Case reports

Localized vasculitis of the gastrointestinal tract: a case report and literature review

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ABSTRACT

Localized gastrointestinal vasculitis is a rare condition. It may be observed as an incidental unexpected pathologic finding at the time of biopsy of an abdominal mass or may present as unexplained abdominal pain with or without unexplained lower gastrointestinal bleeding. In this report we describe a new case of localized polyarteritis nodosa with involvement of peripancreatic middle-sized blood vessels. A literature review of cases of localized gastrointestinal vasculitis was also conducted. A major point of concern is whether a single organ vasculitis of the gastrointestinal tract is actually a localized gastrointestinal vasculitis or simply an initial manifestation of a more severe systemic vasculitis. Due to this, in cases of localized gastrointestinal vasculitis a complete evaluation of the patient to exclude the presence of a systemic a potentially threatening systemic vasculitis is required.

Introduction

The vasculitides are a heterogeneous group of conditions characterized by blood vessel inflammation and necrosis (1). By definition, they are systemic diseases with overlapping clinical and pathologic manifestations (1, 2). However, cases of a vasculitis apparently confined to one organ and mimicking other conditions have previously been reported (3). In this regard, a number of cases of localized vasculitis involving the gastrointestinal tract have also been described (4-8).

In this report we describe a new case of localized vasculitis involving the gastrointestinal tract. We have conducted a review of cases of localized gastrointestinal vasculitis.

Case report

A 75-year-old white man with a past history of smoking, chronic bronchopathy and colonic diverticular disease was operated due to epigastric pain and the presence of a pancreatic mass on ultrasound and computed tomography scans. In the operation a microcystic serous cystadenoma of pancreas was found. Because of that a cephalic duodenopancreatectomy was performed. Excision was curative. Incidentally, a polyarteritis nodosa with involvement of peripancreatic middle-sized blood vessels was observed (Figs. 1 and 2). However, no evidence of systemic vasculitis was found over a 3 year-follow-up period after the diagnosis.

Discussion

The present case is a good example of localized abdominal vasculitis involving middle sized blood vessels. In most cases, however, the vasculitic involvement of the gastrointestinal tract is known to occur in the setting of a primary systemic vasculitis (9, 10). In adults vasculitis of the gastrointestinal tract is more commonly observed in patients with polyarteritis nodosa with typical involvement of middle sized and small arteries (10). Intestinal vasculitis may also occur in patients with other primary systemic necrotizing vasculitides affecting small and middle-sized blood vessels such as Churg Strauss syndrome (9, 10) and more rarely in cases of Wegener's granulomatosis (9). In addition, primary systemic vasculitides involving only small blood vessels (capillaries, venules or arterioles) may also cause gastrointestinal manifestations. In this regard, abdominal pain and gastrointestinal bleeding are typical findings observed in children with Henoch-Schönlein

purpura (11, 12). Individuals classified as having hypersensitivity vasculitis may also have gastrointestinal manifestations due to abdominal vasculitis. However, severe gastrointestinal complications due to small-sized blood vessel vasculitis are less common and generally less severe in patients with hypersensitivity vasculitis than in those with Henoch-Schönlein purpura (13). Children with Kawasaki disease may also present gastrointestinal complications (9). Cases of gastrointestinal manifestations due to primary systemic vasculitis affecting predominantly large-sized blood vessels have been reported in individuals with Takayasu arteritis (9) and more rarely in the context of giant cell arteritis (14).

Population-based studies have disclosed that secondary vasculitides are not uncommon in patients with rheumatic diseases (15). With respect to this, vasculitis involving the gastrointestinal tract may be observed in individuals with rheumatoid arthritis, systemic lupus erythematosus or Behçet's disease (9, 16).

As discussed before, although a vasculitis is generally considered to be a systemic disease, it may present as a localized condition with or without inflammatory signs (5). Localized vasculitis involves blood vessels within a confined vascular distribution or single organ without clinical evidence of generalized inflammation (4, 5).

Burke *et al*. reported a series of 63 patients with localized gastrointestinal vasculitis (4). As with our present case, 33 (52%) of them were classified as having polyarteritis nodosa due to involvement of middle-sized muscular arteries. Burke *et al*. also found 8 (13%) cases of localized gastrointestinal necrotizing vasculitis affecting arteries and veins with numerous eosinophils along with extravascular eosinophilic microabscesses typical of Churg Strauss syndrome (4). Besides 12 (19%) cases in whom the only finding was inflammation of muscular veins, 2 cases with pathological findings of thromboangiitis obliterans and 1 with giant cells within the arterial media layer, these authors reported 6 (10%) patients with small-sized blood vessel vasculitis. In keeping with these observations, we recently reported a patient with isolated colonic vasculitis due to involvement of small-sized blood vessels (8). In that middle-age man, a colonic biopsy disclosed the presence of a necrotizing arteritis involving small arteries with fibrinoid necrosis and neutrophilic infiltration accompanied by extravasation and fragmentation of granulocytes (leukocytoclasia). Of note, the intestinal crypts were normal (8). In the present report we show an unpublished figure (Fig. 3) that emphasizes the involvement of small blood vessels within the colonic mucosa in

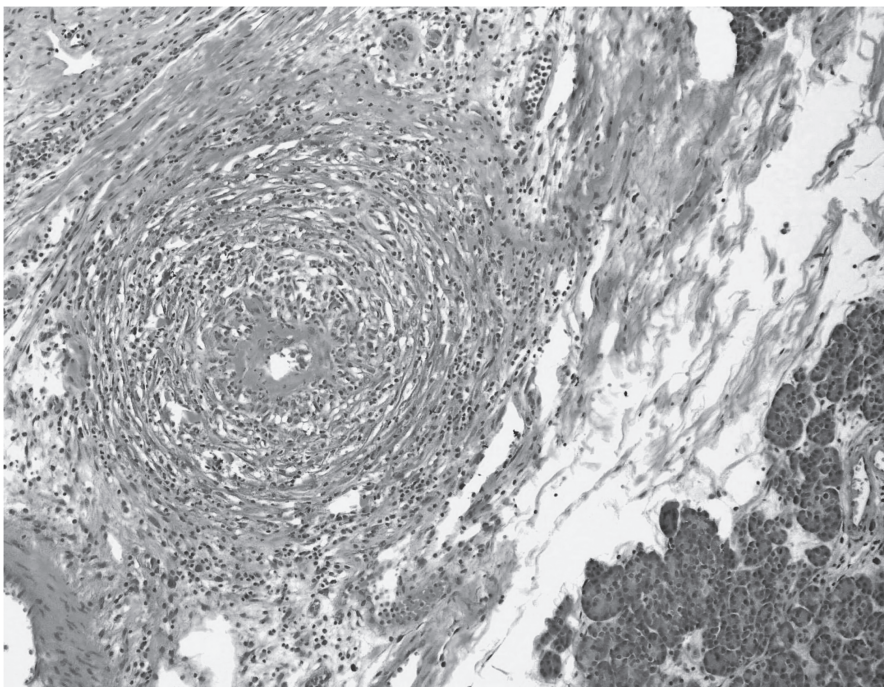

Fig. 1. A pancreatic medium-sized muscular artery showing panmural infiltrate, fibrinoid degeneration and destruction of the wall.

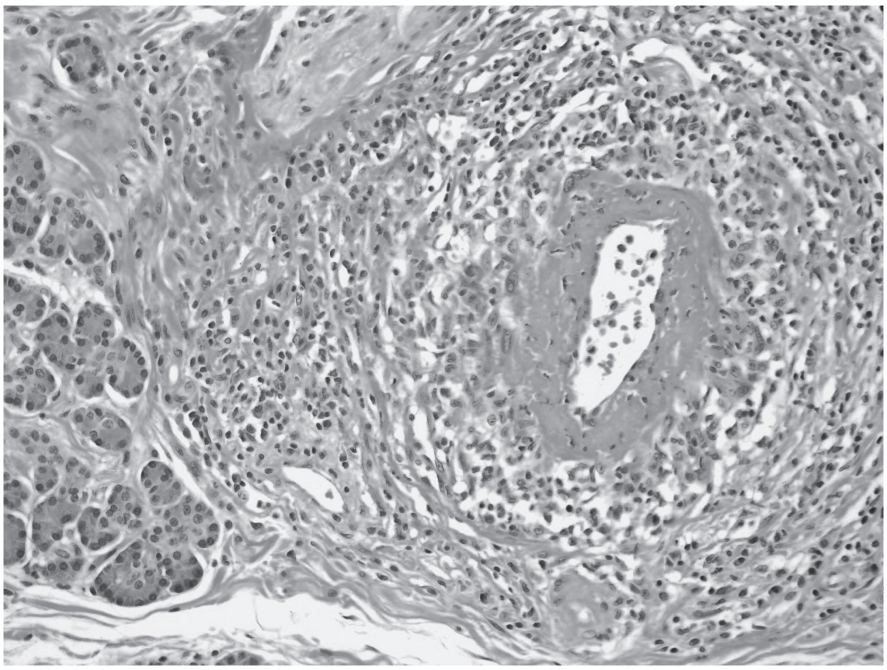

Fig. 2. An artery close to the pancreatic acini with fibrinoide necrosis and a florid inflammatory cell infiltrate consisting mainly of neutrophils around and within the vascular wall.

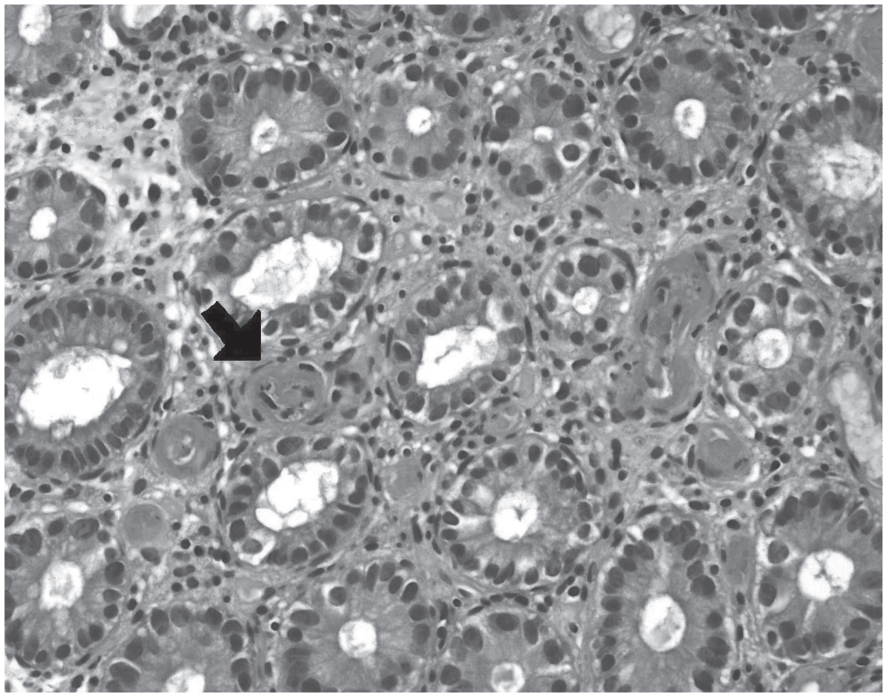

Fig. 3. Among the colonic glands, affected small blood vessels of the mucosa can be seen. Fibrinoid necrosis and nuclear debris within the vascular walls (arrow).

our patient with isolated colonic vasculitis (8).

The most frequently involved organs in patients with localized vasculitis of the gastrointestinal tract are the small and large bowel, gallbladder, appendix, and pancreas (4, 5). As described in our patient with localized polyarteritis nodosa with involvement of peripancreatic middle-sized blood vessels, a localized gastrointestinal vasculitis may be an unexpected pathologic diagnosis at the time of biopsy of a tumor (5). However, a localized vasculitis of the gastrointestinal tract may present as unexplained abdominal pain with or without unexplained gastrointestinal bleeding (8). In these cases anorexia and weight loss are not uncommon.

Patients with polyarteritis nodosa of the gallbladder usually present with acalculous cholecystitis (5). An acute abdomen may be the presenting manifestation of an appendiceal polyarteritis nodosa (5). Small-vessel vasculitis involving the appendix has also been described (4). Moreover, asymptomatic small-vessel vasculitis in the gallbladder and appendix has also been described (4).

The intestine is by far the most common site clinically involved in patients with localized vasculitis (5). The small intestine is more commonly affected than the large intestine (5). In cases of localized bowel vasculitis the most common presenting symptoms are acute abdomen and peritonitis, lower gastrointestinal bleeding or small bowel obstruction (5). Bisceglia *et al*. reported two young patients with acute abdomen caused by necrotizing vasculitis of the cecum and ascending colon with no history of systemic disease or medical treatment (17). A case of toxic megacolon as presenting manifestation of an isolated colonic leukocytoclastic vasculitis that required surgical exploration and subsequent colectomy has also been described (7).

Burke *et al*. reported 5 patients who underwent partial pancreatectomies for biopsy of pancreatic masses (4). Besides areas of chronic pancreatitis, the histologic examination disclosed the presence of a small-vessel vasculitis in one case and a polyarteritis nodosa in the remaining 4 cases. In our case a middle-sized blood vessel vasculitis was also observed following surgery for a pancreatic tumor.

A major point of concern is whether a single organ vasculitis of the gastrointestinal tract is actually a localized gastrointestinal vasculitis or simply an initial manifestation of a more severe systemic vasculitis. Taking into account this consideration, complete evaluation of a patient with localized gastrointestinal vasculitis is mandatory to exclude a systemic vasculitis as the prognosis may differ, and hence, the treatment also. Due to this, in patients presenting with localized gastrointestinal vasculitis laboratory determinations including antineutrophil cytoplasm antibodies, antinuclear antibodies, rheumatoid factor, cryoglobulins, C3 and C4 serum complement levels, HIV, and hepatitis B and C serology should be performed. Moreover, close follow-up, in particular during the first 5 years after the diagnosis of localized gastrointestinal vasculitis, is required to confirm that the vasculitis is strictly restricted to the gastrointestinal tract and no progression to a more threatening systemic vasculitis occur in the following years after the diagnosis of this condition.

In conclusion, localized gastrointestinal vasculitis is a rare condition. It may be observed as an incidental finding in the biopsy or may present as unexplained abdominal pain and bleeding. In cases of localized gastrointestinal vasculitis a complete evaluation of the patient to exclude the presence of a systemic a potentially threatening vasculitis is required.

References

1. GONZALEZ-GAY MA, GARCIA-PORRUA C: Epidemiology of the vasculitides. *Rheum Dis Clin North Am* 2001; 27: 729-49.
2. JENNETTE JC, FALK RJ: The role of pathology in the diagnosis of systemic vasculitis. *Clin Exp Rheumatol* 2007; 25 (Suppl. 44): S52-6.
3. GARCIA-PORRUA C, MATE A, DURAN-MARIÑO JL, FERNANDEZ-MARTINEZ C, GONZALEZ-GAY MA: Localized vasculitis in the calf mimicking deep venous thrombosis. *Rheumatology* (Oxford) 2002; 41:944-5.
4. BURKE AP, SOBIN LH, VIRMANI R: Localized vasculitis of the gastrointestinal tract. *Am J Surg Pathol* 1995; 19: 338-49.
5. QUINET RJ, ZAKEM JM, MCCAIN M: Localized versus systemic vasculitis: diagnosis and management. *Curr Rheumatol Rep* 2003; 5: 93-9.
6. CHARRON P, SMITH J: Mesenteric venulitis in a 71-year-old man after acute appendicitis. *Am Surg* 2005; 71: 574-7.
7. VLAHOS K, THEODOROPOULOS GE, LAZARIS ACH *et al*.: Isolated colonic leukocytoclastic vasculitis causing segmental megacolon: Report of a rare case. *Dis Colon Rectum* 2005; 48: 167-71.
8. GARCIA-PORRUA C, GUTIERREZ-DUQUE

O, SOTO S, GARCIA-RODEJA E, GONZALEZ-GAY MA: Localized vasculitis of the gastrointestinal tract. *Semin Arthritis Rheum* 2006; 35: 403-6.

9. MÜLLER-LADNER U: Vasculitides of the gastrointestinal tract. *Best Pract Res Clin Gastroenterol* 2001; 15:59-82.
10. PAGNOUX C, MAHR A, COHEN P, GUILLEVIN L: Presentation and outcome of gastrointestinal involvement in systemic necrotizing vasculitides: analysis of 62 patients with polyarteritis nodosa, microscopic polyangiitis, Wegener granulomatosis, Churg-Strauss syndrome, or rheumatoid arthritis-associated vasculitis. *Medicine* (Baltimore) 2005; 84: 115-28.
11. DILLON MJ: Henoch-Schönlein purpura: recent advances. *Clin Exp Rheumatol* 2007; 25 (Suppl. 44): S66-68.
12. CALVIÑO MC, LLORCA J, GARCÍA-PORRÚA C, FERNÁNDEZ-IGLESIAS JL, RODRIGUEZ-LEDO P, GONZÁLEZ-GAY MA: Henoch-Schönlein purpura in children from northwestern Spain: a 20-year epidemiologic and clinical study. *Medicine* (Baltimore) 2001; 80: 279-90.
13. GARCÍA-PORRÚA C, GONZÁLEZ-GAY MA: Comparative clinical and epidemiological study of hypersensitivity vasculitis versus Henoch-Schönlein purpura in adults. *Semin Arthritis Rheum* 1999; 28: 404-12.
14. ANNAMALAI A, FRANCIS ML, RANATUNGA SK, RESCH DS: Giant cell arteritis presenting as small bowel infarction. *J Gen Intern Med* 2007; 22: 140-4.
15. GONZÁLEZ-GAY MA, GARCÍA-PORRÚA C: Systemic vasculitis in adults in northwestern Spain, 1988-1997. Clinical and epidemiologic aspects. *Medicine* (Baltimore) 1999; 78: 292-308.
16. GONZALEZ-GAY MA, SANCHEZ-ANDRADE A, PULPEIRO JR, ARMESTO V: Hepatic arteritis in Behçet's disease. *J Rheumatol* 1991; 18: 152-3.
17. BISCEGLIA M, GERMANI G, TARDIO B, DI MATTIA A, LIBERGOLI M: Leukocytoclastic vasculitis of the colon. *Ital J Surg Sci* 1989; 19: 269-72.