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Letter to Editor

Porto-Splenic veins thrombosis and Budd-Chiari syndrome in a patient with essential thrombocythemia – is it always the myeloproliferative neoplasm to blame?

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Dear Editor,

Essential Thrombocythemia (ET) is one of seven Myeloproliferative Neoplasms (MPNs), according to the latest WHO classification [1].

In ET, there is mainly an overproduction in bone marrow of megakaryocytes. This results in the release of a large number of platelets into the blood stream, more than 450,000X10^9/L (the upper limit of normal range). Certain mutations and abnormal karyotypes are frequently found in MPNs. Approximately 50% of patients with ET express mutually exclusive JAK2 mutation (Janus Kinase 2; 9p24), CALR (calreticulin, 19p13.2) mutation is present in 15%-24% of ET and MPL (myeloproliferative leukemia virus oncogene; 1p34) mutation is present in 4% of ET (JAK2, CALR and MPL mutations are called driver mutations). In JAK2 mutated cases, the risk of thrombosis is higher. In ET and Polycythemia Vera (PV, another MPN), arterial and venous thrombosis are the main causes of morbidity and mortality [2]. Venous Thromboembolism (VTE) may involve unusual anatomic sites such as abdominal veins, and is a frequent complication of MPNs [3]. MPNs are the most common underlying prothrombotic disorder found in in patients with SVT. The strong association between SVT and MPN has led to recommendation to screen for MPN when SVT is diagnosed. In ET patients, the estimated prevalence is 9-13% [4]. Hypercoagulable disorders such as antiphospholipid syndrome, protein C or S deficiency. factor V Leiden mutation are independent risk factors for SVT [5]. There is a high prevalence of the JAK2 (V617F) mutation found in SVT. It is possible that JAK2 itself had local effects on the splanchnic venous system [6]. VTE can be presenting symptom of other malignancies, therefore in patients with long-standing and stable MPN, a new VTE should raise the possibility of a new malignancy. Malignancy itself induces a thrombophilic state by increasing the risk of venous stasis, endothelial injury and an imbalance of pro and anti-thrombotic factors leading to a hypercoaguable state. Additional insults to this thrombotic balance are introduced by patient-specific, treatment related and tumor-specific factors [7].

Budd-Chiari Syndrome (BCS), which is one of the Splanchnic Vein Thrombosis (SVT), is a severe potentially fated disease caused by obstruction in the hepatic venous outflow tract [8]. BCS is caused by thrombosis and is divided into "classical BCS" (obstruction of hepatic veins) and "hepatic vena cava BCS" (thrombosis of intra/suprahepatic portion of Inferior Vena Cava (IVC)). Both are classified as "primary BCS" and regarded as hepatic expression of an underlying combination of several pro-thrombotic conditions in blood disorders such

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as MPN. Thus BCS should be excluded in any patient with acute or chronic liver disease.

Portal Vein Thrombosis (PVT) is one of the most common vascular disorders of the liver with significant morbidity and mortality. Like BCS, it is also one of the SVTs. PVT can typically be caused by cirrhosis, hepatobiliary malignancy, abdominal infectious or inflammatory diseases, and MPN. The term PVT refers to the complete or partial obstruction of blood flow in the portal vein, due to the presence of a thrombus in the vassal lumen. The following clinical case highlights the importance of awareness when an SVT appears in a patient with MPN. Frequently the SVT is linked to MPN, yet in rare cases it is linked to a second malignancy that must be considered.

A 19-year-old healthy male was diagnosed with ET. Complete blood count revealed a platelet count of $1.466X10^9$, without anemia, high hematocrit or leukocytosis. Inflammatory diseases and iron deficiencies were all excluded as causes of secondary thrombocytosis. Our patient was a JAK2V617F heterozygote and the bone marrow biopsy was slightly hypercellular with large mature clusters of megakaryocytic hyperplasia, hyperploid nuclei and no reticulin fibrosis, all consistent with ET diagnosis. The possibility of prefibrotic myelofibrosis was excluded. The karyotype analysis was normal. BCR-ABL was negative. LDH level was within normal limits. Von Willebrand factor level and activity were normal. WHO criteria for PV, PMF, CML, and MDS were negative.

The ET was characterized as low risk (age<60, no thrombosis history and JAK2 mutation present). Treatment with Aspirin (100 mg tablet) was started. The ET was stable over the years. Ten years after ET diagnosis our patient developed Budd- Chiari syndrome and portal vein thrombosis. He was treated with anticoagulants and in continuation Transjugular Intrahepatic Portosystemic Shunt (TIPS) was implanted, to treat a huge ascites. Three months later a symptomatic hypercalcemia was investigated. A PET-CT was performed and a tumor in the right chest wall was detected. A biopsy revealed an angiomyosarcoma. Chemotherapy was started but the patient died at age of 29 years.

Our aim is to emphasize that an SVT in a patient with MPN is a frequent finding. It is impossible to deny that SVT in this case was a consequence of ET. Nevertheless, SVT can present symptoms of a secondary malignancy so that, regardless of MPN, this possibility must be carefully pursued.

Compliance with ethical standards

Research involving human participants and/or animals: Our letter is a case report, not a study.

Informed consent: Our letter describes a case report, the patient died before the letter was conceived.

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