventions in a table format could improve clarity.
  3. **Patient Perspective**: Including the patient's perspective on the case and recovery would provide additional context and fulfil the CARE guidelines' reporting criteria.
  4. **References**: Some recent references could be updated to strengthen the discussion, especially around the latest therapeutic protocols for managing cerebral venous thrombosis in PV patients.

Reporting Checklist:

I have also reviewed the manuscript in accordance with the CARE guidelines, as outlined in the Reporting Checklist for Case Report or Case Series. The detailed checklist is attached with specific page references for each reporting item.

In conclusion, the case report is well-documented and provides important clinical insights. With some minor adjustments, I believe this manuscript could contribute significantly to the medical literature on rare neurological complications associated with Polycythaemia Vera.

Thank you for the opportunity to review this manuscript. Please feel free to reach out if you need further clarification on my feedback.

Sincerely,

DR.NAMRATA SRIVASTAVA
ASSISTANT PROFESSOR
ERA UNIVERSITY, LUCKNOW, U.P.INDIA