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

M.A. Gonzalez-Gay, T.R. Vazquez-Rodriguez, J.A. Miranda-Filloy, A. Pazos-Ferro, E. Garcia-Rodeja*

Divisions of Rheumatology and *Pathology, Hospital Xeral-Calde, Lugo, Spain.
Miguel A. Gonzalez-Gay, MD, PhD; Tomas R. Vazquez-Rodriguez, MD; Jose A. Miranda-Filloy, MD; Ana Pazos-Ferro, MD; Eugenia Garcia-Rodeja, MD, PhD.

Key words: Localized vasculitis, localized middle-sized blood vessel vasculitis, abdominal vasculitis, gastrointestinal tract.

*Correspondence to: Dr. Miguel A. Gonzalez-Gay, Division of Rheumatology, Hospital Xeral-Calde, c/ Dr. Ochoa s/n, 27004 Lugo, Spain. E-mail: miguelaggay@hotmail.com

Received on March 6, 2008; accepted in revised form on March 18, 2008.

CLINICAL AND EXPERIMENTAL RHEUMATOLOGY 2008.

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.

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...
Localized gastrointestinal vasculitis

M.A. Gonzalez-Gay et al.

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 thrombocytopenia 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 pannular infiltrate, fibrinoid degeneration and destruction of the wall.

Fig. 2. An artery close to the pancreatic acini with fibrinoid necrosis and a florid inflammatory cell infiltrate consisting mainly of neutrophils around and within the vascular wall.
Localized gastrointestinal vasculitis / M.A. Gonzalez-Gay et al.

CASE REPORT

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

8. GARCIA-PORRUA C, GUTIERREZ-DUQUE M.A. Gonzalez-Gay et al.

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).
CASE REPORT


