Meningeal vein and subarachnoid FLAIR hyperintensities in polycythaemia vera

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DESCRIPTION
A man in his fifties without medical history or cardiovascular risk factors presented with aquagenic pruritus since 3 months, fluctuating visual disturbance and headache since 1 week, followed by acute dysarthria and right-sided motor deficit. Clinical examination showed, in addition to dysarthria and right hemiparesis, facial erythrosis. MRI showed acute infarction in the left anterior choroidal artery territory, and bilateral vascular and subarachnoid fluid-attenuated inversion recovery (FLAIR) hyperintensities predominant in the posterior brain regions (figure 1). Perfusion weighted imaging (PWI) showed bilateral cortical-subcortical occipital perfusion deficit together with global cortical hypoperfusion. Blood analysis showed increased haematocrit value (70%, normal 40%–50%), haemoglobin level (220g/L, normal 130–170g/L) and white cell count (210x10⁹/L, normal 3.9x10⁹/L–10.9x10⁹/L), and normal platelet count and erythropoietin level. Carotid doppler ultrasonography, echocardiography, and cardiac MRI were all normal. A diagnosis of probable polycythaemia vera (PV) was made according to WHO diagnostic criteria.¹

Visual symptoms, headache, pruritus and facial erythrosis rapidly improved after the start of treatment with repeated phlebotomy, hydration and acetylsalicylic acid 160mg once daily and hydroxyurea 1000mg two times daily. One week later, haematocrit value was 56%, haemoglobin level 17g/dL, and white cell count 13700/mm³, and MRI showed disappearance of FLAIR vascular and subarachnoid hyperintensities. Genetic analysis showed a JAK2 V617F mutation. Six months later, the patient fully recovered from right hemiparesis while slight dysarthria persisted.

Polycythaemia vera may be revealed by stroke due to increased blood viscosity, prothrombotic state and microemboli.

Increased blood viscosity in polycythaemia vera may manifest as reversible (when treated) low flow in meningeal veins.

Patient’s perspective
I hope this publication will help young neurologists, radiologists and haematologists have a new perspective at my disease.

Learning points
- Polycythaemia vera may be revealed by stroke due to increased blood viscosity, prothrombotic state and microemboli.
- Increased blood viscosity in polycythaemia vera may manifest as reversible (when treated) low flow in meningeal veins.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

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