ARCHIVES OF HELLENIC MEDICINE: ISSN 11-05-3992

CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

ARCHIVES OF HELLENIC MEDICINE 2021, 38(1):139–140
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2021, 38(1):139–140

Surgery Quiz – Case 30

K. Boulas,
A. Paraskeva,
A. Triantafyllidis,
A. Hatzigeorgiadis

Department of General Surgery, General Hospital of Drama, Drama, Greece

An otherwise-healthy 71-year-old male patient with a known sizable recurrent left retroperitoneal malignant fibrous histiocytoma under pazopanib (*per os* 800 mg daily) with infiltration of the left ureter under double J stent and the descending colon causing incomplete large bowel obstruction, admitted to the emergency department with symptoms and signs of peritonitis. Abdominal computed tomography (CT) revealed the presence of: (a) a solid left retroperitoneal mass (approximate size16×10×12 cm), (b) infiltration of the left ureter with the presence of a double J stent, and (c) infiltration and perforation of the descending colon along with a large quantity of free intraperitoneal air and paracolic fluid (fig. 1). Emergency laparotomy performed which revealed the presence of descending colon perforation and disseminated feculent peritonitis. The patient submitted to left hemicolectomy with end transverse colostomy and intraoperative saline peritoneal lavage. Postoperatively, pazopanib discontinued and tinzaparin (subcutaneously 4,500 anti-Xa IU per day), omeprazole (intravenously [IV] 40 mg daily) and imipenem (IV 1 g q8h) were administered. Regarding surgical complications, postoperative period was uneventfull. On the 19th postoperative day, acute isolated severe thrombocytopenia (PLT <10×10^9/L) accompanied with hematuria occured. WBC count was normal, Hb count was 10.1 g/dL and regarding coagulation tests PT, aPTT, INR were normal, fibrinogen and D-dimmer were 198 mg/dL and 4.25 μg/mL, respectively. Peripheral blood smear showed no red blood cells and leukocytes morphological abnormalities. The patient was afebrile; blood and urine cultures were negative. Chest and abdominal CT revealed (fig. 2): (a) mild bilateral pleural effusion with bibasilar atelectasis, (b) no superficial or deep surgical site infection, (c) no thrombotic and hemorrhagic complications, and (d) progressive disease according to the Response Evaluation Criteria in Solid Tumors (RECIST) 1.1 criteria with rapid and massive increase in the primary tumor volume (approximate size 20×14×15 cm with a 2.5-fold volume increase). Bone marrow biopsy revealed increased number and size of megakaryocytes. The patient treated with combined prednisone (IV 1 mg/kg), intravenous immunoglobulins (IVIg) (400 mg/kg) and transfusion of one unit of apheresis platelets and two units of fresh frozen plasma (FFP) daily along with discontinuation of tinzaparin and imipenem with no response over a 6-day period. The patient gradually presented acute renal and respiratory failure.

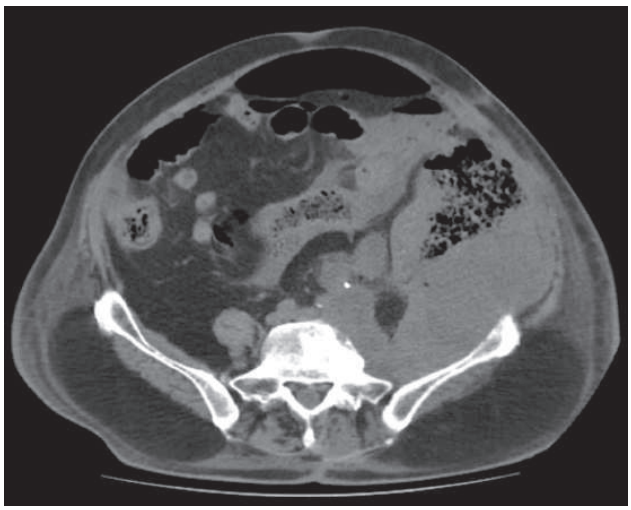


Figure 1

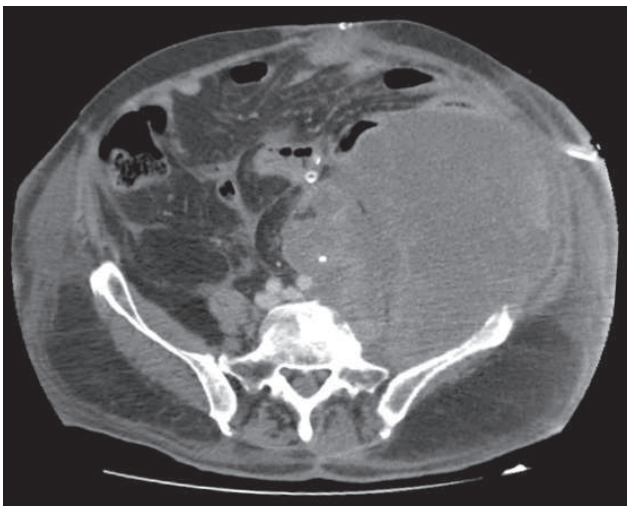


Figure 2

What was the most likely cause of thrombocytopenia?
(a) Sepsis- or drug-induced thrombocytopenia
(b) Heparin-induced thrombocytopenia (HIT)
(c) Immune thrombocytopenia purpura (ITP)
(d) Kasabach-Merritt syndrome

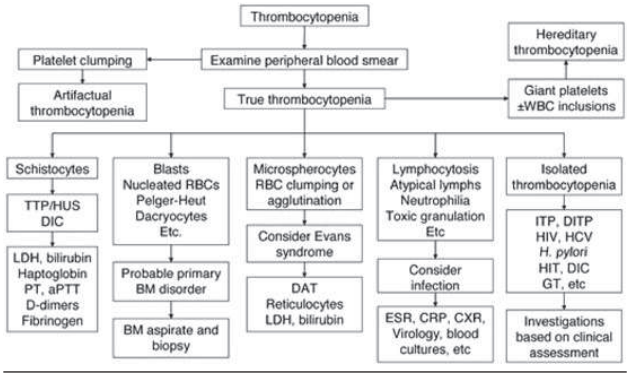
Comment

Drug- or sepsis-induced bone marrow decreased platelet production was excluded as: (a) no sepsis criteria were present, (b) blood and urine cultures were negative, (c) bone marrow biopsy revealed increased number and size of megakaryocytes. HIT-induced increased peripheral platelet destruction was excluded as the 4Ts score was 3 suggestive of low clinical probability for HIT and therefore HIT lab testing was not ordered. Increased peripheral platelet destruction due to ITP and Kasabach-Merritt syndrome were the more prominent diagnoses both relied on exclusion.

The diagnosis of newly diagnosed secondary (drug- or tumor-induced) ITP was supported by exclusion of other causes of thrombocytopenia (tab. 1) as: (a) complete blood cell count revealed isolated thrombocytopenia in the era of chronic disease anemia, (b) peripheral blood smear showed no red blood cells and leukocytes morphological abnormalities, and (c) bone marrow biopsy revealed increased number and size of megakaryocytes without other significant abnormalities. However, non response to initial treatment with corticosteroids and IVIg did not support the immune nature of ITP thrombocytopenia.

The diagnosis of Kasabach-Merritt syndrome was supported by: (a) the presence of the aggressive, rapidly enlarging and highly vascularized recurrent retroperitoneal tumor (fig. 2), and (b) the presence of thrombocytopenia and mild coagulopathy including slightly decrease in fibrinogen and slightly increase in D-dimmer with normal PT, aPTT and INR values. In our patient's case, surgical trauma and vascular endothelial growth factor receptor (VEGFR) inhibitor discontinuation resulted in rapid and massive primary tumor growth with 2.5-fold volume increase on repeated CT. The concomitant presentation of massive primary tumor growth along with profound thrombocytopenia and mild coagulopathy was suggestive of platelet trapping and fibrinogen consumption within the abnormal tumor vascular endothelial architecture and made Kasabach-Merritt syndrome the most probable diagnosis.

Table 1. Algorithm for workup of thrombocytopenia.



References

1. LIEBMAN HA. Thrombocytopenia in cancer patients. *Thromb Res* 2014, 133(Suppl 2):S63–S69
2. O'RAFFERTY C, O'REGAN GM, IRVINE AD, SMITH OP. Recent advances in the pathobiology and management of Kasabach-Merritt phenomenon. *Br J Haematol* 2015, 171:38–51

Corresponding author:

K. Boulas, Department of General Surgery, General Hospital of Drama, Drama, Greece
e-mail: boulaskonstantinos@gmail.com

Diagnosis: *ITP or Kasabach-Merritt syndrome; the most likely diagnosis seemed to be Kasabach-Merritt syndrome*