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PORTAL HYPERTENSION COMPLICATED BY GASTROESOPHAGEAL VARICES IN A PATIENT WITH MYELOFIBROSIS – A CASE REPORT.

General Medicine

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ABSTRACT

Myelofibrosis is a hematological neoplastic condition in which there is inadequate production of blood cells due to fibrosis and scarring of the bone marrow which might lead to the formation of portal venous clots and extramedullary hematopoiesis. It is thought that this extrahepatic portal venous clots are responsible for the cause of noncirrhotic portal hypertension in this population. The incidence of portal hypertension occurs as a complication in around 10-17% of patients [1]. Here, we describe a case of a 54-year-old female patient who presented with one episode of hematemesis. On examination, severe pallor was present and spleen was moderately palpated (figure 1). Upper gastrointestinal endoscopy revealed varices and ligation was done. Radiological imaging showed portal cavernoma formation; secondary to portal hypertension due to extrahepatic portal vein obstruction (EHPVO). Her complete blood count showed bi-cytopenia for which a bone marrow biopsy was done suggestive of myelofibrosis.

KEYWORDS

Myelofibrosis, Portal hypertension, Portal cavernoma, EHPVO, Hematemesis, Varices

INTRODUCTION

Myelofibrosis is a neoplastic disorder that is characterized by fibrotic replacement of the bone marrow, which leads to ineffective hematopoiesis which in turn leads to the development of extramedullary hematopoiesis (EMH). The main site of EMH is the liver and spleen. The emergence of noncirrhotic portal hypertension, which presents as ascites and/or variceal hemorrhage, is a clinically significant result of liver EMH. A microvascular or macrovascular clot, an injury, or the loss of tiny or big portal veins are thought to be the causes of portal hypertension in these cases with myelofibrosis.

CASE REPORT

A 54-year-old female patient presented with one episode of hematemesis. The patient is a known case of diabetes mellitus on medication. On examination, the patient had pallor and mass palpable per abdomen. No previous such episodes in the past and no history of malena. Her complete blood picture shows bi-cytopenia with hemoglobin of 3.3 g/dl with an RBC count of 1.31 x 10^6 /ul and platelet count of 27000. Peripheral smear showed marked anisocytosis with normocytic normochromic and microcytic hypochromic RBCs, polychromasia and tear drop cells(dacrocytes) are seen along with platelets reduced in smear. Reticulocyte count was 8.40 with RPI of 0.8. (reticulocytosis). Rest of the investigations – renal and liver function tests were normal. Four-column massive varices were discovered during the endoscopy along with portal hypertensive gastropathy. Hence Endoscopic variceal ligation (EVL) was therefore performed. Abdominal ultrasonography shows mild hepatomegaly with grade 1 fatty liver, moderate splenomegaly, and portal cavernous transformation(figure 2). Portal Doppler was suggestive of portal hypertension. Abdominal computed tomography revealed gross hepatosplenomegaly with non-visualization of the main portal vein with portal cavernoma formation. A bone marrow biopsy was done to show intramedullary fibrosis with myeloid and megakaryocyte cells, erythroid cells are absent. IHC CD34, C Kit, and CD30 do not highlight any atypical cells /increased blasts. Masson trichrome and reticulin stains highlight fibrosis(figure 3). i.e., suggestive of myelofibrosis as a result of myeloproliferative disorder.

The patient was prescribed Tab. Danazol 100mg, Tab. Thalidomide 50mg, Tab. Prednisolone 40mg in tapering doses and she was advised serum lactate dehydrogenase, Next Generation Sequencing (NGS) for Chronic myeloproliferative disorders, Fluorescent in situ hybridization/RTPCR for BCR-ABL (Philadelphia chromosome) and asked to follow up.

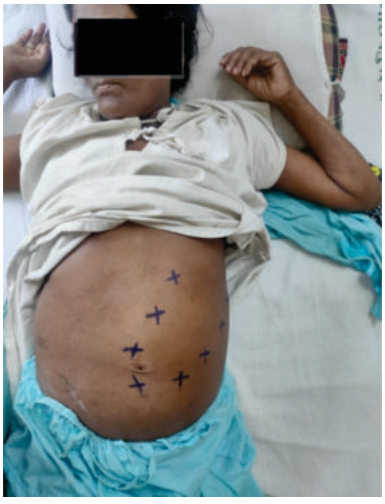

Fig 1: Splenomegaly

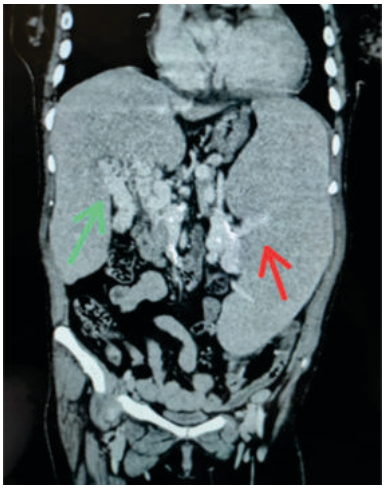

Fig 2: Green Arrow- Portal Cavernoma Red Arrow - Splenomegaly

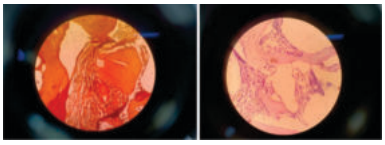

Fig 3: Bone Marrow Biopsy Showing Fibrosis (left – Reticulin Stain)

DISCUSSION

Clonal stem cell disorders include myeloproliferative diseases (MPD). Polycythaemia vera (PV), essential thrombocythemia (ET), and chronic idiopathic myelofibrosis (IMF) are the three main MPDs. Abdominal manifestations include organomegaly i.e., splenomegaly and hepatomegaly. The hepatic manifestations are a result of these MPDs affecting the hepatic circulatory systems. Budd–Chiari syndrome (BCS), portal vein thrombosis (PVT), and nodular regenerative hyperplasia are common hepatic manifestations of MPDs. extramedullary hematopoiesis, increased hepatic blood flow, and secondary hemosiderosis from multiple blood transfusions are a few other features seen in MPDs.

Many disease-specific pathways lead to portal hypertension in myelofibrosis. Non-cirrhotic portal hypertension and its morbid consequences appear mostly in the late stages of myelofibrosis. The exact mechanism is not known. These tumors can cause portal hypertension because of increased portal venous flow, i.e., associated with splenomegaly [2,3]. It is reported that it can also occur due to the increase in the pre-sinusoidal blood flow resistance secondary to extramedullary hemopoiesis in hepatic sinusoids [4]. The other suggested mechanism for the development of portal hypertension in these population groups is due to micro and macrovascular thrombosis with damage and the loss of the portal vein [5]. This also might be accompanied by rupture of varices leading to gastroesophageal variceal hemorrhage [6].

In conclusion, noncirrhotic portal hypertension's morbid effects might manifest as early as a year after diagnosis, but they typically manifest in the late stages of myelofibrosis and related conditions (ET and PV). Given the high prevalence of varices at autopsy and the recurring case reports of variceal bleeding in this community, patients with myelofibrosis, ET, or PV may benefit from primary variceal screening, keeping in mind that portal hypertension may develop. To more accurately categorize those in this particular cohort who are at risk for developing portal hypertension, more research is needed.

While the patient's overall health is good, preventive endoscopic therapy or endovascular treatment should be taken into consideration when a risk of rupture is possible. The median survival period for patients ranges from 3 to 7 years, and only 20% of patients can expect a median survival of 10 years due to the dismal prognosis of myelofibrosis [6]

Next Generation Sequencing testing should be expanded in order to determine chronic myeloproliferative disorders. Ruxolitinib, a JAK2 inhibitor, has been shown to be effective in reducing splenomegaly and easing constitutional symptoms in the majority of Myelofibrosis patients while also extending survival; however, the only therapeutic option that provides a potential cure is allogeneic hematopoietic stem cell transplantation.

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