

Isolated Superior Mesenteric Artery Thrombosis

A Rare Cause for Recurrent Abdominal Pain in a Child

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Abstract

A 4-year-old boy was evaluated for recurrent abdominal pain and failure to thrive over a 1-year period in a pediatric subspecialty clinic. Results of the extensive workup mostly were unremarkable. Eventually, imaging studies of the abdominal aorta revealed an isolated thrombosis of the superior mesenteric artery trunk and compensatory hypertrophy of the inferior mesenteric artery. He had been having abdominal angina symptoms and fear of eating. A detailed family history suggested a possible hypercoagulable state. However, an extensive hematologic evaluation did not reveal a recognizable defect that could produce thrombotic events. He was treated by arterial graft bypass surgery and started on conventional anticoagulants. Several months later, he developed repeat, near-total thrombosis of the graft with recurrence of his symptoms. After balloon dilation of the graft and starting him on appropriate anticoagulant maintenance regimen, he had good symptom relief, and the graft remained patent. This presentation was unusually prolonged for the type of vascular problem identified. The possibility of vascular problems in children, therefore, should be considered. Unidentified cause of hereditary clotting tendency is another challenging aspect of this case.

Key Words: Superior mesenteric artery thrombosis—Recurrent abdominal pain—Children.

bleeding, vomiting, severe or acute abdominal pain, and obvious abnormalities on physical examination or initial screening tests. Organic causes of recurrent abdominal pain in children are numerous, with some more common than others. Reasonable stepwise approach to such a clinical presentation has been emphasized in the pediatric gastroenterology literature, and most organic causes can be identified in the process. Diagnostic dilemmas often arise in the following scenario: the physician needs to ensure that the patient does not have any of the multiple potential organic causes of abdominal pain; the physician also needs to limit the workup to appropriate studies to avoid unnecessary exposure to radiation and to prevent further escalating the cost of health care while considering the increasing limitations of managed care. However, when a common symptom has an uncommon cause, it may be difficult to recognize initially and requires more than the usual steps. We present the case of a child experiencing recurrent abdominal pain with an unusual cause.

CASE REPORT

A 4-year-old boy had a 4-month history of recurrent midabdominal pain, refused to eat, and demonstrated poor growth. He reported abdominal pain daily that usually was periumbilical, usually occurred shortly after meals, and sometimes was associated with vomiting. The vomitus was not bloody or bilious and occurred two to three times a week. He increasingly refused his food with a resultant 1.8-kg weight loss. There were no other behavioral problems. He had no history of constipation, diarrhea, fever, or frequent respiratory problems. He had been given acetaminophen and antacids with little relief. He had no significant medical or surgical history. There was no family history of inflammatory bowel disease, acid peptic disorder, clotting abnormalities, or connective tissue disorders.

His physical examination revealed a thin child with wasted buttocks who otherwise was normal. His weight was 12.7 kg (<5%); height, 99 cm (10–25%); blood pressure, 88/52 mm Hg; pulse, 90 beats per minute; and temperature, 36°C. Examination of the head, ears, eyes, nose, and throat; neck; chest; heart; extremities; central nervous system; and skin all produced results within normal limits. His abdominal examination revealed mild tenderness over the epigastrium, and there were no abdominal bruits or palpable masses. Rectal examination revealed no perianal lesions and only a small amount of soft stool, which was negative for occult blood.

He disappeared from follow-up and missed the scheduled 6-month workup without explanation. When he returned, he had gained 2 kg yet reported that his midabdominal pain and occasional vomiting had continued and increased.

Recurrent abdominal pain is a common problem in childhood with many possible causes. Its evaluation is challenging, even to the most experienced clinicians. Careful assessment and close attention to details often are the most important aspects in finding the true cause in many cases. Functional abdominal pain is one of the commonest causes for chronic recurrent abdominal pain in children, particularly if the pain is periumbilical, is associated with altered bowel pattern, and seldom awakens the child from sleep. Result of physical examination usually is unremarkable, and reasonable screening tests give reassuringly normal findings. These tests usually include blood count, sedimentation rate, routine chemistry with liver profile, and urinalysis and simple stool tests for occult blood and parasites. Recognizable “red flags” for possible organic cause include young age at onset, associated weight loss or failure to thrive,

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Results of this workup were as follows: complete blood count was unremarkable except for mild eosinophilia (absolute eosinophil count, 704 cells/ μ L; reference range [RR], 50–600), erythrocyte sedimentation rate was 7, and his serum chemistry profile was unremarkable except for low bicarbonate level (14; RR, 20–28). His urine pH was inappropriately high (>8.5). Therefore, he was believed to have renal tubular acidosis, and a referral to a pediatric nephrologist was recommended. His serum lipase and amylase and his liver enzymes all were within normal limits. His stool tested negative for ova and parasites, his sweat chloride test was borderline abnormal (37; RR, 0–40), and the markers for *CF* gene mutations were negative. Tests for celiac markers, antigliadin, antireticulin, and antiendomysial antibodies also gave negative results. His serum total IgE was somewhat elevated (90; RR, <50), but results of a radioallergosorbent test for multiple food allergens were negative. Findings on plain abdomen radiographic study were within normal limits. Screening results of abdomen ultrasound study of the liver, gall bladder, kidneys, and pancreas were unremarkable. An upper gastrointestinal endoscopy study with biopsy was done and revealed mild chronic duodenitis, lymphoid aggregates in the gastric mucosa, and chronic reflux esophagitis with some submucosal hemorrhage. He was started on treatment with ranitidine, metoclopramide, and daily PediaSure formula supplement (Ross Labs, Abbot Park, IL). He continued to refuse food as before and continued to lose weight over the next 4 weeks; his abdominal pain persisted. A 2-week trial of therapy with the proton pump inhibitor, omeprazole, also was equally ineffective.

Because of the persistence of his symptoms, a computed tomography scan of the abdomen with intravenous contrast was performed, which revealed abnormal appearance of the superior mesenteric artery (SMA) trunk with a low-density mass and calcification within it, suspicious for thrombosed SMA. There also was a compensatory enlargement of the inferior mesenteric artery to approximately 25% of the size of his aorta. A subsequent magnetic resonance angiography study was done (Fig. 1), which revealed a focal segmental proximal occlusion of the SMA near its origin and resultant hypertrophy of the inferior mesenteric artery with a well-developed arch of Riolan on the left and a prominent marginal artery of Drummond seen along the region of the ascending colon on the right. The distal SMA seemed to be reconstituted, likely through the collateral flow. There was an incidental duplicated left renal artery.

Additional family history then was elicited and revealed several female relatives who had deep venous thrombosis, a young relative who had a stroke, and another relative who had an arterial thrombosis of the lower limb requiring amputation. No known risk factor was identified.

Detailed hematologic evaluation then was done to rule out hypercoagulable state, and normal values were obtained for tests on levels of homocysteine, protein S, protein C, activated protein C, antithrombin 3, factor VII, factor V (Leiden), anticardiolipin (antiphospholipid), anti-double-stranded DNA; lipid profile; and analysis for the prothrombin 20210 mutation.

The patient had a visceral angiogram that confirmed the diagnosis of proximal SMA thrombosis with collaterals feeding its distal branches. He then underwent surgical exploration with subsequent arterial graft to bypass the thrombosed part of the SMA. There was no other reported vascular thrombosis or malformation noted during the surgical exploration. In the absence of any specified hypercoagulable state, he also was started on the oral anticoagulant warfarin. He did well with resolution of his abdominal pain and fear of eating, and, subsequently, correction of the initial biochemical abnormalities for 6 months postoperatively. He then developed recurrence of his abdominal pain and was readmitted with



FIG. 1. Magnetic resonance angiogram of the abdominal aorta showing abrupt occlusion in a proximal segment of the superior mesenteric artery trunk (arrow) and distal filling by hypertrophied collateral circulation from the inferior mesenteric artery.

near-total thrombosis of the SMA graft. This was managed by angiographic balloon dilation. On readmission, the international normalized ratio was within therapeutic range. He then was switched to maintenance with low molecular weight heparin and was doing well for at least 2 months after discharge.

DISCUSSION

Thrombosis or occlusion of the SMA has been reported in the medical and surgical literature as a cause of acute abdominal pain and catastrophic intestinal ischemia or infarction.^{1–7} However, most of the reported cases were of adults with advanced arteriosclerosis⁷ or recurrent embolus formation in the setting of chronic atrial fibrillation⁷; were associated with clotting dysfunction or generalized thrombotic events, as in antiphospholipid syndrome⁴; or were related to vasculitis⁸ or fibromuscular dysplasia.⁹ There is one published case report on a child with acute thrombosis of the SMA that was related to abdominal sepsis and perforated appendicular abscess⁵ and another report about neonatal presentation of SMA thrombosis complicating congenital nephrotic syndrome of the Finnish type with a fatal outcome.⁶ Hamed and Ghandour⁹ report a case of a 5-year-old child with acute abdominal pain who had catastrophic gangrene of the entire small bowel. The child was diagnosed with fibromuscular dysplasia of the SMA and had a fatal outcome within weeks.⁹ Abdominal angina with severe postprandial pain typically is seen in older patients

with advanced atherosclerosis of the mesenteric vessels and may respond to surgical treatment.¹⁰ Superior mesenteric artery thrombosis has been reported in systemic vasculitis with isolated onset in abdominal vessels.⁸ In our patient, the pathohistologic features of the resected portion of the thrombosed SMA showed no evidence of vasculitis or fibromuscular dysplasia.

This is the first published case report of an isolated SMA thrombosis in an otherwise healthy child in the absence of any of these known risk factors, with this unique chronic presentation and protracted symptoms of recurrent abdominal pain and growth failure over a full year with a subsequent successful outcome. This may represent an unrecognized hereditary hypercoagulable tendency. Further evaluation in the patient and affected family members may be needed.

As mentioned earlier, the diagnosis was suspected once contrast abdominal computed tomography was performed, which seems to be a reliable method of detecting this condition.¹¹ However, in our patient, subsequent delineation of the problem and its associated vascular changes was made using magnetic resonance angiography. Contrast computed tomography of the abdomen needs to be considered in the evaluation of significant abdominal pain in children, even in the absence of radiologic abnormalities on initial ultrasound evaluation.

Metabolic acidosis with relatively alkaline urine in our patient initially was believed to reflect renal tubular acidosis but probably was related to bowel ischemia and recurrent lactic acidosis, considering that the chemical abnormality of the serum and urine both corrected after the surgical correction. The peripheral eosinophilia may have been related

to the same cause, but a proposed mechanism for this finding is less well understood. Mildly elevated eosinophils often are seen during allergy season in Oklahoma.

This diagnosis of SMA thrombosis is not likely to become a common differential diagnosis in the workup of young children with recurrent abdominal pain, but it needs to be recognized when it is present.

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REFERENCES

1. Sharma BD, Singh RP. Thrombosis of the mesenteric artery [case report]. *Indian Pract* 1966;19:317-8.
2. Glicic V, Sindjic M, Tasic D, et al. [Contribution to the study of the etiology of thrombosis of the superior mesenteric artery: report of a case]. *Srp Arh Celok Lek* 1968;96:435-40.
3. England RJ, Woodcock B, Zeiderman MR. Superior mesenteric artery thrombosis in a patient with the antiphospholipid syndrome. *Eur J Vasc Endovasc Surg* 1995;10:372-3.
4. Hamilton ME. Superior mesenteric artery thrombosis associated with antiphospholipid syndrome. *West J Med* 1991;155:174-6.
5. Al Neimi K, Boulet E, Imbaud P, et al. [Thrombosis of the superior mesenteric artery and perforation of the appendix in a young girl]. *Ann Chir* 1990;44:524-6.
6. Tsai MJ, Wu TJ, Teng RJ, et al. Mesenteric arterial thrombosis complicating congenital nephritic syndrome of Finnish type: report of one case. *Zonghau Min Guo Xiao Er Ke Yi Xue Hui Za Zhi* 1995;36:445-7.
7. Altman KA. Superior mesenteric artery occlusion: a review and case report. *Am J Gastroenterol* 1971;55:152-7.
8. Chang YJ, Kerr LD. Isolated abdominal vasculitis as an atypical presentation of Wegener's granulomatosis. *Am J Gastroenterol* 2000;95:297-8.
9. Hamed RM, Ghandour K. Abdominal angina and intestinal gangrene, a catastrophic presentation of arterial fibromuscular dysplasia: case report and review of the literature. *J Pediatr Surg* 1997;32:1379-80.
10. Danesh S. Abdominal angina and mesenteric insufficiency. *Angiology* 1979;30:281-3.
11. Salazano A, De Rosa A, Carbone M, et al. [Computerized tomography features of intestinal infarction: fifty-six surgically treated patients of which 5 with reversible mesenteric ischemia]. *Radiol Med (Torino)* 1999;97:246-50.