Polycythemia vera presenting with bilateral papilledema: A rare case report

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A 45-year-old male patient presented with gradual onset of headache, vomiting and blurring of vision of 28 days duration. Ophthalmological examination revealed normal anterior segment and pupillary reflex. No abnormality was detected in the vitreous. Optic disc showed features of advanced papilledema with normal macula and retinal periphery in both eyes. Visual acuity was 20/200 in the right eye and counting fingers close range in the left eye. Non-contrast computed tomography of brain was normal and magnetic resonance imaging showed sagittal sinus thrombosis without any evidence of venous infarction or intracranial mass. Routine hematological investigations revealed increased hemoglobin level, packed cell volume and leucocytosis. Further investigation revealed increased Vitamin B12 and decreased serum erythropoietin. A diagnosis of polycythemia vera was made from the above findings. This case is being presented for the rarity of association of polycythemia vera with bilateral advanced papilledema due to sagittal sinus thrombosis.

Key words: Papilledema, polycythemia vera, sagittal sinus thrombosis

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Polycythemia vera, also commonly called polycythemia rubra vera, is a chronic clonal disorder characterized by elevated red cell mass, normal arterial oxygen saturation, leucocytosis, thrombocytosis and splenomegaly.[1] The signs and symptoms of polycythemia vera are due to slowing of blood flow and formation of thrombosis as a result of increased viscosity.

Cerebral venous thrombosis is a rare and life-threatening condition that should be considered in all individuals with unusual and intractable headache. Young adults and children are typically affected and the superior sagittal sinus is the most commonly affected. Septic sagittal sinus thrombosis may develop in relation to infection of ear or paranasal sinuses or to bacterial meningitis. Etiological factors for aseptic sagittal sinus thrombosis are head injury, tumors, oral contraceptive pill, pregnancy and coagulopathies.[2] We report a rare case of bilateral papilledema with thrombosis of posterior part of sagittal sinus due to polycythemia vera.

Case Report

A 45-year-old male presented with sudden onset of headache, vomiting and blurring of vision of 28 days duration. Past medical history was insignificant. Detailed ophthalmological examination revealed normal anterior segment findings. The pupillary reaction to both direct and consensual light reflex was normal. Relative afferent pupillary defect was not detected. Visual acuity in both eyes was reduced to 20/200 in the right eye and counting fingers close range in the left eye. Visual field could not be tested due to low vision. However, fundoscopic examination revealed a clear optical media with significant advanced papilledema in both eyes [Fig. 1]. The macula, fovea and periphery of fundus in both eyes were normal. Non-contrast computed tomography (CT) scan of the brain was found to be normal, though magnetic resonance imaging (MRI) revealed features of sagittal sinus thrombosis without any evidence of venous infarction or mass lesion [Fig. 2]. Routine hematological and biochemical investigation revealed the following significant features: Differential count- (Neutrophil 90 Eosinophil 02 Basophil 00 Lymphocyte 08 Monocyte 00); Total leucocyte count-(164,000/mm^3), Hemoglobin- 18 gm%; Packed cell volume-56% total platelet count-4.4 lakhs/mm^3; prothrombin time -25.55sec and INR (International normalized ratio)-2.04.

High level of hemoglobin, packed cell volume and a raised platelet count with leucocytosis aroused the suspicion of polycythemia vera and on further hematological investigation, it was found that serum Vitamin B12 level was increased (796.0 pg/ml) and serum erythropoietin level was decreased (2.19 U/L). Taking all the above parameters into account and in consultation with a hematologist, the diagnosis of polycythemia vera with bilateral papilledema was made.

The patient was treated initially with subcutaneous injection of Heparin (40 mg twice daily) and acetazolamide 250 mg orally twice daily. Therapeutic venesection was done followed by systemic hydroxyurea (500 mg four times daily to start followed by 500 mg twice daily). Headache and vomiting subsided within 10 days of initiation of treatment. On follow-up visit after three weeks, a marked recovery of vision was observed in the right eye with visual acuity 20/30 and no improvement was seen in the left eye due to post papilledema optic atrophy.

Discussion

Cerebral venous thrombosis is a rare clinical entity. Septic sagittal sinus thrombosis may develop in relation to infection of ear or paranasal sinuses or to bacterial meningitis. Etiological factors for aseptic sagittal sinus thrombosis are head injury, tumors, oral contraceptive pill, pregnancy and coagulopathies. It affects sagittal sinus (71%) more often than any other dural sinuses because of its high position, low pressure and slow flow.[2,3] The symptoms and signs of sagittal sinus thrombosis are related to the site of occlusion. It is minimal in occlusion of the anterior part of the sagittal sinus and the symptoms are headache and paresthesia. When the posterior part of the sagittal sinus is affected, the neurological consequences are devastating.[3] Such patients complain of severe headache which is often worse at night or when arising and may be increased by bending or performing the Valsalva maneuver. In many patients, vision becomes transiently obscured. These episodes usually affect both eyes, last 10 to 15 seconds, sometimes precipitated by bending, coughing or standing. Constant blurring of vision, diplopia, tinnitus and syncopal spells may occur, but less frequently.[4] Decreased visual acuity in one or both eyes, enlarged blind spots, concentric decrease in visual fields can also be present in chronic papilledema.[4,5]

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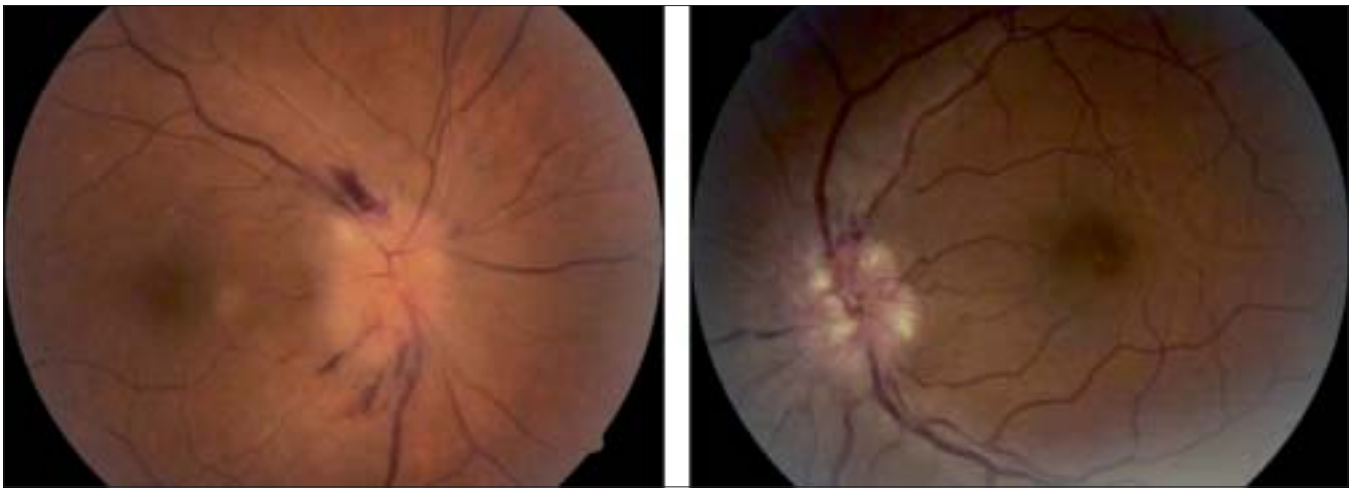

Figure 1: Fundus showing advanced papilledema in the right eye and left eye

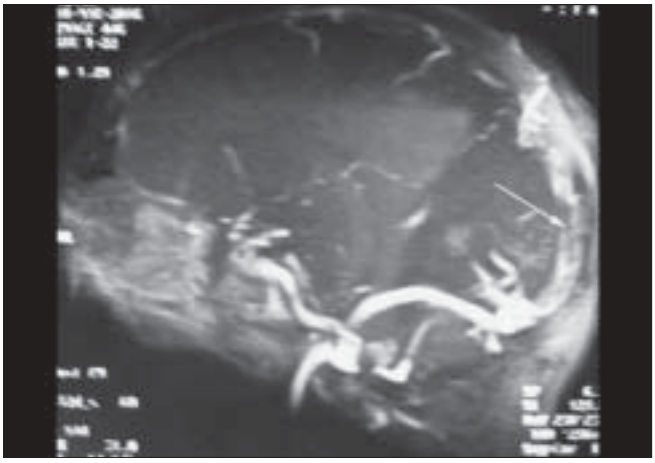

Figure 2: MRI showed features of sagittal sinus thrombosis without any evidence of venous infarction or mass lesion

The papilledema is usually bilateral and symmetric, however it may be unilateral and asymmetric; the visual sensory function is usually not affected unless macular hemorrhage and exudates or papilledema become chronic. In aseptic thrombosis due to polycythemia vera, the raised intracranial pressure usually results from occlusion of the superior sagittal sinus producing the symptoms of papilledema, which results from impaired cerebrospinal fluid (CSF) absorption and consequent elevation of intracranial pressure.

Cerebral sinus thrombosis is diagnosed by CT scan or MRI. On non-contrast CT scan a thrombosis is visualized as a linear area of hyperdensity. On contrast imaging, a dark triangle may be observed in the sinus, where flow is blocked and this is termed as the *empty delta sign*. Magnetic resonance imaging and magnetic resonance venography may demonstrate acute thrombosis and lack of blood flow in involved sinus. It remains the best diagnostic tool and should not be delayed if there is a clinical suspicion of thrombosis.[6,7]

Medical management of cerebral venous thrombosis should include attention to the treatment of seizures, broad-spectrum antibiotics for septic thrombosis, detection and management of metabolic derangements and management of cerebral edema and elevated intracranial pressure. Patients with aseptic thrombosis are treated with anticoagulant to combat risk of hemorrhage. Prolonged anticoagulation may be required for refractory cases or for patients with an identified pro-thrombotic state.[2]

For patients who complain of progressive or rapid loss of vision, the procedure currently favored by ophthalmologists is fenestration of optic nerve sheath which consists of partial unroofing of the orbit and intraorbital incision of the dural-arachnoid sheaths surrounding the optic nerve.[5] In some cases surgical thrombectomy can be done. In patients who do not respond to these conservative measures, lumbo-peritoneal shunt, ventriculoperitoneal or ventriculoatrialshunt, and subtemporal decompression is necessary to reduce CSF pressure.[8,9]

In a patient of polycythemia vera, the management is only to prevent thrombotic complication and to maintain normal hematocrit level by doing periodic phlebotomies and the correction of raised intracranial pressure. Hydroxyurea or Interferon-α or Anegrelide can be given to control thrombocytosis.[1] Visual prognosis in patients with papilledema treated medically or surgically is generally good.

In our case the patient was diagnosed and treated earlier, because of which he regained his vision. Hence any case presenting with bilateral papilledema should always necessitate a thorough clinical, hematological, biochemical and radiological workup and prompt treatment in order to save vision and life. The ophthalmologist may play an important role in the care of patients with polycythemia vera since ocular lesions may precede potentially serious extra-ocular disease.

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