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Papilledema with Lateral Rectus Palsy in a Young Patient with Polycythemia Vera: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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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 underscores the critical need for early recognition and comprehensive management of neurological symptoms, particularly in the presence of underlying hematological disorders such as Polycythemia Vera. A multidisciplinary approach, involving hematological and neurological expertise, is essential to address both the immediate and systemic complications effectively.

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Keywords: Papilledema; lateral rectus palsy; polycythemia vera; cerebral venous thrombosis.

1. INTRODUCTION

Polycythemia vera (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 [1,2].

2. CASE PRESENTATION

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, 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.

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).

Neurology consultation was obtained and patient was initiated on Injection Low Molecular Weight Heparin (LMWH) 60 mg subcutaneous stat, Tab. Acetazolamide 250 mg 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.

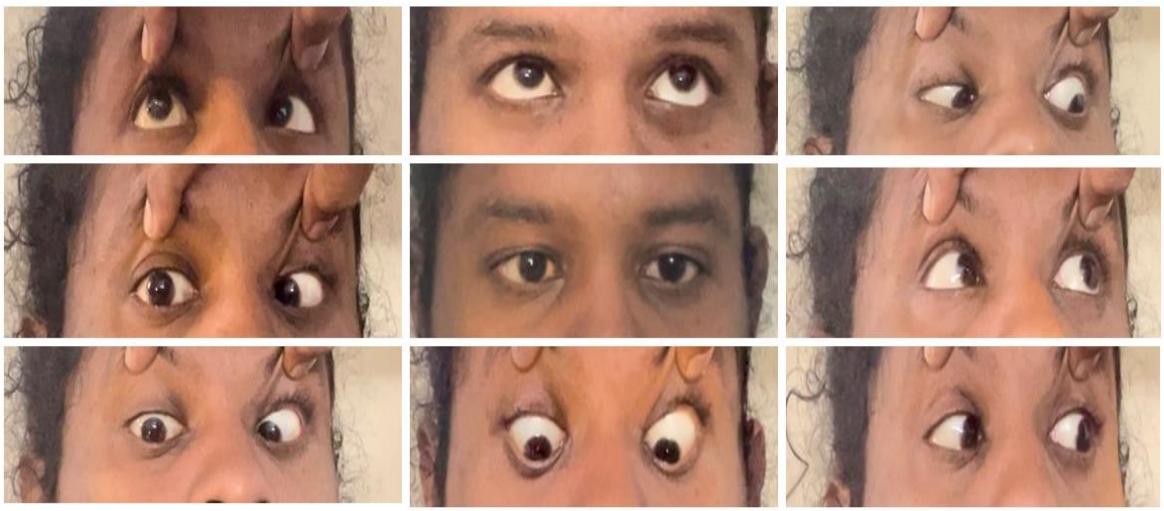

Fig. 1. Limitation of abduction in RE in dextroversion and dextroelevation

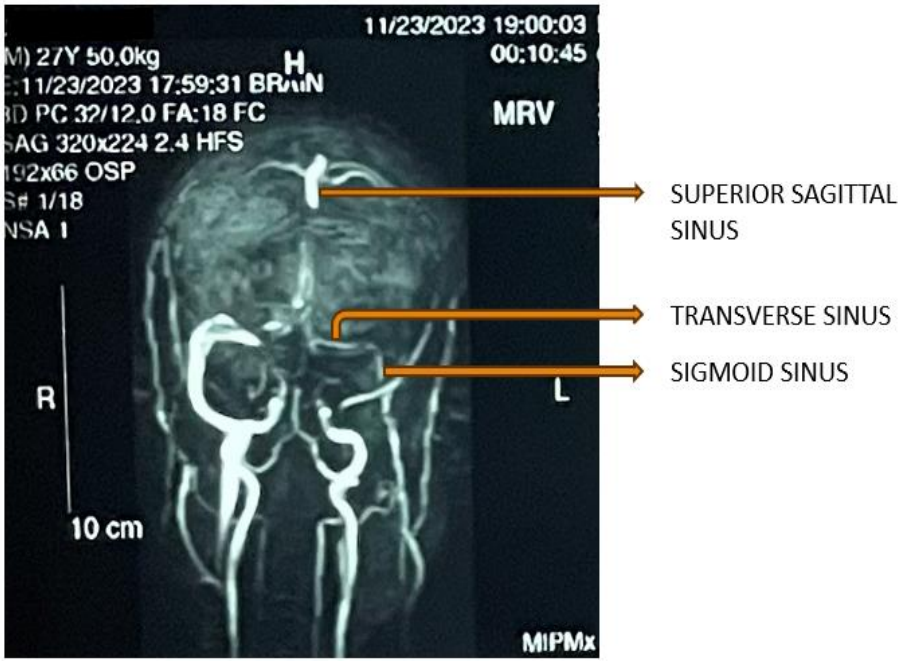

Fig. 2. Thrombus in superior sagittal sinus

3. 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 [3,4].

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 [2,3].

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 [3,1,5].

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 [5-8].

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 [7].

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 [7,9].

4. CONCLUSION

Timely diagnosis and intervention in cases of papilledema are vital to prevent long-term sequelae, including permanent visual impairment

and increased intracranial pressure. This case further highlights the necessity of recognizing the association between Polycythemia Vera and cerebral venous thrombosis, as both conditions can present with subtle but critical symptoms. Early imaging, combined with prompt therapeutic anticoagulation, can significantly reduce the risk of irreversible damage. Comprehensive management is essential to improving patient prognosis.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

CONSENT

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.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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