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 (220 g/L, normal 130–170 g/L) and white cell count ($210x10^9$/L, normal $3.9x10^9$/L–$10.9x10^9$/L), and normal platelet count and erythropoietin level. Carotid doppler ultrasonography, ECG monitoring, transthoracic and transesophageal 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 160 mg once daily and hydroxycarbamide 500 mg two times daily. One week later, haematocrit value was 56%, haemoglobin level 17 g/dL, and white cell count 13 700/mm^3, and MRI showed disappearance of FLAIR vascular and subarachnoid hyperintensities. Genetic analysis showed a JAK2 V1617F mutation. Six months later, the patient fully recovered from right hemiparesis while slight dysarthria persisted.

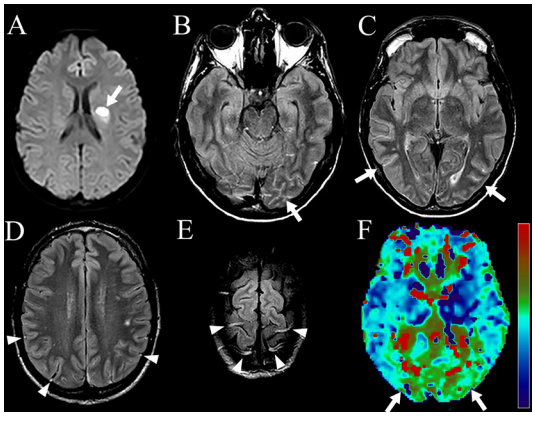

Figure 1 MRI showing (arrows) acute infarction on diffusion weighted imaging (DWI) (A), bilateral subarachnoid (B, C) and meningeal vein (D, E) hyperintensities on FLAIR, and bilateral occipital perfusion deficit (time-to-peak map, (F)).

Risk factors for thrombosis in PV include increased haematocrit (10% increase of haematocrit has been estimated to increase blood viscosity by 20%) and elevated white =cell count (ie, >15 000/mm^3). PV can result in acute brain infarction in more than 15%, probably related to increased blood viscosity, prothrombotic state (endothelial dysfunction and platelet activation are thought to be underlying mechanisms precipitating thrombus formation) and/or microemboli. Increased density on CT of cerebral venous sinuses (correlating with haematocrit and haemoglobin levels) in PV patients have been reported in earlier studies.[2–4]

The reversible vascular and subarachnoid FLAIR hyperintensities observed in our PV patient were most likely related to low flow in the meningeal veins due to increased blood viscosity, whereas perfusion deficit was attributed to venous congestion (related to reduced outflow, identical to earlier report observed in patients with cerebral venous thrombosis).[5]

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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