CASE REPORTS/CLINICAL VIGNETTES

Giant Cell Arteritis Presenting as Small Bowel Infarction

Aniyizhai Annamalai, MD, Mark L. Francis, MD, Sriya K. M. Ranatunga, MD, MPH, and David S. Resch, MD

Department of General Internal Medicine, Southern Illinois University School of Medicine, P.O. Box 19636, Springfield, IL 62794-9636, USA.

Giant cell arteritis predominantly affects cranial arteries and rarely involves other sites. We report a patient who presented with small bowel obstruction because of infarction from mesenteric giant cell arteritis. She had an unusual cause of her obstruction and a rare manifestation of giant cell arteritis. In spite of aggressive therapy with steroids, she died a month later because of multiple complications. We discuss the diagnosis and management of small bowel obstruction and differential diagnosis of vasculitis of the gastrointestinal tract. We were able to find 11 cases of bowel involvement with giant cell arteritis in the English literature. This case report illustrates that giant cell arteritis can be a cause of small bowel obstruction and bowel infarction. In the proper clinical setting, vasculitides need to be considered early in the differential diagnosis when therapy may be most effective.

KEY WORDS: giant cell arteritis; temporal arteritis; small bowel infarction; vasculitic disorders.

BACKGROUND

In this case report, we describe a patient who presented with small bowel obstruction. She had an unusual cause of her obstruction, small bowel infarction from giant cell arteritis, and a rare manifestation of giant cell arteritis. Most typically, giant cell arteritis predominantly affects cranial arteries, which leads to headache, scalp and temporal artery tenderness, jaw claudication, and visual disturbances. Systemic symptoms of fever, fatigue, weight loss, myalgias are present in half of patients. Less commonly, it can present as neuropathies, transient ischemic attack, or stroke. Rare manifestations of giant cell arteritis include limb claudication, aortic dissection, myocardial infarction, pericarditis, tongue necrosis, interstitial lung disease, breast mass, or bowel infarction.[1,2]

The present case reminds us, in the proper setting, to consider giant cell arteritis along with other vasculitic disorders in the differential diagnosis of small bowel obstruction.

CASE REPORT

A 78-year-old white female with a past medical history significant for migraine headaches, 2 previous abdominal surgeries, and carotid artery disease, presented to the emergency department with a 2-week history of lower abdominal pain and nausea as well as a 4-day history of obstipation. At this time, she denied a new headache or visual symptoms. On examination, she was tachycardic with a heart rate of 126/min and hypertensive with a blood pressure of 151/76 mmHg. Her oral mucosa was dry. Her left pupil was round and reactive to light, while the right was irregular secondary to a recent cataract surgery. Funduscopic examination was not done. She had a diffusely tender abdomen, more so, in the periumbilical region and in the left lower quadrant. Bowel sounds were decreased. The rest of the physical exam was normal. The white blood cell count was elevated at 25.2 k/cumm. The platelet count was mildly elevated at 484 k/cumm. The hemoglobin and hematocrit were normal at 12.7 g/dL and 39%, respectively. Serum lactic acid was normal at 1.1 mg/dL. Other laboratory values were also normal with sodium, 133 meq/L; potassium, 3.5 meq/L; chloride, 90 meq/L; bicarbonate, 30 meq/L; blood urea nitrogen, 17 mg/dL; creatinine, 0.7 mg/dL; alkaline phosphatase, 86 U/L; bilirubin, 0.6 mg/dL; aspartate aminotransferase, 39 U/L; alanine aminotransferase, 21 U/L; lactate dehydrogenase of 190 U/L; amylase, 58 U/L; and lipase, 15 U/L. A plain abdominal film and computerized tomography (CT) scan of the abdomen showed dilated small and large bowel loops. A diagnosis of partial small bowel obstruction was made, and she was treated conservatively.

After 2 days of observation with no clinical improvement, her white blood cell count continued to rise with a left shift, and she also developed rebound tenderness. Repeat CT scan of the abdomen showed multiple fluid and gas filled loops of small bowel without a definite point of obstruction. There was some small bowel wall thickening. There was vascular congestion seen in the mesenteric vessels. Retroperitoneum was unremarkable. There was new development of ascites. There were multiple diverticula in the sigmoid colon as seen previously. She underwent an exploratory laparotomy, and a section of the mid-jejunum was found to be necrotic without perforation. Ninety-nine centimeters of bowel was resected and submitted for pathological examination.

Two days after the surgery, the patient complained of a shade obscuring her vision in her left eye, and on examination, she had decreased visual acuity in that eye. The next day, her vision in that eye deteriorated to only hand movement and then rapidly to no light perception. Funduscopic examination
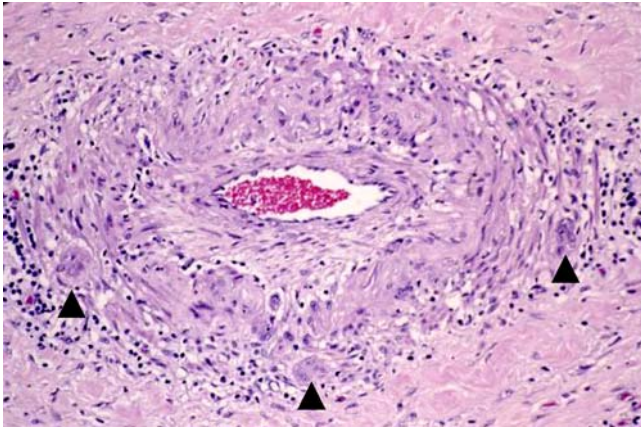
Figure 1. Narrowed lumen of the mesenteric artery with multiple giant cells (arrows) in the tunica media.

by an ophthalmologist showed left ischemic optic neuropathy and a normal right optic disc. On the same day, the pathological examination of her bowel was reported as giant cell arteritis. On repeated questioning at this time, she reported intermittent blurred vision in her left eye about a week before admission. She also recalled having a severe headache 4 days before admission that was different from her usual migraines, but this was on the right side, opposite to the side of her blindness. Examination of her temporal arteries did not show any thickening or tenderness.

Her erythrocyte sedimentation rate and C reactive protein were 99 mm/hour (normal range, 0–33 mm/hour) and 131 mg/L (normal range, 0–3 mg/L), respectively. As we had a definite pathology, a temporal artery biopsy was not done. She was started on intravenous Methylprednisolone, 24 mg twice daily. A few days later, she was discharged home on Prednisone, 60 mg daily. She did not recover from her visual loss in the affected eye. A month later, she was readmitted with steroid-related complications. She had developed impaired glucose tolerance, proximal myopathy, methicillin sensitive staphylococcus bacteremia, and a decubitus ulcer. Over the next 10 days, her steroid dose was tapered rapidly down to 12.5 mg daily. Despite treatment with antibiotics, nutritional support, and other supportive care, she did not improve and died about 20 days after admission. An autopsy concluded that the cause of death was ischemic heart disease with hypertensive cardiomyopathy. The major vessels of the heart, lungs, kidney did not show any evidence of giant cell arteritis. However, the temporal artery was not examined.

Gross pathology of the small bowel showed focal transmural necrosis and extensive mucosal necrosis. Proximal and distal segments of the involved bowel had viable margins. Histology of the mesenteric vessels showed focal giant cell arteritis. The arteritis was seen involving numerous mesenteric vessels of varying sizes. There was infiltration of the artery walls by multinucleated giant cells and inflammatory cells, predominantly lymphocytes. The lumen of these vessels was significantly narrowed with thrombosis.

Ideally, it would have been desirable to correlate these histological findings with a biopsy of the temporal artery. Unfortunately, the patient's clinical status did not allow us to do this when the diagnosis was first made. Even if the temporal artery had been examined at autopsy, the findings of giant cell arteritis

would likely not have been present after 4 weeks of steroid therapy. While this is a limitation of our report, the classical histological findings along with the sudden blindness strongly support a diagnosis of giant cell arteritis. Sections of the mesenteric vessels in our patient are shown in Figures 1 and 2.

DISCUSSION

Small bowel obstruction is a common clinical problem that accounts for 12% to 16% of surgical admissions annually. Mechanical causes are the most common with adhesions causing 60% of cases. Other etiologies are malignant tumors (20%), hernias (10%), inflammatory bowel disease (5%), volvulus (3%), and other miscellaneous causes (2%).[3] Initial management includes fluid resuscitation, nasogastric decompression, and possibly, broad spectrum antibiotics. It is important to differentiate partial from complete obstruction. Continued passage of stool or flatus and persistence of colonic air 6 to 12 hours after onset of symptoms indicate partial obstruction. CT imaging determines the degree of intestinal collapse distal to the obstruction and may also show a transition zone. The majority of cases of partial small bowel obstruction resolve spontaneously, and therefore, a trial of nonoperative management is appropriate in most cases. Management of complete bowel obstruction is more controversial. A delay of surgery for 12 to 24 hours is reasonable, but the incidence of strangulation and ischemia significantly increases after longer periods of nonoperative management. Some signs indicating infarction are continuous abdominal pain, fever, tachycardia, peritoneal signs, leukocytosis, acidosis, absence of bowel sounds, and blood in stools. However, none of these signs are pathognomonic.

Patients with acute mesenteric ischemia may initially present with relatively benign abdominal findings because peritoneal signs do not develop until transmural bowel necrosis occurs. Acute mesenteric ischemia carries a mortality rate exceeding 70% despite advances in patient management.[4] Early recognition and treatment is important for intestinal viability and survival. CT imaging findings, although not specific, may aid in differentiating bowel ischemia from infarction. Some radiographic features that indicate ischemia rather than infarction are bowel wall thinning, dilatation of bowel lumen, and pneumatosis.[5] Older patients with comorbid
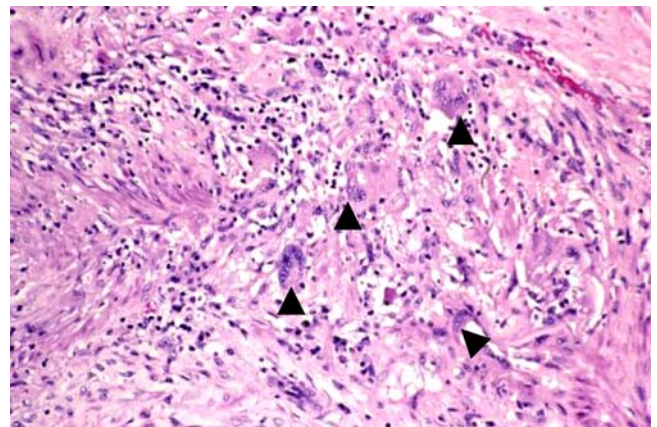

Figure 2. A section of the media of the mesenteric blood vessel showing several multinucleate giant cells (arrows).

illnesses account for a majority of cases of mesenteric ischemia. Acute occlusion may be related to atherosclerosis, low cardiac output states, cardiac arrhythmias, severe valvular heart disease, aortic or mesenteric dissection, and different types of vasculitis.[4,5]

Localized vasculitis of the gastrointestinal tract has been reported.[6] More commonly, however, it occurs as part of a systemic vasculitis. Polyarteritis nodosa is the vasculitis most frequently associated with abdominal manifestations. Other vasculitides associated with bowel infarction are giant cell arteritis, Churg–Strauss syndrome, rheumatoid vasculitis, Kawasaki disease, Takayasu disease, and Buerger disease.[7] Vasculitis should be considered early in the differential diagnosis if the patient is young, has no other predisposing factors for bowel ischemia, and there is evidence for multi-organ involvement. CT imaging findings that suggest vasculitis are skip lesions, involvement of multiple anatomic regions, and evidence of ischemia at various chronological stages. Involvement of duodenum almost always indicates vasculitis. Mesenteric angiography is considered the gold standard for acute mesenteric ischemia and can help differentiate vasculitis from other causes of acute arterial occlusion.

Giant cell arteritis involving the bowel has been reported in a small number of cases. Our PubMed search http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?DB=pubmed *National Library of Medicine and National Institutes of Health* (words used: giant cell arteritis, bowel; extracranial giant cell arteritis) revealed 11 cases of bowel involvement with giant cell arteritis in the English literature, the earliest of which was in 1976. Stenwig [8] reviewed 64 cases of extracranial giant cell arteritis

Table 1. Cases of mesenteric giant cell arteritis

Author	Age	Comorbidities	Gastrointestinal symptoms	Cranial symptoms	Mesenteric biopsy	Temporal biopsy
Current case	78	Migraine headaches, carotid artery disease	Abdominal pain, nausea, obstipation	Sudden blindness	Giant cells and lymphocytes in arterial wall; luminal thrombosis	Not done
8	77	Dyspepsia	Abdominal pain, vomiting; bowel obstruction and perforation	None	Giant cells in mesenteric arterial wall; luminal thrombosis	Giant cell arteritis
9	67	Osteoarthritis	Abdominal pain, vomiting, diarrhea; bowel perforation	None	Chronic inflammatory infiltrate with giant cells in media	Negative
10	87	None mentioned	Abdominal pain; ischemia of sigmoid colon	None	Luminal narrowing; panarteritis; giant cells in media	Not done
11	68	None mentioned	Periumbilical pain, anorexia, nausea, vomiting; ischemia of small bowel	Bitemporal headache	Arteritis in small and medium arteries; giant cells in media	Marked thickening and narrowing of lumen; giant cells in media
12	65	None mentioned	Abdominal pain with fever and arthralgias; bowel necrosis	Headaches	Giant cell angiitis with eosinophilic infiltration	Negative
13	78	None mentioned	Abdominal pain, vomiting; bowel infarction	None	Giant cell arteritis	Giant cell arteritis
14	63	Hepatitis C, aplastic anemia-PNH syndrome	Abdominal pain, melena; bowel perforation	Neck stiffness, jaw claudication, fever	Arteritis of small arteries	Granulomatous arteritis with giant cells
15	73	None mentioned	Abdominal pain, nausea, vomiting; bowel necrosis	Bitemporal headache, blurring of vision, jaw claudication	Active arteritis; giant cells in media	Not done
16	43	None mentioned	Abdominal pain; bowel gangrene	None	Granulomatous inflammation with fibrinoid necrosis; giant cells in vessel wall	Not done
17	82	Congestive heart failure, renal calculi, osteoarthritis	Abdominal pain; bowel infarction	Tongue pain with necrosis; no other cranial symptoms	Not done	Inflammatory infiltrate with giant cells in intima and media
18	69	Depression	Diarrhea, rectal bleeding, abdominal pain; bowel infarction	None	Not done	Arteritis with disruption of elastic lamina and giant cells

before 1976. These were autopsy studies; of the 64 cases with extracranial giant cell arteritis, 13 cases showed mesenteric involvement with 4 of these 13 cases showing intestinal gangrene.

Among the 11 cases since 1976, 9 were biopsy proven in the mesenteric vasculature.[8–16] The 2 other cases reported giant cell arteritis in other sites in patients with intestinal infarction, in which the infarction was therefore presumed to be due to giant cell arteritis [17,18] (See Table 1). There are 4 case reports in other languages of mesenteric giant cell arteritis, 2 biopsy proven [19,20] with 2 possible cases. [21,22] Of the 9 biopsy proven cases, 4 patients presented with abdominal symptoms but no symptoms of temporal arteritis (See Table 1). One patient presented with both abdominal pain and throbbing headache, and both the temporal artery and mesenteric vessel biopsy were positive.[11] In 2 patients, bowel involvement was the initial presentation; interestingly, temporal artery biopsy was done in spite of no cranial symptoms and was positive for giant cells.[8,13] In 2 other patients, however, the mesenteric vessels showed giant cell arteritis, whereas their temporal artery biopsy was negative.[9,12] Thus, a temporal artery biopsy cannot reliably exclude giant cell arteritis of the mesenteric artery.

Histologically, giant cell arteritis has a granulomatous inflammation. The lumen is narrowed because of intimal proliferation. The adventitia is infiltrated by mononuclear and occasionally polymorphonuclear cells. The media is dominated by giant cells, which can vary from cells with 2 nuclei to masses with multiple nuclei. An early feature of the inflammation is fragmentation of the internal elastic lamina. An uncommon pattern of inflammation is the absence of granulomas with a mixed inflammatory infiltrate and no giant cells. The presence of fibrinoid necrosis is rare and is an indication to consider a different vasculitis.[23]

While examination of the affected mesenteric vessels may sometimes not be diagnostic of a specific vasculitic syndrome, our patient showed extensive arteritis with giant cells. Among the vasculitic disorders, giant cells are most commonly seen in giant cell arteritis. Granulomatous inflammation with giant cells can also be seen in other vasculitides, notably Takayasu arteritis and Wegener's granulomatosis. Our patient, however, did not have other clinical and histological findings to suggest these alternate diagnoses. Giant cells can occasionally be seen in Churg–Strauss and Buergers disease (See Table 2). While Polyarteritis nodosa is the vasculitis most likely to cause abdominal ischemia, it is not a granulomatous disease.

In cases where histology is inconclusive for a specific vasculitic disorder, diagnosis is made on demographic characteristics, site of vessel involvement, and clinical manifestations. Our patient had a granulomatous inflammation with giant cells in the mesenteric vessels. Moreover, she developed ischemic optic neuropathy consistent with cranial arteritis. Based on her age, ethnicity, clinical manifestations, and pathology, giant cell arteritis is the most likely diagnosis.

Steroids are effective treatment for the inflammatory lesion in giant cell arteritis. Cyclophosphamide or other immunosuppressive therapy is typically needed for vasculitis because of polyarteritis nodosa but not giant cell arteritis. In all the cases we reviewed of giant cell arteritis involving the mesenteric artery, the diagnosis was only made postoperatively, and the outcomes were poor. In each case, steroids were started after the diagnosis was made.

This case report, along with a few others in the literature, illustrates that intestinal infarction can occur as a rare

Table 2. Differentiating features of common vasculitides

Syndrome	Involved vessels	Symptoms	Histology
Giant cell arteritis	Large and medium sized arteries, commonly carotid artery branches	Systemic symptoms plus temporal headache, jaw claudication, visual loss	Giant cells and granuloma formation
Takayasus arteritis	Aorta and its main branches	Claudication of extremities, coronary or pulmonary involvement	Granulomatous inflammation largely similar to above
Polyarteritis nodosa	Medium and small sized arteries of kidney, heart, liver, GI tract	Systemic symptoms with end organ damage	Acute necrotizing reaction with fibrinoid necrosis
Wegener's granulomatosis	Small to medium sized arteries of respiratory tract	Varying degrees of lung damage and kidney involvement	Necrotizing granulomas with occasional giant cells in intra- and extravascular sites
Churg–Struass angiitis	Medium and small sized arteries and veins especially of lungs	End organ involvement, rarely kidneys	Necrotizing granulomas with more eosinophils

manifestation of giant cell arteritis. Thus, vasculitic disorders should be considered in the differential diagnosis of bowel infarction, especially in the absence of other obvious causes. When a definite diagnosis is made, aggressive treatment for the specific disorder should be started.

Corresponding Author: *Aniyizhai Annamalai, MD, Department of General Internal Medicine, Southern Illinois University School of Medicine, P.O. Box 19636, Springfield, IL 62794-9636, USA (e-mail: aannamalai@siumed.edu).*

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