Original Research Paper

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Diplopia as initial manifestation of Polycythemia Vera

Medicine

KEYWORDS:

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ABSTRACT

Diplopia is rare initial presentation of polycythemia Vera. If diagnosed early, treatment can reduce mortality & morbidity significantly often it may present with headache as only complaint thus diagnosis is likely to be missed. Our Patient presented with only complaints of diplopia. This case is being presented for rarity of association of polycythemia Vera leading to cerebral venous sinus thrombosis with papilledema in a young adult with normal platelet counts.

Introduction

Polycythemia Vera is blood disorder in which bone marrow makes too many red blood cells. It is a chronic progressive & ultimately fatal disease. Polycythemia can be secondary due to high altitude, chronic obstructive pulmonary disease, tumors. True Polycythemia Vera is a rare entity. Symptoms are caused by increased blood volume, viscosity & by thrombotic hemorrhagic complication.

Case Report

35 years old male, exsmoker presented with complaints of Diplopia since 10 days duration. On examination patient was afebrile , pulse – 60/min, regular, central nervous system examination – NAD, No cranial nerve involvement on admission, Respiratory system – no abnormality detected, Cardiovascular examination no murmur. Fundoscopy suggestive of bilateral papilledema which was confirmed with ophthalmologist. Multiplanar multiecho MRI of brain & Venography was brain showed extensive thrombosis of entire superior sagittal sinus, right transverse & sigmoid sinuses, straight sinus & cortical veins of high frontoparietal lobes. Complete blood count (CBC) showed Hemoglobin – 20.2g/dl, hematocrit – 55.6%. Serum Homocysteine was normal. Electrocardiogram was normal. 2 D echocardiography no evidence of valvular heart disease. Arterial blood gas showed normal oxygen saturation. Serum Erythropoietin levels were decreased. JAK 2 gene mutation testing were present.

Patient was started on heparin with warfarin. Ecosprin was also started. Patient developed Lateral rectus palsy on day 3 of admission, Diamox was started for the patient. Alternate day pbletomy was done. Repeat CBC count after phlebotomy showed HB – 16g/dl, hematocrit – 50% . Patient was also advised to increase fluid intake orally to decrease the viscosity of blood. Patient was discharged on Ecosprin, warfarin & Diamox. Patient showed improvement in vision. His diplopia was improved on day 12 of treatment.

Discussion

Polycythemia is a myeloproliferative disorder. It can be primary caused by JAK2 gene mutation or secondary caused by increased Serum erythropoietin levels in response to chronic hypoxia leading to excess production of erythrocytes from bone marrow.

Cerebral venous thrombosis is a serious complication of Polycythemia Vera. The mechanism by which polycythemia causes thrombosis is still under investigation but based on previous research, stasis of blood causes hyperviscosity possibly leading to development of thrombosis. Our Patient had history of smoking for 1 year which had further aggravated his risk of having thrombosis. His arterial blood gas (ABG) did not show any evidence of hypoxia, also he had stopped smoking 3 months before admission that rules out smoking as primary cause for his polycythemia. Polycythemia Vera was diagnosed based on increase in Hemoglobin levels, increased hematocrit. Serum Erythropoietin levels were reduced & JAK2 mutation was present in our patient as seen in Polycythemia Vera.

This case report is unique in a way: - A young adult, Exsmoker, presented with only complaints of Diplopia as initial manifestation of Polycythemia Vera. Early diagnosis & treatment saved vision of the patient & other serious neurological complication.

In aseptic thrombosis due to polycythemia vera, increased intracranial pressure (ICP) usually result in occlusion of superior sagittal sinus producing symptoms papilledema which results from impaired cerebrospinal fluid absorption & consequent elevation of ICP.

In a patient with Polycythemia Vera, management is only to prevent thrombotic complication & to maintain normal hematocrit levels by doing periodic phlebotomy & correction of raised ICP. Hydroxyurea & Interferon alpha or Anegrelide can be given to control thrombocytosis. Visual prognosis in patients with papilledema treated medically or surgically is generally good.

In our case, Patient was diagnosed & treated early because of which his vision had improved.

Conclusion

Any case presenting with diplopia & bilateral papilledema should always necessitate a thorough clinical, hematological, biochemical & radiological workup & prompt treatment in order to save vision & life of patient.

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