CASE REPORT

Ischaemic Stroke as A Presenting Feature of Polycythemia RubraVera - A Case Report

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ABSTRACT

Stroke secondary to polycythemia rubra vera (PRV) is very uncommon. A 55 years old man presented to us with ischaemic stroke, imaging revealed one recent and multiple previous Lacunar infarcts. Evaluation for underlying cause proved him as a case of PRV. Effective treatment of PRV can prevent the further recurrence of Ischaemic stroke.

Key Words: polycythemia rubra vera, ischaemic stroke, hypercoagulable state

Introduction

Stroke is a clinical syndrome characterized by sudden onset of focal rather then global neurological signs that lasts more than 24 hours or leads to death and has a presumed vascular cause, which includes both infarction and haemorrhage.[1] It is the third leading cause of death worldwide and major cause of adult neurological disability.[1] About 80% are caused by primary cerebral ischaemia resulting in infarction, on the other hand only 20% are caused by cerebral haemorrhage.[1] Ischemia occurs when there is a decrease in blood flow to less than 20 mL/100 g of brain tissue per minute. Reduction of blood flow to less than 16mL/100g of brain tissue per minute leads to tissue death within one hour. In the absence of blood flow, death of brain tissue can occurs within 4 to 10 minutes[2]. Majority of ischaemic stroke are duo to local damage to a vessel wall from atherosclerosis and thrombosis. Rests of them are embolic cause and about one-quarter are cardioembolic[1]. There are many other rare causes of ischaemic stroke. Several hematological disorders and haemostatic defects also increase risk of ischemic stroke. A common feature of these disorders is the creation of a prothrombotic state (hypercoagulable state). Hematological diseases such as essential thrombocythemia, polycythemia rubra vera (PRV), and thrombotic thrombocytopenic purpura can cause stroke,[3] though it is very rare. They usually produce their own features before development of stroke. Here we report a case who had Ischaemic stroke as a presenting feature of Polycythemia rubra vera.

Case Report

A 55 years old normotensive, nondiabetic, nonsmoker, previously healthy man from Mymensingh presented with sudden onset of weakness of the left side of his body. On quarry, he gave history of global dull headache and fullness of upper abdomen for last 1 month. There was no history of chest pain, chest tightness, palpitation, respiratory distress, skin rash, bleeding manifestation, itching, blurring of vision, leg swelling and fever. On clinical examination, he was alert, well oriented with plethora and injected conjunctivae, no cyanosis, blood pressure was 130/80 mm of Hg.

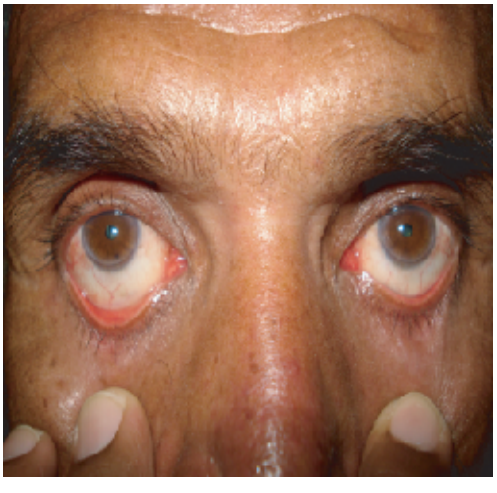

Fig 1: *Plethora with injected conjunctivae*

Neurological examination revealed, dysarthria, left sided upper motor type of facial palsy, muscle power in left upper and left lower limb were 4/5 with exaggerated all deep tendon reflexes in the same side. Planter reflex was extensor and Hoffmann's sign was positive in left side. Abdominal examination revealed palpable huge spleen, 15 cm from left costal margin along its long axis. Liver was not palpable. Otherwise all other examination findings were normal.

Investigation revealed Hb%- 17.1 g/dl, total red blood cell count - 8.1 x 1012/L (normal 4.5-5.5 x 1012/L), total white blood cell count - 23.5 x 109/L (4.00-10.00 x 109/L) with Neutrophils 87%, Pack cell volume - 53% (normal 37-53%), Neutrophilic leukocytosis on peripheral blood film, ESR - 02 mm in 1st hour. On the other hand total platelets count, blood sugar, blood urea, serum creatinine, serum electrolytes all were normal. ICT for Kala azar, HBsAg, Anti HCV were negative. Ultrasonography of the whole abdomen showed hugely enlarged spleen with increased coarse parenchymal echo texture all over without any focal lesion, splenic vessels were mildly dilated. Chest X-ray, ECG and Echocardiography were normal. MRI of the brain showed hypointense signal changes in T1 and hyperintense signal changes in T2 and flair images in the area of right middle cerebral artery distributions, There were also bilateral multiple old Lacunar infarct in thalamo-ganglionic regions. Finally he was diagnosed as a case of Ischaemic stroke with left sided hemiparesis secondary to Polycythemia rubra vera. Ischaemic stroke is a rare first presenting feature of Polycythemia Rubra Vera and uncommon cause of Ischaemic stroke.

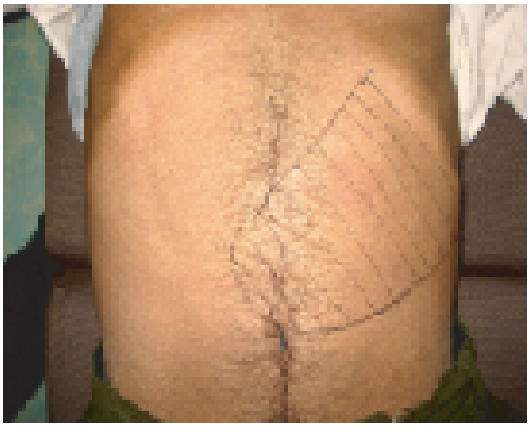

Fig 2: *Huge splenomegaly (surface marking).*

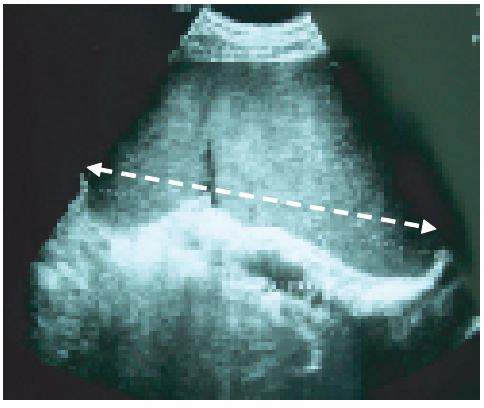

Fig 3*: Ultrasonography showing splenomegaly*

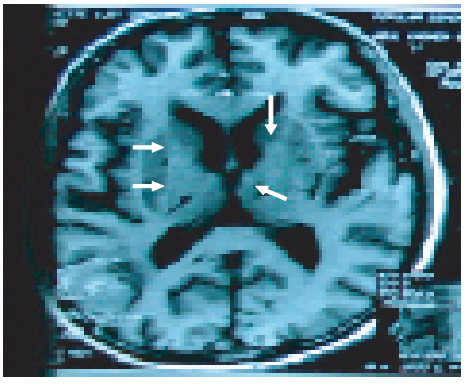

Fig 4: *Axial T1 weighted MRI of brain showing multiple bilateral hypointense signal changes in thalamoganglionic regions (arrow).*

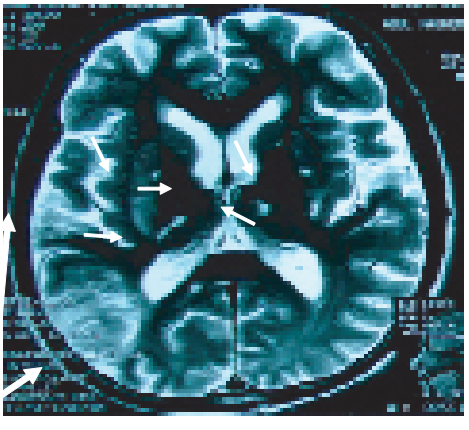

Fig 5: *Axial T2 weighted MRI of brain showing multiple bilateral small hyperintense signal changes in thalamoganglionic regions (small arrow) and large hyperintense signal changes in right parietal region (large arrow).*

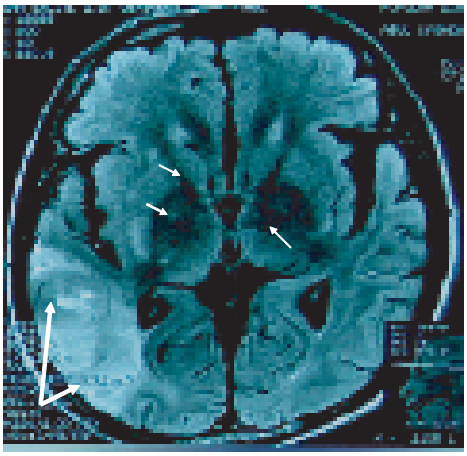

Fig 6: *Axial Flair image in MRI of brain showing multiple bilateral small hyperintense signal changes in thalamoganglionic regions (small arrow) and large hyperintense signal changes in right parietal region (large arrow).*

Discussion

Polycythemia Rubra Vera (PRV) is a rare myeloproliferative disorder that has a high risk of stroke. PRV is often found after the manifestation of cerebral infarction.[4] Our case of PRV had multiple old lacunar infarcts which were unrecognized and came to us with a major infarct in left middle cerebral artery territory. About one month before of this event he felt mild global dull headache and fullness of upper abdomen for one month, which was due to hyper viscosity and splenomegaly. An inverse relationship between cerebral blood flow and the packed cell volume has been shown in PRV. Cerebral blood flow is significantly reduced in polycythaemia and predisposes to thrombus formation; it impairs normal uptake of oxygen by tissues and impairs the microcirculation.[5] Which initially present as Lacunar stroke and followed by major neurological events.[5]

With increase in the plasma red cell volume, the viscosity of the blood increases. Increase in viscosity present as complications of polycythemia including stroke, acute coronary syndrome, pulmonary emboli, deep vein thrombosis and other thrombotic phenomena. However, patients may present with nonspecific symptoms and signs including plethora, headache, itching, chest tightness and weakness.[6] Neurological disorders associated with PRV include transient ischaemic attacks, cerebral infarction, and cerebral haemorrhage. Less specific symptoms include dizziness, paraesthesiae, visual disturbances, tinnitus, and headache explained on the basis of reduced cerebral blood flow and increased blood viscosity.[5] Fluctuating dementia, confusional states, and chorea attributed to multiple small vessel occlusions in the cortex and basal ganglia have also been recorded.[5]

In PRV there is an absolute increase in red cell mass caused by an intrinsic problem in the red cell lineage that increases erythropoiesis. Serum erythropoietin is usually reduced or absent in PRV, but elevated in secondary polycythaemia. In our case we could not find any secondary cause of polycythemia.

The aim of treatment in PRV with stroke is to prevent further clot formation by reducing high blood viscosity. Stroke secondary to PRV should be treated with stroke regimen as well as PRV therapy, and hydroxycarbamide might have stable benefit and few side effects.[7] Phlebotomy, chemotherapy drugs (eg. Hydroxyuria) or radiation therapy can be used to suppress the bone marrow.[6] Allopurinol may be used for high uric acid levels.[6] Aspirin should be used if associated Neurological features present. Hematocrit assay should be used as a routine item in stroke patients.[7] Effective treatment of Polycythemia rubra vera can prevent the recurrence of further stroke.

References

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