

Case report

Acute superior mesenteric artery thrombosis in a 34-year-old male with thrombophilia: a case report

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Abstract

Introduction: Thrombophilic state, marked by extensive formation of blood clots within the vascular system poses a formidable peril, leading to loss of circulatory integrity and systemic well-being. Superior mesenteric artery (SMA) thrombosis is a serious medical disorder characterized by the obstruction of the arterial blood flow to the gastrointestinal tract, which is associated with significant morbidity and mortality. The condition has a plethora of etiological causes. Vague abdominal pain is a common clinical manifestation, and there is a very high mortality rate.

Case Presentation: A 34-year-old Pakistani male presented with rapidly worsening acute abdominal pain over a 10-hour period. Clinical examination revealed elevated blood pressure, while other vital signs remained within normal limits. Abdominal examination findings were inconclusive compared to the patient's severe abdominal pain complaint. Contrast-enhanced CT scan indicated a large thrombus in the superior mesenteric artery with radiological features suggestive of intestinal ischemia, extending from the duodeno-jejunal flexure to the mid-transverse colon. Subsequently, the patient underwent exploratory laparotomy with extensive bowel resection and received anticoagulant therapy. He subsequently experienced compensated right lower limb ischemia, treated conservatively with high-dose anticoagulant therapy. Within days, bowel anastomotic leak developed, requiring multiple surgeries. Eventually, the patient progressed to septic shock, followed by multiorgan failure, culminating in a fatal cardiac arrest.

Conclusion: Acute mesenteric ischemia, a rare abdominal emergency with an incidence of 6 per 100,000, is associated with a substantial mortality rate, ranging from 58% to 93% among diagnosed cases. The challenges posed by SMA thrombosis include diagnostic complexities, as it frequently manifests with symptoms disproportionate to clinical signs, and results in catastrophic complications, necessitating high suspicion index and prompt intervention.

Keywords: Mesenteric Ischemia, Thrombosis, Ischemia, Abdominal Pain

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Introduction

Thrombophilic state is characterized by the extensive development of blood clots within blood vessels, leading to the potential blockage or complete interruption of blood circulation. This can manifest in different regions of the body, such as the deep veins of the legs [known as deep vein thrombosis (DVT)], arteries, or critical organs like the heart and brain [1].

The superior mesenteric artery (SMA) is a branch of the abdominal aorta responsible for blood supply of the midgut, extending from the second part of the duodenum or the duodeno-jejunal (DJ) flexure to the junction between the right 2/3 and the left 1/3 of the transverse colon [2]. The colon is primarily supplied by the distal branches of the superior mesenteric artery, specifically, the right colic, ileocolic, and middle colic arteries, while the distal colon from the distal 3rd of the transverse colon downwards is supplied by the inferior mesenteric artery [3].

SMA thrombosis is a critical medical condition characterized by the blockage of the arterial blood supply to the small and large intestines. According to a systematic review and meta-analysis, acute mesenteric ischemia (AMI) is a rare condition, the incidence being 0.0062% [4]. This condition is severe and potentially life-threatening. Several etiological factors can lead to SMA thrombosis, including atherosclerotic occlusion, embolic occlusion, thrombophilia, hypercoagulable states, infections, vasculitis, mesenteric dissection, or the development of a mycotic aneurysm [5].

In a clinical context, severe abdominal pain is the most frequently reported symptom, with 80% of patients experiencing it characteristically when abdominal pain is out of proportion when compared to the clinical signs. Other symptoms, in descending order of occurrence, include vomiting (41%), nausea (34%), constipation (23%), abdominal distention (21%), fever (19%), diarrhea (15%), and rectal bleeding (hematochezia) in 9% of cases. As a complication of thromboembolic phenomena, infarction and ischemia of other systems can occur, for example cerebral infarction, limb ischemia etc. [6]. Acute thromboembolic occlusion of the SMA is a condition associated with a poor prognosis [7]. Despite notable advancements in medical diagnosis and treatment options in the last four decades, mesenteric vascular occlusion continues to have an unfavorable sequela,

with in-hospital mortality rates ranging from 59% to 93% [8].

The potential differential diagnosis for mesenteric artery thrombosis is extensive and includes conditions such as acute colitis, bowel obstruction, abdominal aortic aneurysm, biliary obstruction, intestinal perforation, and malignancies [5].

Case Presentation

A 34-year-old Pakistani male, with a medical history significant for left nephrolithiasis, presented to the Emergency Room (ER) with sudden onset of severe, progressively worsening epigastric pain lasting for 10 hours. Subsequently, the pain diffused throughout the abdomen and was accompanied by nausea and four episodes of vomiting.

Upon examination, vital signs were stable except for elevated blood pressure (174/93 mmHg). Abdominal assessment revealed a soft, lax abdomen with mild tenderness in the epigastrium and the right iliac fossa, along with mild rebound tenderness. No palpable masses were detected, and bowel sounds were audible.

Initial laboratory results showed elevated inflammatory markers, with a total white blood cell count of 14,000/uL and a C-reactive protein level of 10 mg/dL, while other labs were within normal limits (Table 1). An initial computerized tomography scan of the kidneys, ureters and bladder (CT KUB) demonstrated the persistence of left renal stones and mild left hydronephrosis.

Initially admitted under urology care, a general surgery consultation was sought due to the emergence of abdominal tenderness. A repeat CT with intravenous contrast revealed features suggestive of acute small bowel ischemia, attributable to arterial occlusive thrombus, observed 2.5 cm beyond the origin of SMA, extending into the proximal SMA, along with a floating thrombus observed in the abdominal aorta [Figure 1: A, B]. Whole body CT did not reveal any evidence of any mass in the body. Echocardiography revealed no abnormalities.

The patient was commenced on treatment with high-dose enoxaparin (60 mg BID) and a heparin infusion (1000 units/hour) for anticoagulation. Plans for an endovascular intervention were abandoned due to

concerns about the elapsed time. Subsequently, a diagnostic laparoscopy was performed, revealing evidence of ischemic bowel extending from the DJ flexure to the mid transverse colon. The procedure was converted to an open laparotomy, involving the excision of the non-viable necrotic small and large bowel sections, followed by side-to-side bowel anastomosis between the transverse colon and the 4th part of the duodenum. Microscopic examination of the resected bowel revealed extensive infarcted gangrene consistent with arterial thrombosis.

On the fifth postoperative day, the patient developed right leg pain, erythema, and edema, with non-palpable pulses in the dorsalis pedis and popliteal arteries. CT angiogram revealed complete blockage of the right internal iliac artery and occlusion of the distal segment of the right superficial femoral artery, leading to acute compensated right lower limb ischemia (Figure 2: A,

B). Conservative management by the vascular surgeon included anticoagulation, analgesia, and vigilant limb monitoring for possible decompensation.

By the eighth postoperative day, the patient exhibited signs of acute abdomen, fever (38°C), and drainage of 100 mL of pus from the right drain. A second exploratory laparotomy revealed an abdomen filled with more than 2L of feculent fluid and colo-duodenal anastomosis dehiscence. The wound was left open after peritoneal lavage, with subsequent saline wash procedures.

The patient developed septic shock, resulting in refractory hyperkalemia, cardiac arrest, and eventual demise. Hematological consultation was sought to investigate the hypercoagulability state, but despite specialized hematological investigations, no definitive diagnosis was reached.

Table 1: Laboratory investigations

Investigations	Result	Normal Range	Timing of the results
White Blood Cell (WBC) count	14 x 10 ³ /mcl	4-10 x10 ³ /mcl	Day 1
Hemoglobin (Hgb)	14.1 g/dl	13.0-18.0 g/dL	Day 1
Hematocrit (Hct)	41.2%	37.0-49.0%	Day 1
Platelet count	239.00 x 10 ³ /mcl	150-400 x 10 ³ /mcl	Day 1
Sodium	141 mmo/L	135-145 mmol/L	Day 1
Potassium	3.10 mmol/L	3.4-5.0 mmol/L	Day 1
Creatinine	113.7 µmol/L	61.9 to 114.9 µmol/L	Day 1
Blood Urea Nitrogen (BUN)	4.9 mmol/L	2.9-8.9 mmol/L	Day 1
Estimated Glomerular Filtration Rate (eGFR)	63 ml/min/1.73m2	90 to 120 mL/min/1.73 m ²	Day 1
C-Reactive Protein (CRP)	10 mg/dL	< 0.3 mg/dL	Day 1
Partial Thromboplastin Time (PTT)	34.7 sec	25 to 35 sec	Day 1
International Normalised Ratio (INR)	1.1	<1.1	Day 1
Random Blood Glucose	93 mg/dL	70 - 100 mg/dL	Day 1
Glycated Hemoglobin A1C (HbA1C)	5.1%	4% - 5.6%	Day 1

Total cholesterol	153 mg/dL	125 – 200 mg/dL	Day 1
Non-HDL cholesterol	102 mg/dL	<130 mg/dL	Day 1
LDL cholesterol	68 mg/dL	<100 mg/dL	Day 1
HDL cholesterol	70 mg/dL	>40 mg/dL	Day 1
Rapid Plasma Reagin (RPR)	Non-reactive	Non-reactive	Day 3
Fibrinogen	9 g/L	2.0 to 4.0 g/L	Day 1
D- dimer	2.7 mg/L	<0.50 mg/L	Day 1
Anti-thrombin III	117.6%	80% to 120%	Day 3
Protein C	125.0%	70%- 150%	Day 3
Protein S	135.0%	60% -150%	Day 3
Factor V Leiden mutation	Negative	Negative	Day 5
Double stranded DNA (dsDNA)	Negative	Negative	Day 5
Extractable nuclear antigen (ENA)	Negative	Negative	Day 5
Antineutrophil cytoplasmic autoantibodies (ANCA)	Negative	Negative	Day 5
Janus Kinase gene 2 (JAK2) V617F mutation	Negative	Negative	Day 7
C3	98 mg/dl	75 - 175 mg/dl	Day 3
C4	25 g/dL	15 to 45 g/dL	Day 3

Discussion

Despite the fact that acute mesenteric ischemia is a rare abdominal emergency (incidence being 6.2/100,000 persons), it has a significant mortality rate (between 59% to 93%). Among the key etiologic causes of acute mesenteric ischemia are Behcet disease, protein C, protein S deficiency, atherosclerosis, thrombocytosis, malignancies that promote thrombotic tendency, and antithrombin III deficiency [8]. But it can also be idiopathic, with inability to reach a conclusive hematological diagnosis, which restricts the treatment to only anticoagulation without any definitive treatment.

Patients may present with nonspecific complaints, but commonly present with rapid onset abdominal pain, which is out of proportion when compared to clinical findings. Laboratory investigations may be normal but high lactic acid and d-dimer may be encountered. Although direct abdominal X-rays and ultrasonography

can aid in diagnosis, angiography is the gold standard to determine the exact location of the vascular obstruction and the state of collateral circulation [9].

Thrombotic or embolic etiologies can be the cause of arterial thromboembolic events in non-cancer patients. The embolic etiologies being atherosclerosis of the aortic arch, aneurysms (carotid, aortic, popliteal), atrial fibrillation, acute myocardial infarction, mitral stenosis, valvular prosthesis, infective endocarditis, non-bacterial thrombotic endocarditis. Atherosclerosis, shock/low flow state, thrombophilia, antiphospholipid syndrome, protein C and S deficiency, antithrombin deficiency, and heparin-induced thrombocytopenia are among the thrombotic etiologies [10].

The patient's young age, as well as the absence of carotid/abdominal bruit and normal LDL cholesterol levels made atherosclerosis implausible, and echocardiography revealed no abnormalities. The hematological data for our patient indicated normal

platelet counts, and hematological profiles were also normal, other than raised fibrinogen and D-dimer levels, attributable to inflammation. The full blood count parameters were not in keeping with myeloproliferative disease, and additionally, JAK 2 negativity proved polycythemia vera unlikely. Since the levels of Antithrombin III, Protein C, and Protein S were likewise normal, it was determined that these inadequacies were not the cause of the extensive thrombosis. Rapid Plasma Reagin (RPR), done to rule out syphilis (as a cause of aortitis) was also non-reactive. The surgical pathology report also revealed evidence suggestive of gangrene of the bowel wall consistent with arterial thrombosis, with no atherosclerotic changes in the vessel walls and no evidence of vasculitis, ruling both these diagnoses out.

The reported case is intriguing because of the involvement of multiple large arteries throughout the body without any plausible cause and without any identifiable risk factors. This is one of the few, if any such reported cases,

To the best of our knowledge this is the first case of extensive arterial thrombosis that has been documented that does not have a plausible cause or recognizable risk factors. An idiopathic floating ascending aortic thrombus was the cause of a patient presenting with acute abdominal pain, according to a case report found on PubMed [11]. However, no other reports of the condition's co-occurrence included superior mesenteric and superficial femoral artery occlusion.

If there is a possibility of acute mesenteric ischemia, prompt revascularization and emergency angiographic imaging can save lives. The recommended courses of treatment include surgical bypass and/or endovascular balloon angioplasty. Because endovascular procedures have fewer complications than surgical treatment, they have grown in popularity recently. For mesenteric ischemia, mesenteric bypass surgery is a preferable long-term therapy option [12].

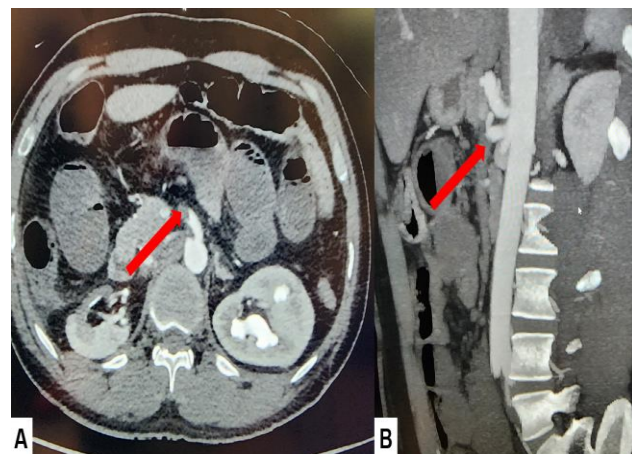


Figure 1: A. CT Abdomen w/ contrast (axial) demonstrating occlusion at the Superior Mesenteric Artery (red arrow), B. CT Abdomen w/ contrast (sagittal) demonstrating occlusion at the Superior Mesenteric Artery (red arrow)

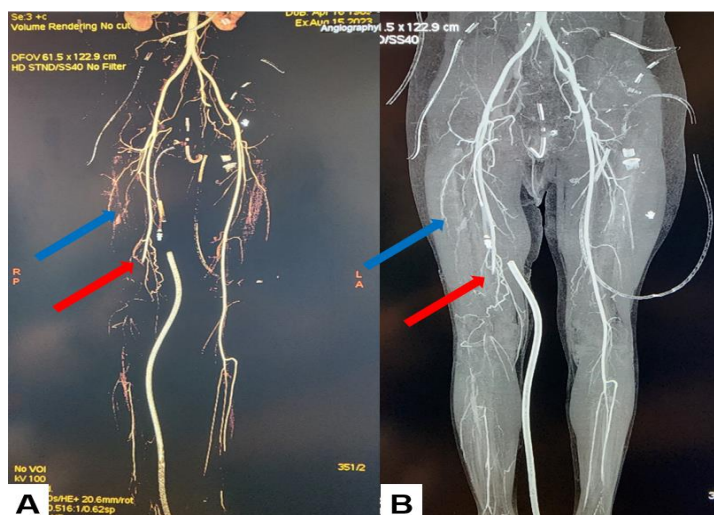


Figure 2: A. CT Angiography (volume rendered) demonstrating occlusion of the right Internal Iliac Artery (blue arrow) and right Superficial Femoral Artery (red arrow), B. CT Angiography demonstrating occlusion of the right Internal Iliac Artery (blue arrow) and right Superficial Femoral Artery (red arrow)

Conclusion:

Since cases of massive thrombosis, including AMI and acute limb ischemia (ALI) in the younger population are uncommon, inquiries into the underlying cause should be made. An intricate, multidisciplinary team is required to diagnose and treat unexplained arterial thrombosis, while maintaining a high suspicion index to reach the diagnosis early and prevent the catastrophic complications. It is necessary to conduct an evaluation in a methodical manner, taking into account both common etiologies and unusual ones. Management is based mostly on patient preference and patient-specific thrombotic and bleeding risk, and therefore necessitates a review of the literature that is currently accessible for certain vascular areas or disorders. It is also crucial to maintain continuous monitoring and to regularly reevaluate the treatment plan because new information or symptoms may emerge over time, as well as new risk factors for bleeding and thrombosis. Clinical protocols and indications must be followed when doing medical therapy and revascularization techniques.

Ethical Approval:

Not applicable as this is a case report.

Human and Animal Guidelines:

Not applicable.

Consent for publication:

Informed written consent was obtained from the patient before his unfortunate demise.

Abbreviations:

SMA: Superior Mesenteric Artery

ER: Emergency Room

BP: Blood pressure

DJ Flexure: Duodenojejunal flexure

CT angiogram: Computed Tomography angiogram

AMI: Acute Mesenteric Ischemia

ALI: Acute Limb Ischemia

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