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Recurrent Ischemic Stroke Revealing Polycythemia Vera

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Abstract

Background: Polycythemia vera is a possible cause of recurrent ischemic stroke which can be prevented. **Aim:** Describe a junctional ischemic stroke without large arterial trunks stenosis associated with an acute coronary syndrome. **Case Presentation:** A 66-years-old man was admitted for abrupt recurrent right hemiparesis related to bilateral and junctional ischemic stroke lesions. He had a medical history of a vertebrobasilar ischemic stroke concurrent with an acute coronary syndrome with normal coronary arteries. Transthoracic echocardiogram showed small apical akinesia. Hemoglobin level was 18.9 g/dl with a hematocrit of 57.6%. The endogenous erythropoietin was 1.3 mIU/ml with JAK2 V617F mutation positivity (37%). After eight months of treatment (hydroxycarbamide + aspirin + allopurinol) hemoglobin was 12.5 g/dL. **Conclusion:** This case illustrates the most suggestive features of PV particularly the ischemic stroke junctional topography.

Keywords

Ischemic Stroke, Acute Coronary Syndrome, Recurrence, Polycythemia Vera, Antiplatelet Therapy

1. Introduction

Polycythemia vera (PV) is a myeloproliferative disorder associated with an increased risk of cerebrovascular diseases including cerebral ischemia and hemorrhage. It is a rare but likely underdiagnosed etiology of recurrent ischemic stroke (IS), sometimes associated with non-neurological arterial events. Cerebral ischemic events are due to increased blood viscosity and platelet activation within the central nervous system arterial vessels [1] [2] [3]. We report a case of recur-

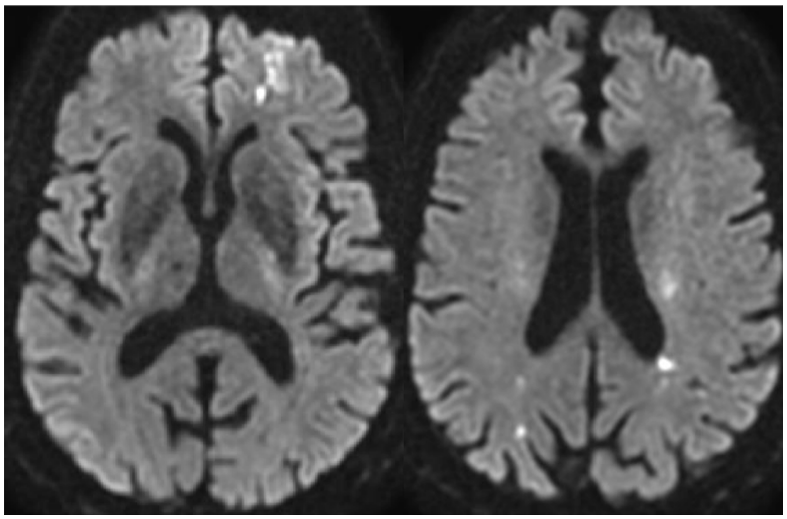
rent IS secondary to PV diagnosed nine months after the first neurovascular episode, which was associated with a coronary event.

2. Case Report

A 66-years-old man was admitted on 11/06/2019 for rapidly regressive right hemiparesis with abrupt onset, revealing a bilateral junctional ischemic stroke (IS), predominant on the left (Figure 1). Patient had a medical history of a colonic adenocarcinoma in long-standing remission, hypertension, dyslipidemia, and a first vertebrobasilar IS 9 months prior to presentation (09/2018), concurrent with a non ST segment elevation acute coronary syndrome (NSTE-ACS) with normal coronary arteries. After these two simultaneous vascular episodes, he received warfarin and aspirin as secondary prevention. Patient's hemoglobin (Hb) level was then 17.3 g/dl with a hematocrit (Hct) of 53.9% and a platelet count at 466,000/mm^3, compared to normal levels recorded in February 2017 (Hb: 14.6 g/dl; Hct: 44.4%).

As part of the etiological assessment of recurrent IS, transthoracic echocardiogram was performed and showed small apical akinesia, septal and lower wall dyskinesia, with reduced left ventricular ejection fraction (34%), and without intracavitary thrombus. ECG, 24-hour Holter-ECG, and supra-aortic trunks CT angiography were normal. Hemoglobin level was 18.9 g/dl with a hematocrit of 57.6% and a platelet count of 314,000/mm^3.

Thirteen days later, on 24/06/2019, the patient presented again with abrupt right hemiparesis, partially regressive within 30 minutes. Clinical examination revealed right brachial monoparesis (NIHSS = 1) and discrete facial erythrosis. Cerebral MRI showed multiple small bilateral ischemic lesions with mainly junctional topography (Figure 2). Hemoglobin and hematocrit levels were elevated



MCA: middle cerebral artery; ACA: anterior cerebral artery; PCA: posterior cerebral artery.

Figure 1. Brain MRI of 11/06/2019, DWI sequence and axial sections: left (MCA/ACA and MCA/PCA) and right (MCA/PCA) junctional infarcts.

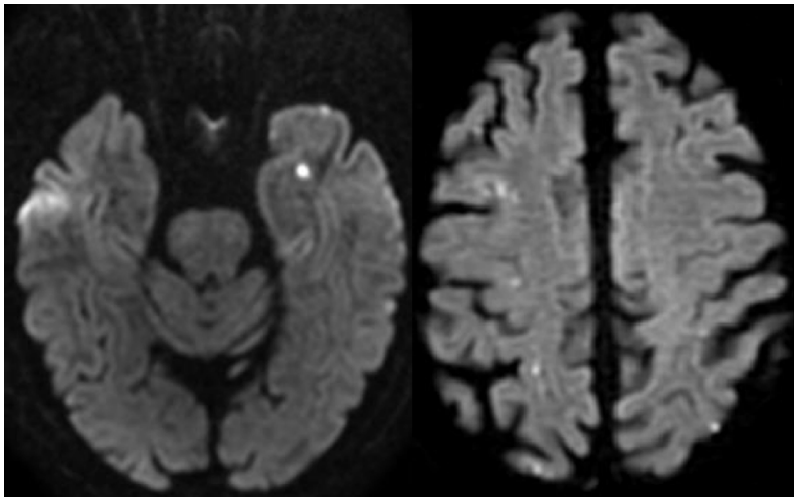


Figure 2. Brain MRI of 24/06/2019, DWI sequence and axial sections: bilateral ischemic stroke recurrence of mainly junctional topography.

again at 18.6 g/dl and 56.6% respectively, leading to suspicion of polycythemia vera. This diagnosis was confirmed by the breakdown of endogenous erythropoietin (1.3 mIU/ml) [N: 2.6 - 18.5] and positivity of the JAK2 V617F mutation at 37%, with no evidence of secondary polycythemia.

Cytoreductive treatment with hydroxycarbamide was then introduced, combined with aspirin and allopurinol. Anticoagulation was stopped. After 12 hospitalization days, the patient became asymptomatic and was discharged home. After eight months of medical treatment, hemoglobin (12.5 g/dL) and hematocrit (36.6%) had normalized. At one year follow-up, no ischemic recurrence was observed and the modified Rankin scale was 0.

3. Discussion

Arterial ischemic complications of polycythemia vera (PV) are common, usually early on, ranging from 15% in the two years prior to diagnosis and up to 40% throughout the disease course [1] [2]. The main risk factors for vascular complications in patients with PV are advanced age, history of thrombosis, dyslipidemia and hypertension, as found in our patient. IS can be indicative of PV in 15% of cases [3]. Elevated hemoglobin levels can precede the diagnosis of IS by at least three months [4].

Other ischemic complications include acute coronary syndrome which, as in our case, may be concurrent with IS or occur shortly after [5].

The mechanism of ischemic stroke in PV mainly involves blood stasis and hyperviscosity as well as an increase in peripheral resistance, leading to infarcts with predominant junctional topography, as seen in our patient [6]. Elevated hematocrit, a determinant of blood hyperviscosity, could also induce an attack on the arterial vascular wall, similar to atheromatous lesions, which results in platelet activation and triggering of the inflammatory cascade that leads to arterial stenosis and in situ thrombi formation [7].

Once PV has been diagnosed, antiplatelet therapy combined with cytoreductive therapy generally prevents ischemic recurrence, particularly with low-dose aspirin [8], which can be used in cases where Clopidogrel is ineffective [5].

4. Conclusion

Polycythemia vera is a possible cause of recurrent ischemic stroke, which can be prevented with antiplatelet therapy combined with cytoreductive therapy. Our case illustrates the most suggestive features of PV, which are the junctional topography of the IS without large arterial trunks stenosis, and a possible association with ACS with healthy coronary vessels.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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