Idiopathic acute mesenteric venous thrombosis causing ischemic enteritis: A case report

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A R T I C L E   I N F O
Article history:
Received 14 June 2020
Received in revised form 13 August 2020
Accepted 15 August 2020
Available online 29 August 2020

Keywords:
Mesenteric venous thrombosis
Idiopathic
Infarction
Surgery

A B S T R A C T

INTRODUCTION: Though mesenteric venous thrombosis (MVT) causes bowel ischemia far less frequently
than arterial thrombosis, it still has the potential to cause life-threatening bowel infarction.

PRESENTATION OF CASE: Presented here is a case of idiopathic MVT of the superior mesenteric vein
and multiple distal venous branches causing diffuse peritonitis secondary to small bowel infarction in a
64 year old male. History and physical exam demonstrated severe persistent abdominal pain, hema-
tochezia, and diffuse abdominal tenderness to palpation with guarding. Venous filling defects and
segmental enteritis were noted on CT. The patient was treated with immediate IV heparin therapy with
subsequent laparotomy and excision of 45 cm of ischemic ileum. The patient had an uncomplicated recov-
ery. Post-operative thrombophilia screen was negative. The patient was discharged on indefinite warfarin
therapy.

DISCUSSION: MVT is often idiopathic in nature, with up to 49% having no identifiable cause. Risk factors
include abdominal inflammation and systemic thrombophilias. Importantly, bowel infarction is more
common with occlusion of more distal, smaller caliber mesenteric vessels. The standard of diagnosis is
contrast-enhanced abdominal CT, and management is prompt anticoagulation with surgical intervention
if severe. If the cause remains unclear, outpatient anticoagulation is continued indefinitely.

CONCLUSION: This case provides a valuable demonstration of several important MVT concepts –
specifically the high rate of idiopathic etiology, the need for indefinite anticoagulation in idiopathic cases,
and the increased risk of infarction in occlusion of smaller, more distal mesenteric veins.

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1. Introduction

Mesenteric venous thrombosis (MVT) is a comparatively rare cause of mesenteric ischemia. While arterial thrombosis accounts for the majority of mesenteric ischemia, venous thrombosis underlies only 6–9% of mesenteric ischemia cases. It is typically discovered either as an incidental finding on abdominal CT or during the workup for abdominal pain. Acute presentation is generally characterized by persistent, intense abdominal pain with progression to peritonitis and sepsis in more severe cases [1,2]. Treatment involves prompt and prolonged anticoagulation. Surgical resection of ischemic bowel is indicated in the most severe presentations [1,2]. Historically acute MVT has had a mortality rate of 44%, however in the more recent era of CT scans and aggressive anticoagulation mortality has dropped to 10–20% [3].

Here we present a case of severe acute MVT requiring emergent surgical intervention in a 64 year old African-American male with no known past medical history or prior predisposing factors for MVT.

This case report has been prepared in accordance with SCARE criteria [4].

2. Case presentation

A 64 year old Trinidadian male with no known significant medical or surgical history presented to the emergency department complaining of severe sharp, diffuse abdominal pain. The pain began eight days prior to admission with on-and-off pain. These symptoms were accompanied by anorexia, nausea, vomiting, and watery diarrhea with scant blood. The patient denied alcohol use, smoking, illicit drug use, and had no personal or family history of similar complaints, DVT or clotting disorders.

Pertinent physical exam findings included a distended abdomen with diffuse guarding to palpation. Laboratory tests revealed hemог-
raphy (CT) revealed severe wall thickening involving multiple small bowel loops consistent with a severe segmental enteritis; thrombosis of the SMV to the portal-splenic confluence with further thrombosis of several more distal mesenteric venous branches; and perihepatic, perisplenic, and pelvic ascites (Fig. 1).

The patient was started on IV heparin, piperacillin-tazobactam, and IV fluids. He was subsequently taken emergently to the OR for diagnostic laparotomy and exploratory laparotomy by the acute care surgery attending on call. A 45 cm segment of ischemic ileum was excised and sent to pathology. The bowel and mesentery had a woody appearance with significant lymphatic drainage and edema. The bowel appeared to have transmural ischemia. The bowel was left in discontinuity with temporary abdominal closure obtained via an ABThera® wound vac, in order to gain a second look and ensure that there was no ongoing ischemia from MVT. Time elapsed from arrival to the ER to arrival in the OR was 10 h and 21 min, as the patient required transfer to our higher-acuity facility from the initial hospital’s emergency department. The following day a successful anastomosis via side-to-side functional enterostomy was performed by the acute care surgery attending. IV heparin was resumed 4 h post-procedure with the addition of warfarin therapy until an INR of 2.0 was achieved.

Prior to discharge, the patient underwent postoperative evaluation by hematology-oncology, who performed a thrombophilia screen for anti-cardiolipin, anti-B2 glycoprotein, anti-phosphatidylserine, protein C deficiency, protein S deficiency, factor V Leiden mutation, and prothrombin G20210A mutation; all results were negative. Further workup in outpatient hematology was undertaken. The patient is now maintained on lifelong warfarin anticoagulation for idio-pathic MVT.

3. Discussion

3.1. Epidemiology

MVT accounts for 1:5000–15,000 of admissions and 1:1000 ED admissions, making it an uncommon symptomatic entity. Among causes of acute mesenteric ischemia, MVT accounts for only 6–9% of cases. Diagnosis of MVT is thought to have increased in the 21st century due to greater use of CT abdominal imaging. It is typically discovered either as an incidental finding on abdominal CT or during the workup for abdominal pain [1,2].
3.2. Risk factors and pathogenesis

Typical age of onset is 45–60 years with a slight preference for males. The most common risk factors include malignancy, thrombophilia, localized abdominal inflammation (eg. pancreatitis, inflammatory bowel disease), recent intra-abdominal surgery, and venous stasis (eg. cirrhosis, CHF). Of these, malignancy—especially myeloproliferative neoplasia associated with JAK2 mutations—is the most common cause of MVT secondary to hypercoagulability. Notably, 17–44% of MVT patients have a personal or family history of DVT. Thrombosis of the superior mesenteric vein (SMV) is more highly associated with localized abdominal pathology, while thrombosis of the distal venous branches are more highly associated with systemic prothrombotic states. Involvement of the IMV is rare, occurring in 0–11% of cases [1,2]. Despite the extensive list of risk factors, 21–49% of MVTs are idiopathic. This number is expected to decrease as thrombophilia screens become more sophisticated [1–3].

Congestion of the mesenteric veins secondary to thrombosis can lead to bowel ischemia, and in acute settings may precipitate transmural bowel infarction. Bowel ischemia of venous origin tends to produce a more gradual transition zone from normal to ischemic regions, in contrast to the sharp borders produced by acute arterial occlusion [1,2]. This gradual transition zone was demonstrated well upon pathologist examination of our patient’s excised small bowel segment. Transmural infarction may have grave consequences due to disruption of bowel mucosa and translocation of enteric bacteria to the peritoneal cavity [1,2]. Ileum and jejunum are by far the most commonly involved bowel regions [3].

3.3. Presentation

Acute MVT presents almost universally with abdominal pain, often out of proportion with physical exam. Occult fecal blood is present in 50% of cases. Increasing abdominal tenderness, distention, and ascites are worrisome for bowel ischemia, while progression to fever, peritoneal signs, and hemodynamic instability are concerning for bowel infarction and perforation. Anywhere from 6 to 29% of cases may lead to hemodynamic instability [1,2]. A number of case series conducted between 1994–2009 showed a weighted arithmetic mean incidence of peritonitis of 33.2% [3]. MVT of the more distal mesenteric veins tend to produce higher rates of bowel infarction versus MVT of the more proximal SMV and portal vein [1,2]. This may help explain the particularly acute nature of the case presented above.

3.4. Diagnosis

The mainstay of diagnosis is contrast-enhanced multidetector abdominal CT [1,2,5,6]. Labs are typically used not diagnostically but to evaluate for complications related to bowel ischemia. Lab findings may include hemococoncentration, non-specific leukocytosis, hyperamylasemia, and modest transaminitis. Hypoxemia, lactic acidosis, and positive blood cultures are associated with worse prognosis [1,2]. Findings on contrast-enhanced abdominal CT fall into three categories: circumferential bowel wall thickening, vascular changes, and non-mural/non-vascular signs. Circumferential wall thickening is the most common CT finding in acute MVT. Hypoattenuated thickened bowel wall is highly suggestive of venous bowel infarction. A “halo sign” indicates stratification of the bowel wall into layers of attenuation by a central zone of edema. Pneumatosis intestinalis may also be visualized in cases of advanced ischemia. Venous filling defects are apparent in over 90% of cases. Venous enlargement around the clot is more suggestive of acute MVT, while venous atrophy is more suggestive of chronic MVT. Mesenteric fat edema and ascites are particularly prominent in MVT as opposed to arterial thrombosis due to the congestive nature of the disease. Bowel dilatation secondary to ischemic ileus is also seen [5,6].

3.5. Management

Anticoagulant therapy is the mainstay of MVT management. Prompt and prolonged anticoagulation decreases mortality and hospital stay. Heparin therapy (or LWMH in appropriate nonoperative patients) is indicated. Heparin alone is continued until symptoms resolve and operative management becomes unlikely. Then, warfarin is initiated to an INR of 2–3; treatment lasts 6 months in patients with reversible MVT etiologies and indefinitely in idiopathic cases or irreversible prothrombotic states. Direct thrombin inhibitors and factor X inhibitors are likely also appropriate therapeutic choices, given prior reviews regarding use in portal venous thrombosis [7]. The choice of warfarin vs a novel oral anticoagulant is influenced by a patient’s renal function as well as financial considerations. Pain control, bowel rest, nasogastric decompression and fluid and electrolyte replenishment are provided as appropriate. Antibiotic therapy and surgical resection of ischemic bowel are reserved for patients with a high suspicion for bowel infarction, perforation, or peritonitis as seen in the presented case [1,2]. If viable bowel is seen on surgical exploration with evidence of total venous occlusion, thrombectomy with subsequent anticoagulation and a second-look laparotomy may be considered. With viable bowel and partial venous occlusion, anticoagulation with a second-look laparotomy is also an option [3]. From a logistical perspective, endovascular interventions for MVT are not a preferred approach, as access to the mesenteric venous system would require the performance of a transjugular intrahepatic portosystemic shunt (TIPS) procedure.

In the absence of an apparent predisposing factor for MVT, thrombophilia screens should be performed. Particularly important is screening for JAK2 sequence variations, which are commonly seen in myeloproliferative neoplasms. The incidence of gain-of-function JAK2 mutations in neoplasia-free patients with MVT ranges from 5 to 46% [1,2]. It is useful to draw blood for a thrombophilia screen prior to anticoagulation, as anticoagulants such as warfarin may cause artificial alterations of clotting factor levels [2].

Acute mortality from MVT varies from 0% to 20% which is decreased from 44% reported between 1966–2002 [3]. The most common cause of death is sepsis. Several factors increase short-term mortality risk: most commonly bowel infarction. Others include advanced age, lack of anticoagulation, management by a nonsurgical team, and colonic ischemia.

Long-term mortality increases with short bowel syndrome, recurrent thrombosis, and anticoagulation-related bleeding [1,2,8]. Recurrent MVT typically occurs within 30 days, at a rate as high as 25%. However, recurrence rates may be as low as 0–3% in patients receiving continuing anticoagulation. Rates of anticoagulation-related bleeding are less than 10% [1,2].

4. Conclusions

This is a case of acute MVT that is notable for its idiopathic origin and severity of presentation requiring urgent surgical intervention. It is important to remember that despite its association with prothrombotic states, it is not uncommon for acute MVT to “come out of the blue”. Also an important consideration is the fact that venous thrombosis causes bowel infarction at lower rates than arterial thrombosis, but in instances of distal venous thrombosis as in this case, life-threatening ischemia is possible.
**Declaration of Competing Interest**

The authors report no declarations of interest.

**Sources of funding**

No funding sources reported.

**Ethical approval**

The Crozer Keystone Healthcare System IRB does not require ethical approval of case reports per institutional standards and therefore, IRB approval was not sought and is exempt in this case.

**Consent**

Written informed consent was not obtained from the patient. The head of our medical team has taken responsibility that exhaustive attempts have been made to contact the family and that the paper has been sufficiently anonymised not to cause harm to the patient or their family. A copy of a signed document stating this is available for review by the Editor-in-Chief of this journal on request.

**Author contribution**

Andrew Samoyedny – record review, case report authorship.
Sai Sajja – advisor, case report authorship.
Asanthi Ratnasekera – surgeon in reported case, advisor, case report authorship.

**Registration of research studies**

This is a case report, not a research study.

**Guarantor**

Asanthi Ratnasekera.

**Provenance and peer review**

Not commissioned, externally peer-reviewed.

**Acknowledgements**

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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