



M FHAISOL YUSOF¹, SAFREEDA SF SALIM²

^{1,2} *UiTM HOSPITAL, SUNGAI BULOH, SELANGOR, MALAYSIA*

INTRODUCTION

Non cirrhotic **portal vein thrombosis (PVT)** is an unusual cause of abdominal pain. In addition, a patient who frequently visited ED may have something to befall if we do not apprehend the case immediately.

CASE

A previously healthy **27-year-old gentleman** had multiple visits to our ED complaining of **abdominal pain** especially over epigastric region. During his 1st visit, he also had vomiting and 1 episode of minimal **hematemesis**. The platelets showed severe **thrombocytosis**, and per rectal examination revealed no melanic stool. He was then discharged by surgical team with an outpatient OGDS to rule out severe gastritis and Mallory-Weiss tear. Prior to OGDS, he had 2nd ED visit due to epigastric pain but no more hematemesis. The pain was controlled with analgesia and was discharged well. His thrombocytosis was investigated as outpatient. His 3rd ED visit occurred at midnight few days after, complaining of severe persistent epigastric pain which was not resolved with analgesia. A thorough examinations by ED team showed **hepatosplenomegaly** with **persistent thrombocytosis**. Grey-scale imaging of bedside ultrasound abdomen demonstrated no ascites, numerous clusters of periportal veins, and hepatosplenomegaly. He was referred to gastroenterology team as severe gastritis to rule out hematological malignancy likely essential thrombocytosis for further in patient workups such as OGDS and ultrasound doppler of abdomen. CT Abdomen was done which demonstrated hepatosplenomegaly, chronic portal vein and splenic vein thrombosis with portal hypertension after formal doppler showed similar findings (**Figure 1-3**). OGDS revealed large varices (**Figure 4**). **JAK2 V617F mutation** was positive. He was planned for *Bone Marrow and Trepine Biopsy (BMAT)* by hematologist to confirm the diagnosis. Anticoagulant, beta-blocker and hydroxyurea were started. His abdominal pain was resolved and platelet counts decreased in trend over time.

	1 st visit	2 nd visit	3 rd visit	Follow up 1	Follow up 2
Platelet (x10 ⁹ /L)	1234	774	864	479	557
ALT (U/L)	41.2	-	-	100.1	21.5
ALP (U/L)	117	-	111	86	53
GGT (U/L)	79	-	-	80	46
AST (U/L)	-	-	-	42.5	19
Amylase (U/L)	67	-	61	-	-
LDH (U/L)	-	308	-	-	-

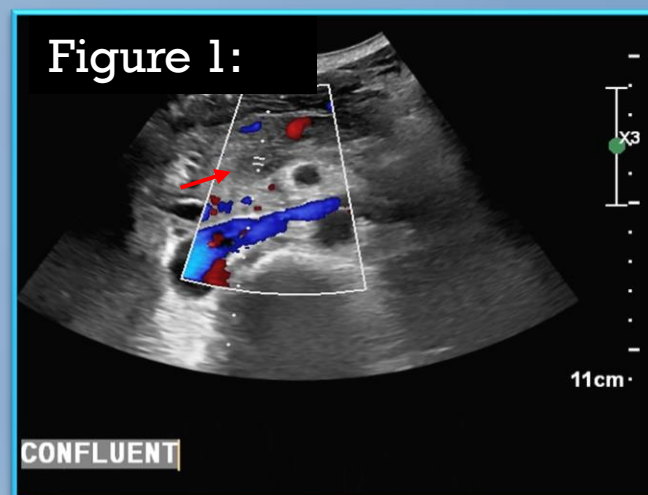


Figure 1: Power doppler image demonstrates no pulse wave and color doppler in portal confluence (arrow).

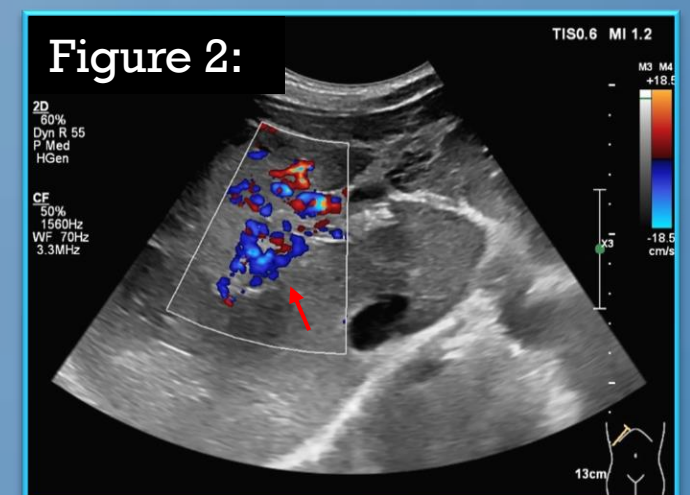


Figure 2: Ultrasound shows no color doppler within the portal vein with adjacent cavernous malformation and collaterals (arrow).

DISCUSSION

Myeloproliferative disorder (MPN) can be classified into polycythemia vera, essential thrombocytosis and myelofibrosis. The diagnosis requires positive **JAK2 V617F** mutation test and BMAT. MPN can cause PVT but it is rare. Typically, PVT occurs in patient with liver cirrhosis. The prevalence of non-cirrhotic PVT ranges from 0.7-3.7 per 100,000 populations. It is believed that hypercoagulable state (thrombocytosis) predisposes the venous system for thrombosis. As the thrombus builds up, portal venous shunting occurs as a result of portal hypertension. At this point, patient may have abdominal pain due to thrombosis and upper gastro-intestinal hemorrhage due to variceal bleed. Our patient was diagnosed as MPN likely essential thrombocytosis complicated with PVT and portal hypertension.

It is important to recognize and treat PVT early with anticoagulant to recanalize the portal vein to improve patient's prognosis. In chronic case like our patient, the aim is to control portal hypertension, treat MPN, and to prevent further unwanted complications such as variceal bleed.

One of the goals of the *Malaysian Emergency Medicine service* is to deliver an early, accurate and definitive emergency service including patient's disposition for ongoing treatment. Thus, it is crucial to practice a good clerking, examination and to brainstorm with clinical reasoning especially in patient with undifferentiated symptoms such as abdominal pain.

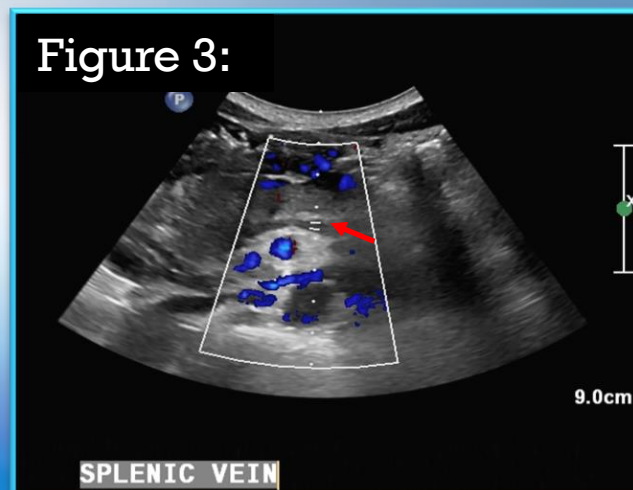


Figure 3: Doppler shows splenic vein thrombosis with no color flow seen (arrow).

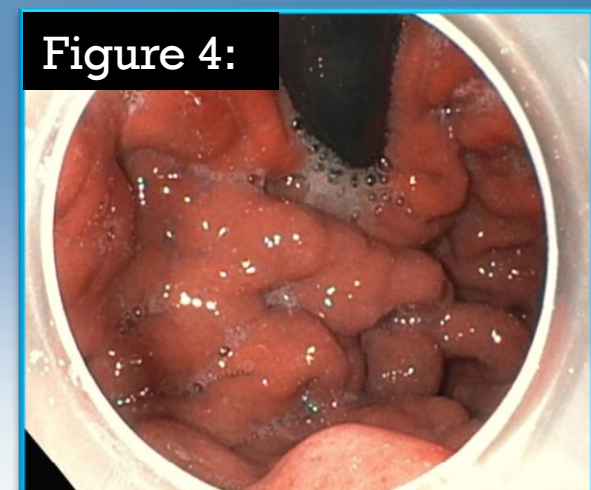


Figure 4: OGDS shows large esophageal varices with no stigmata of bleeding.

CONCLUSION

Life long education and developing competency is well accepted in medicine. Clinical reasoning requires understanding, and only by learning someone can acquire knowledge and skills. Considering this case, a good decision making skill is paramount importance after analyzing patient's history, with the involvement of correct in-depth physical examinations and supportive workups. In this case, early recognition, referral and prompt treatment are salient to stave off further complications.

ACKNOWLEDGEMENT

We would like to thank Emergency Department of Hospital UiTM for supporting us throughout the case report preparation. There is no conflict of interest.

References:

- Grinfeld J, Nangalia J, Baxter EJ, Wedge DC, Angelopoulos N, Cantrill R, et al. Classification and Personalized Prognosis in Myeloproliferative Neoplasms. *N Engl J Med.* 2018;379(15):1416-30.
- Gameiro AF, Robalo Nunes A, Guerra P, Mateus E, Fernandes F. Portal Vein Thrombosis Secondary to Occult Polycythemia Vera. *Eur J Case Rep Intern Med.* 2020;7(12):002003.
- Colaizzo D, Amitrano L, Tiscia GL, Scenna G, Grandone E, Guardascione MA, et al. The JAK2 V617F mutation frequently occurs in patients with portal and mesenteric venous thrombosis. *J Thromb Haemost.* 2007;5(1):55-61.