Research Paper

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An Uncommon Presentation of Polycythemia Vera as Ischemic Stroke—A Rare Case Report

Medical Science

KEYWORDS : Polycythaemia Vera, Ischemicstroke, Hypercoaguble state

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ABSTRACT

Ischemic stroke as an initial presentation of PolycythaemiaVera is rare. It usually presents with an unspecific neurological and vascular symptoms. We herein report a case of 32year old male presenting with hemianopia in left eye and magnetic resonance imaging revealed a foci of restriction diffusion measuring between 3-4 mm in posterior watershed area.

In our evaluation we diagnosed the case as polycythaemia Vera by JAK2V6 17F mutation analysis and haematocrit greater than 60%, haemoglobin greater than 20 gm/dl. The case is reported here in view of its rare initial presentation.

INTRODUCTION

Stroke or cerebrovascular accident is defined as the abrupt onset of a neurological deficit that is attributable to a focal vascular cause [(1)]. It is the third leading cause of death worldwide and major cause of adult neurological disability. About 80% of strokes are caused by primary cerebral ischemia and rest are due to haemorrhagic [(2)]. Majority of ischemic strokes are due to local damage to a vessel wall from atherosclerosis and thrombosis rest of them are due to embolic causes and about one quarter are cardio embolic[(2)]. Haematological diseases such as essential thrombocythemia, polycythaemia rubra Vera (PRV), and thrombotic thrombocytopenic purpura can cause stroke [(3)] through it is very rare. Patients with polycythaemia Vera are at high risk for vaso-occlusive events ranging from digital ischemia to cerebral ischemia. We here report a case of polycythaemia Vera presenting as ischemic stroke as initial complaint (a rare cause).

CASE REPORT

A 32 year old male with Body mass index of 20kg/m^2 was brought to emergency room with sudden loss of consciousness followed by hemianopia of left eye and dizziness from 2 hours. The onset was sudden and there was no progression .There was no motor weakness, no sensory deficit and no coordination abnormalities, no signs of cranial nerve involvement (except for hemianopia). His gait was normal

There was no history of trauma to the head and fever

He had history of headache since 1year.headaches were generalised, lasted for 1-2 hours and subsided spontaneously.

There was no previous history of seizure disorder, pulmonary tuberculosis, and cardiac problems .He was not a known diabetic or hypertensive. He was not a smoker, non-alcoholic and did not have any history of illicit drug usage. He is a professional photographer by occupation. There were no neurological or haematological complaints in the family.

General examination revealed conjunctivalcongestion(Fig 3). Erythema of palms and soles were seen, no enlarged lymph nodes. .

Physical examination showed he was afebrile, normotensive and had a pulse rate of 84/minute, normal sinus rhythm and had a respiratory rate of 16/minute.

Higher mental functions were normal .speech was fluent Motor system showed no focal weakness, no abnormal movements, coordination was normal, sensory system examination was normal. All cranial nerves were intact except second cranial nerve .deep tendon reflexes were normal.

Laboratory evaluation revealed a haemoglobin of 20.6 grams/deci litre .Red blood cell count was 7.1million /cumm.white blood cell count was 4600/cumm, with 75%neutrophils, platelet count was 3.2 lakh /cumm.

Echocardiogram was within normal limits. Liver function tests, Renal function tests and urine analysis were normal. A transthoracic echocardiogram showed a 60% ejection fraction and no abnormalities.

An abdominal ultrasound showed mild splenomegaly. Chest rotenjogram was normal

Arterial blood gas analysis showed blood P^H of7.42, $Pco_2$was 44 mm of mercury, Po_2 was 76 mm of mercury,

Assay of JAK2V617F mutation was positive. Erythropoietin level was 12.4 MIU/L (normal range 2-22 MIU/L)

A brain magnetic resonance imaging study revealed a foci of restricted diffusion measuring between 3-4 mm in posterior watershed area(Fig 1 and Fig2). Carotid Doppler was performed and it showed no signs of any athermanous plaque.

Withraised haemoglobin %,packed cell volume, red blood cell count, normal serum erythropoietin level and absence of any apparent secondary cause of erythrocytosis, wediagnosedthe case as polycythaemiaVera .In a middle aged male who is normotensive and non-diabetic, withoutdyslipidaemia, and normal BMI and no other significant risk factors for atherothombosis and without any evidence of source of embolism,polycythaemiaVera was thought to be the underlying cause of ischemic stroke. Patient was treated with anti-platelet agents. Phlebotomy was

done twice, and hydroxyurea 500mg/day was started.

DISCUSSION

PolycythaemiaVera is a clonal disorder involving a multipotent haematopoietic progenitor cells in which phenotypically normal red cells granulocytes and platelets accumulate in the absence of a recognisable physiologic stimulus.[4].

PolycythaemiaVera occurs in 2 per 1, 00,000 persons [4]. ischemic stroke may be the first presenting symptom of polycythaemiaVera in 15% or more of those affected [5,6]. Assumptions had been previously made that cerebral ischemia in polycythaemia was due to increased viscosity of the blood flow, leading to poor cerebral blood flow along with platelet activation creating an environment for thrombus formation in local cerebral arteries and arterioles[7,8,9]. The increased haematocrit of polycythaemiaVera is main determinant of blood viscosity. As viscosity increases ,cerebral blood flow decreases [8,9].Platelet marginalization with increased contact to vessel walls occur , along with local effect of high haematocrit on vessel wall[8,9] ,this fulfils all three components of virchows triad[10],and is consistent with the thought that many strokes in polycythaemia are due to propagation of local thrombus. [8,9,15,16]

With increase in the plasma red cell volume, theviscosity of blood increases leading to complications of polycythaemia including stroke. Acute coronary syndrome, pulmonary emboli, deep vein thrombosis and other thrombotic phenomenon. However patients may present with nonspecific symptoms including plethora, headache,itching,chest tightness and weakness [11].Neurological disorders associated with polycythaemia rubra Vera include transient ischemic attacks, cerebral infraction and cerebral haemorrhage. Less specific symptoms include dizziness,paraesthesia, visual disturbances, tinnitus and headache explained on basis of reduced cerebral blood flow and increases blood viscosity[12].

In our case a middle aged man with BMI of 20kg/m^2normotensive, non-diabetic, not a smoker or alcoholic, no dyslipidaemia and no haematological abnormalities, except for haematocrit of 62% and HB of 20.6gm/dl and erythropoietin levels were normal and as we could not find any secondary causes of polycythaemia .we treated him as polycythaemia Vera presenting as ischemic stroke.

Management include anti platelet,phlebotomy and chemotherapy .Hydroxyurea is used as an adjunct to phlebotomy especially in patients prone to thrombosis.pegyted interferon alpha produce complete remissions in polycythaemia Vera patients , and its role in this disorder may be expanded. Radioactive phosphorus (32P) are leukemogenic in polycythaemia Vera, and their use should be avoided[4].low dose aspirin (75-150 mg/day) should be given to all the patients if not contraindicated[13]. Haematocrit should be used as routine investigation in stroke patients[14] .allopurinol may be used for high uric acid levels. Effective management of polycythaemiaVera rubra can prevent further stroke.

CONCLUSION

Proper management of polycythaemiaVera can prevent recurrent stroke. Early diagnosis of polycythaemia rubra Vera, phlebotomy, and chemotherapy and interferon therapy can prevent complications.

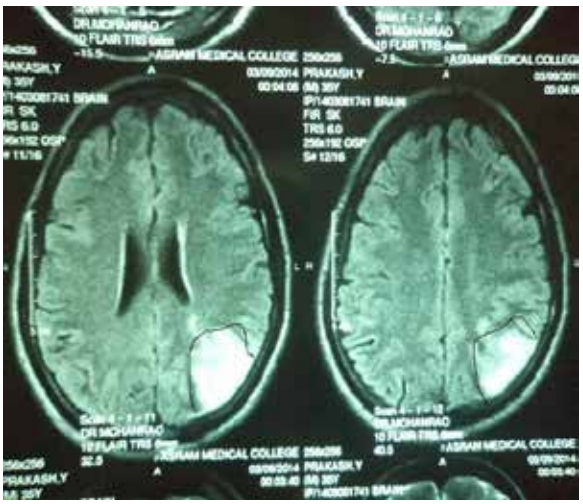

Fig 1. MRI SCAN OF THE PATIENT, Acute infract in posterior watershed area

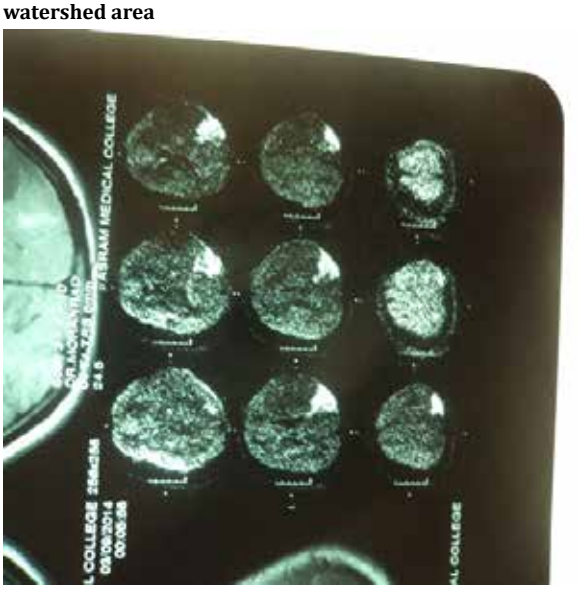

Fig 2. Diffusion images in MRI of the patient

Fig 3.CONJUCTIVAL CONGESTION of the patient

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