Twists and Turns: Unmasking Myeloproliferative Neoplasms Through Surgical Complications of Wandering Spleen

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INTRODUCTION

Myeloproliferative neoplasms (MPNs), particularly advanced myelofibrosis, often present with splenomegaly due to extramedullary hematopoiesis. This can lead to abdominal discomfort, early satiety, or cytopenias from sequestration of blood cells, though it may also be detected incidentally and occasionally precede the diagnosis of chronic myeloid leukemia (CML) or primary myelofibrosis (PMF). Massive splenomegaly may contribute to the development of a wandering spleen, a rare condition caused by weakened supporting ligaments, with torsion being its most serious complication.

CASES

Case 1

A 56-year-old man had a 15-year history of a left upper abdominal mass with recent postprandial pain. Imaging revealed an ectopically located, enlarged spleen with vascular torsion (Figure 1). CBC showed anemia, thrombocytosis, and dacryocytes. He underwent splenectomy, after which bone marrow biopsy confirmed primary myelofibrosis (WHO Grade 3) (Figure 2). Postoperatively, platelets rose to 1,378 × 10^9/L, controlled with hydroxyurea and anagrelide

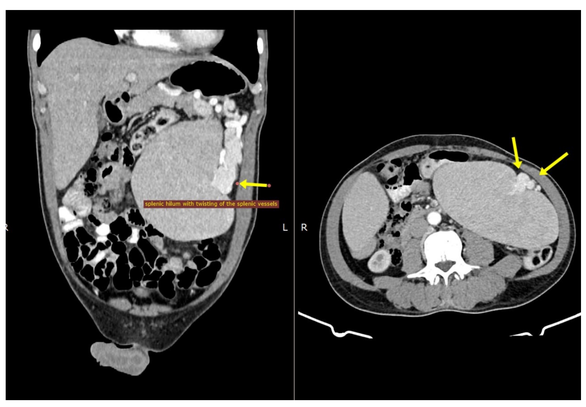

Figure 1

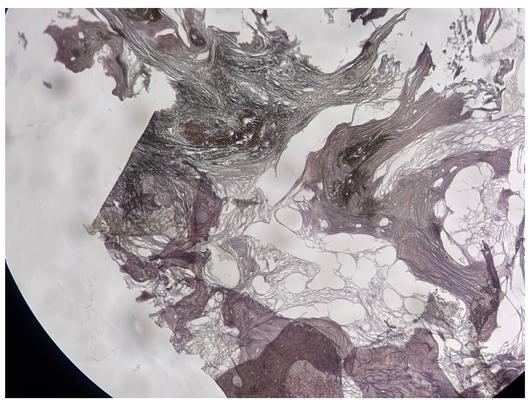

Figure 2

Case 2

A 26-year-old woman presented with abdominal distension and obstruction from massive splenomegaly. CT scan confirmed displacement of the spleen into the pelvis. (Figure 3) Laparotomy with bowel resection and colostomy was performed. (Figure 4) FISH confirmed CML (BCR::ABL1+), and imatinib therapy led to spleen reduction and blood count normalization.

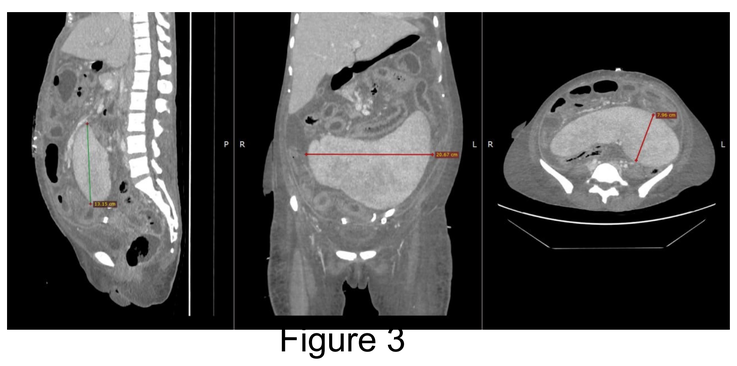

Figure 3

Figure 4

CONTACT

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DISCUSSION AND CONCLUSION

Physiologic Role of the Spleen

The spleen is a highly vascular organ essential for immune defense, clearance of aged blood cells, and storage of erythrocytes, reticulocytes, and platelets.

Splenomegaly in MPNs

In myeloproliferative neoplasms (MPNs), extramedullary hematopoiesis frequently causes massive splenomegaly. This can result in cytopenias from sequestration, abdominal discomfort, and rare but serious complications such as torsion, infarction, rupture, or bowel obstruction.

Splenic Torsion and Obstruction

Splenic torsion, though rare, may present with chronic pain or acute abdomen. Management options include detorsion with splenopexy or splenectomy when preservation is not feasible. Massive splenomegaly can also compress adjacent structures, occasionally leading to bowel obstruction.

Role of Splenectomy

In myelofibrosis, splenectomy is considered for refractory pain, portal hypertension, transfusion dependence, or severe thrombocytopenia, though it carries substantial risks. In chronic myeloid leukemia, splenectomy was historically used but now plays a limited role in the tyrosine kinase inhibitor (TKI) era, reserved mainly for palliation or select post-transplant complications.

CONCLUSION

Massive splenomegaly is a recognized complication of MPNs and may necessitate surgical intervention in select cases. Whenever possible, spleen-preserving strategies should be prioritized before vascular compromise or infarction occurs. Clinicians should maintain vigilance, as splenomegaly may precede abnormal blood counts. Careful evaluation with blood smear and bone marrow studies is essential to uncover underlying myeloid disease and guide timely management.

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