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RESEARCH ARTICLE

SIMULTANEOUS CEREBRAL VENOUS THROMBOSIS AND ISCHEMIC STROKE REVEALING VAQUEZDISEASE:A CASE REPORT

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Abstract

A 53 years old Caucasian woman, with no personal medical history. She was admitted to the emergency room, for a sudden onset of severe vertigo, intense headache, vomiting and blurred vision. Physical examination found a confused patient with Glasgow Coma Scale (GCS) of 13/15; her pulse was 98 beats/min, blood pressure of 240/140 mmHg, respiratory rate of 22 per minute. Neurological assessment showed left hemiparesis with mild dysarthria. The ophthalmological examination revealed bilateral papillary edema. Brain MRI done at the emergency department showed cerebral thrombophlebitis, with signal amputation from the left lateral sinus and superior longitudinal sinus (figure1),associated to right ischemic stroke fronto-parieto-temporal and lenticular nucleus.Complete blood count showed high red blood cell with thrombocytosis (hemoglobin 17.2 g / 100 ml; hematocrit 53,2 p. 100; platelets 814,000), erythropoietin at 0.9 mIU / mL (normal: 2.6 - 8.5), serum electrolytes, glucose, renal function, and serologieswerenormal.The tests for antithrombin III, protein C, protein S, ANA, ANCA and lupus anticoagulant were normal. Her cardiac assessments did not reveal any anomaly. Lumber puncture revealed high intracranial pressure (37mmgh) with normal cerebrospinal fluid (CSF) study. The diagnosis of polycythemia vera (PV) was retained based on the result of blood count, the low level of erythropoietin, and the JAK2V617F mutation. The patient was treated by phlebotomy, and subtractive lumbar puncture, associated with a medical treatment based on hydroxyurea and anticoagulant treatment. The follow-up was marked by clinical and biological improvement.

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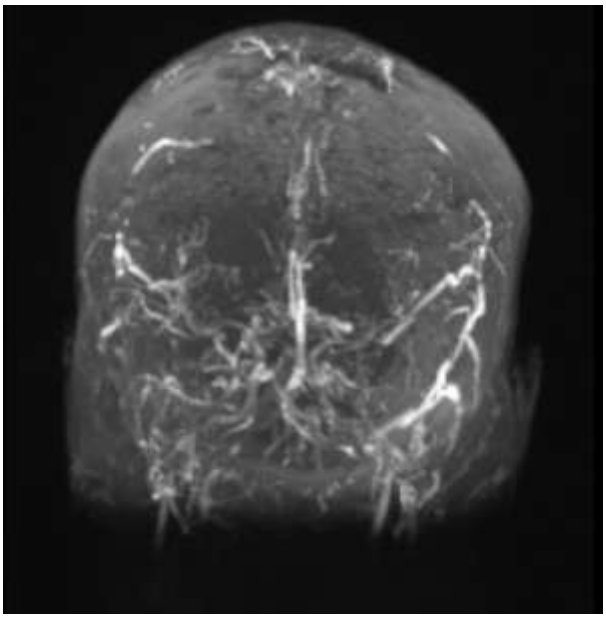

Figure 1:- Magnetic resonance venography (MRV) showed that the flow void signal of superior sagittal sinus and left lateral sinus disappeared.

Polycythemia Vera (PV) is a chronic progressive myeloproliferative neoplasm (MPN) characterized by the clonal proliferation of myeloid cells [1]. PV is expressed primarily as an excess of erythrocytes. It can, sometimes, be accompanied an excess of the other two hematopoietic lines (hyperleukocytosis and thrombocytosis). Its evolution is gradual [2]. The JAK2V617F mutation on the JAK2 gene is frequently found in PV. The clinical manifestations of PV are frequently insidious and unspecific, which may cause delayed diagnosis. However, the diagnosis and management of CVT and stroke in PV remains challenging due to low incidence and limited evidence [1]. Thrombosis is a major cause of morbidity and mortality in PV but bleeding complications can be associated [3]. Recent study confirmed that cerebrovascular complication was 10.2% (11/108), which was significantly higher than the incidence in general population [2]. Most patients (8/11, 72%) had at least two conventional vascular risk factors (eg hypertension, hyperlipidemia, diabetes and obesity). But in simultaneous cerebral venous thrombosis and ischemic stroke revealing Vaquez disease has never been described to the best of our knowledge. The presence of JAK2V617F mutation was correlated to a higher risk of thrombotic events [4]. This mutation is confirmed in our patient. The aims of the current treatment for PV are to improve symptoms and reduce thrombotic risk by keeping hematocrit level less than 45%. First-line treatment for low-risk patients is aspirin and phlebotomy. High-risk patients require aspirin and cytoreduction, most often combined with hydroxyurea or interferon (IFN) [5].

References:-

[1]Shen HX, Huang XQ, Fan CQ, Cao YX, Zhao H, Duan JG. Cerebral venous sinus thrombosis in polycythemia vera patients with JAK2V617F mutation. Chin Med J 2021;134:1251–1253. doi: 10.1097/CM9.0000000000001484.
[2] Marton I, Pósfai E, Csomor A, Vécsei L, Borbényi Z, Sas K. Cerebrovascular Complications and Polycythaemia Vera. Pathol.Oncol.Res. (2019) 25:439–442.https://doi.org/10.1007/s12253-017-0329-9.
[3]Zangari M, Tolomelli G, Lee J CH, Stein BL, Hickman K, Swierczek S, and al.Could Hypoxia increase the prevalence of thrombotic complications in Polycythemia Vera? Blood Coagul Fibrinolysis. 2013 April ; 24(3): 311–316. doi:10.1097/MBC.0b013e32835bfdb9.
[4] Malak S, Labopin M, Saint-Martin C, Bellanne-Chantelot C, NajmanA,and The French Group of Familial Myeloproliferative Disorders. Long term follow up of 93 families with myeloproliferative neoplasms: life expectancy and implications of JAK2V617F in the occurrence of complications. Blood Cells, Molecules, and Diseases 49 (2012) 170–176. 10.1016/j.bcmd.2012.06.004.
[5]Marcellino BK, Hoffman R. Recent advances in prognostication and treatment of polycythemia vera. 12 Mar 2021, Faculty Reviews 10:(29) https://doi.org/10.12703/r/10-29.