Essential thrombocytosis 40 years after splenectomy

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DESCRIPTION

A 69-year-old man presented at our Department of Neurology in December 2016 with acute aphasia. His medical history was notable of splenectomy after a spleen rupture due to a car accident at the age of 25, a subarachnoidal haemorrhage (SAH) without evidence of cerebral aneurysm at the age of 35 and a non-ST elevation myocardial infarction at the age of 63. For this vascular event, a drug-eluting stent was placed into the ramus interventricularis anterior, followed by combined antiplatelet treatment for 2 years, and up to admission acetylsalicylic acid was given at a dose of 100 mg per day.

Now MRI revealed a left hemispheric ischaemic stroke (figure 1A). No major vascular risk factors were found. The only pathological finding was a thrombocytosis of 1700×10^3 per µL, reference value $150–450\times10^3$ per µL. A post-splenectomy syndrome was suspected. However, previous thrombocyte levels as documented during the SAH and during routine assessments had been below 840×10^3 per µL until December 2012 (figure 1B). Other causes of thrombocytosis, such as reactive thrombocytosis or inflammatory diseases or autoimmune or paraneoplastic thrombocytosis, have not been found. Bone marrow examination and genetic testing confirmed the V617F mutation in exon 14 of the JAK2 gene consistent with essential thrombocytosis.

After stroke, antiplatelet treatment with acetylsalicylic acid 100 mg per day was continued. For treatment of the essential thrombocytosis, hydroxyurea was started (500 mg twice daily) practically normalising the thrombocyte counts.

After rehabilitation, mild aphasia remained which is currently treated with speech therapy.

Splenectomy can induce thrombocytosis.[1] However, other causes including haematological diseases, especially myeloproliferative neoplasms, should be considered. In this patient, the evaluation of thrombocyte counts over time helped us in making the diagnosis. Splenectomy after so many years leads to a moderately elevated platelet count of approximately $450–500\times10^3$ per µL.[2] The markedly raised platelet count in this patient is therefore mainly due to the clonal haematopoietic disease, marked by the acquired (somatic) JAK2 mutation. Occasionally, stroke is the first clinical manifestation of essential thrombocytosis.[3]

Learning points

- Splenectomy can induce thrombocytosis.
- However, in the diagnostic work-up of thrombocytosis secondary causes should be considered.
- Thrombocytosis can be an independent risk factor for myocardial infarction and stroke.

Contributors Both authors were involved in clinical care and investigative workup of the patient. ON conceived and designed the study and developed and wrote the neurology part of the manuscript. GK wrote the haematology part and revised the manuscript.

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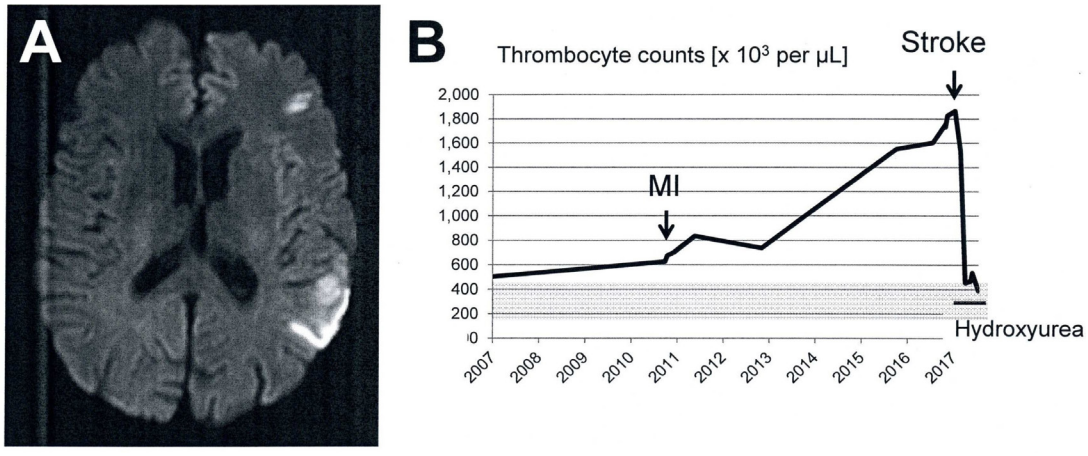

Figure 1 (A) Diffusion-weighted MRI of a left hemispheric stroke in this patient. (B) Thrombocyte counts over 10 years. Myocardial infarction occurred in November 2010 at a thrombocyte count of 625×10^3 per µL (reference value $150–450\times10^3$ per µL, grey bar). Stroke occurred in December 2016 at a thrombocyte count of 1755×10^3 per µL. Hydroxyurea treatment was started in March 2017.

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