Annals of Short Reports

Editorial
Published: 15 Apr, 2020

Polycythemia Vera Presenting with Multiple Strokes

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Editorial

A 68-year-old right-handed woman presented with sudden onset of speech disturbance (Broca aphasia) and gait instability. Over the preceding month, she noticed unusual headaches. Magnetic resonance imaging of the brain revealed multiple strokes (left middle cerebral artery, vertebro-basilar territory) without intracranial stenosis or thrombosis (Figure 1). Carotid ultrasound was normal. Cardiac monitoring revealed no arrhythmia. Laboratory tests demonstrated hemoglobin of 21.4 g/dL (12 to 16) and hematocrit of 70% (37 to 47). The peripheral-blood smear showed an elevated red blood cell count (Figure 1). Platelets (302000/mm^3), white blood cell counts (9500) and EPO level were normal. The occurrence of multiple ischemic strokes, unusual headache, the absence of cardioembolic etiology or steno-occlusive vascular disease and polyglobulia, evoked a thrombophilic state. Genetic analysis confirmed the diagnosis of Polycythemia Vera (PV) (*JAK2* V617F mutation), presenting with ischemic strokes in multiple arterial territories. She improved significantly after phlebotomy, aspirin and hydroxyurea (the hematocrit decreased from 70% to 50%). She had no recurrent events over the ensuing 11 months. PV is a chronic myeloproliferative neoplasm, which is primarily characterized by elevated erythrocyte count with high risk of thrombosis [1,2]. First described in the 19th century the understanding of the molecular pathogenesis, its natural history and the basis for its many complications continues to improve. Thrombosis is the presenting symptom in 20% of patients with PV. The incidence is estimated at 1 in 100,000 people with median age at diagnosis of 60 years [3]. This patient met the World Health Organization criteria of PV, with fulfillment of the 2 major criteria (hemoglobin >18.5 g/dL, *JAK2* 617V mutation) and 1 minor criterion (low or normal serum EPO level). The brain is the main organ involved. Seventy per cent of the thrombotic events are transient ischemic attack or strokes [4]. Hemorrhage in the nervous system is also reported. Furthermore, erythromelalgia is also a common symptom and can occur in 30% of patients with PV. The mechanism of thrombosis in PV is multifactorial [5]. High blood viscosity is the main suspected factor, in conjunction with disseminated intravascular clots and thrombocytosis. The headaches in PV are related to the increase in blood viscosity. *JAK2* V617F mutation occurs in 95% of cases of PV. *JAK2* V617F is also seen in other myeloproliferative disorders including essential thrombocythemia and primary myelofibrosis, but its presence is not diagnostic of this disease. Cytoreduction obtained by aspirin, phlebotomy and hydroxyurea allows reducing the future thrombotic events. A hematocrit <45 L/L is associated with lower rate of cardiovascular death and major thrombosis. Targeted therapies with two non-myelotoxic agents (ruxolitinib and pegylated interferon) look promising. Clinicians should be aware of rare etiology of stroke including PV. Patients remain at high risk for future cerebrovascular events and should be monitored frequently. A complete blood count is essential in the assessment of stroke.

Author Contributions

Alexis Demas - Drafting the manuscript, study concept or design, analysis or interpretation of data, acquisition of data. Laurence De Menibus Demas - Drafting the manuscript, acquisition of data.

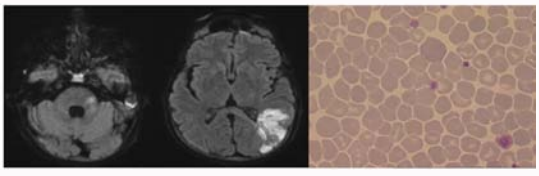

Figure 1: Cerebral MRI (diffusion weighted imaging) and peripheral-blood smear (elevated red blood cell count).

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Received Date: *23 Mar 2020*
Accepted Date: *13 Apr 2020*
Published Date: *15 Apr 2020*

Citation:
Alexis D, Laurence DMD. Polycythemia Vera Presenting with Multiple Strokes. Ann Short Reports. 2020; 3: 1049.

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