Case Report

Cerebral Hemorrhage of a 50-Year-Old Female Patient with Polycythemia Vera

Lei Chen, MM, Han Xiao, MD, PhD, and Zhiping Hu, MD, PhD

Polycythemia vera is a chronic myeloproliferative neoplasm, which is primarily characterized by elevated erythrocyte count with the risk of thrombosis, hemorrhage, and vasomotor symptoms. More common reported about bleeding events are gastrointestinal, mucosal, and cutaneous bleeding. Spontaneous cerebral hemorrhage/bleeding is seldom reported. Here, we report the case of a 50-year-old female with polycythemia vera who developed a spontaneous cerebral hemorrhage. She improved significantly after hydroxyurea agent and red cell apheresis, and the hematocrit decreased from 74% to 40%.

Key Words: Polycythemia vera—cerebral hemorrhage—hydroxyurea—red cell apheresis

Introduction

Polycythemia vera (PV) is a myeloproliferative disorder characterized by erythremia in the blood. Thrombosis and bleeding can also be one of the initial manifestations although the reported prevalence varied considerably across some studies. The prevalence of hemorrhage at diagnosis in patients with PV lower than the prevalence of thrombosis. And the cerebral hemorrhage is rarely mentioned.

Case Report

A 50-year-old female was admitted to our hospital with acute parietal headache and blurred vision. She had hypertension 6 years and no history of smoking, hyperlipidemia, diabetes mellitus, or family history of cardiac-cerebral vascular disease. On the primary physical examination, her vital sign were stable. There are no abnormalities for four limbs' muscle tone and strength, and no pathological reflexes were detected. On rough examination, the left visual field appears to be a homonymous hemianopia. Emergent computed tomography (CT) imaging was notable for cerebral hemorrhage in the parietal and occipital lobe (Fig 1, A). Cerebrovascular malformation, cerebral tumor, or hypertension at the first consideration which result in the hemorrhage, while there was no enhancement around the lesion in enhanced magnetic resonance (Fig 1, B) and CT angiogram was negative for any vascular abnormalities (Fig 1, C). Surprisingly, laboratory blood examination displayed an elevated red cell (9.41×10^{12}/L) and hematocrit level (74%), and white cell count and platelet count were slightly increased, and other examination including coagulation function, serum chemistry, and D-dimer level were not remarkable. According to the high level of the erythrocyte, we thought it may be the PV result in cerebral hemorrhage. The diagnosis of PV was established by bone marrow biopsy (Fig 1, D) and the positive gene Janus kinase 2 (JAK2) V617F mutation. The hydroxyurea agent (2000 mg/day) was initiated and the count of red cell decreased gradually. But after taking the approach for 10 days, the white cell count decreased to 2.77×10^9/L, so we reduced the dosage of hydroxyurea to 1000 mg/day and red cell apheresis therapy and other symptomatic treatment was applied. Eventually, the patient was discharged with the normal red cell count (4.41×10^{12}/L), hematocrit level (40%), and leukocyte count (4.0×10^9/L).

From the Department of Neurology, The Second Xiangya Hospital of Central South University, Changsha 410011, Hunan, PR China.

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Address correspondence to Zhiping Hu, MD, PhD, Department of Neurology, The Second Xiangya Hospital of Central South University, 139 Renmin Road, Changsha 410011, Hunan, PR China. E-mail: zhipinghu@csu.edu.cn.

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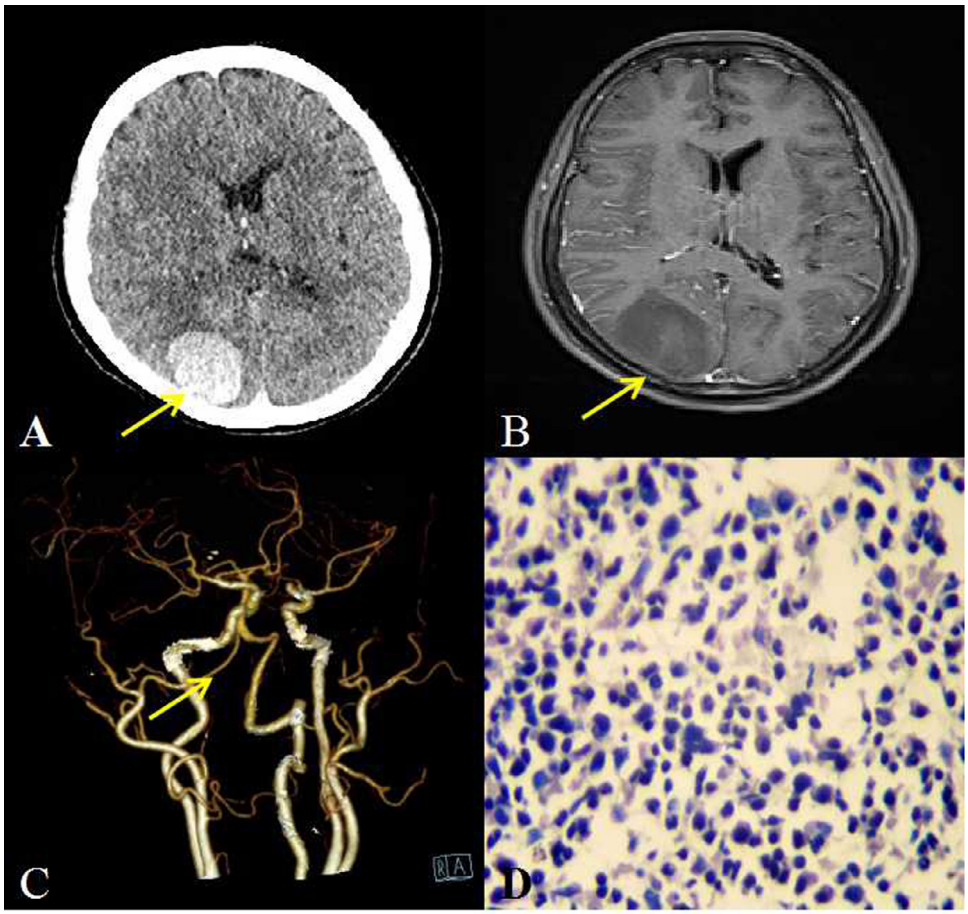

Figure 1. *(A) Computed tomographic (CT) showing the high density hemorrhage lesion in the parietal and occipital lobe. (B) Gadolinium-enhance magnetic resonance displaying no enhancement around the lesion. (C) CT angiogram showing the physiologic angusty in right vascular vertebral artery of the intracranial segment but no cerebrovascular malformation. (D) Bone marrow biopsy showing active proliferation of bone marrow cells especially the erythrocyte series and megakaryocyte series.*

Discussion

Polycythemia vera is one of the Philadelphia chromosome negative myeloproliferative neoplasms that is characterized by elevated red cell counts, increased risk of thrombosis, and bleeding, and a tendency to progress to myelofibrosis.[1] The discovery of JAK2 mutations in PV patients has revolutionized the diagnosis of PV. The JAK2 V617F mutation can be found in over 95% of PV patients.[2] As expected, there is a higher risk of thromboembolic event than bleeding. According to some literature, approximately one third of patients with PV have thrombotic events prior to or at diagnosis.[3] On the contrary, bleeding events as an initial presenting symptom at an estimated prevalence of between 3% and 8.1% in PV.[4] Although the exact mechanism of hemorrhagic manifestation remains unclear, it is believed that acquired von Willebrand disease from excessive binding of von Willebrand factor with the abnormal platelets is the most likely pathogenesis.[5] Hematological diseases are a rare cause of central nervous system emergencies, and it first reported about the cerebral hemorrhage with PV in 1923.[6] It is rare for the subsequent reports. For our patient, after ruling out other causes, we considered the cerebral hemorrhage related with PV for the definite diagnosis. The initial use of hydroxyurea has acquired certain therapeutic effects, meanwhile, some side effects have also been caused. Some literature suggests selective red blood cell apheresis is recommended alternatively, which is more appropriate to those patients who have contraindications or unwillingness to use cytoreductive therapy.[7] After the decrement of hydroxyurea and the application of red blood cell apheresis, the patient improved gradually. But there are more preferable treatments that need further research.

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