CASE REPORT

Mesenteric inflammatory veno-occlusive disease as a rare cause of acute abdomen

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Abstract
Mesenteric inflammatory veno-occlusive disease is a rare but recognized cause of intestinal ischemia, who can be defined as phlebitis or venulitis affecting mesentery or the bowel, without any evidence of coexisting of an obvious predisposing cause or a coexisting arterial inflammatory involvement. We report the case of a male patient, 63 year old, admitted in the Emergency County Hospital of Craiova, who after presenting with an acute abdomen, underwent exploratory laparotomy and resection of the ischemic sigmoid, temporary colostomy and after four months we reintroduced descendent colon in the digestive transit. The resected specimen of the patient was examined histopathologically, and distinctive histopathological characteristics of the mesenteric inflammatory veno-occlusive disease were identified.

Keywords: mesenteric inflammatory veno-occlusive, intestinal ischemia.

Introduction
In up to 5–15% of patients, mesenteric venous thrombosis is identified as the underlying cause of acute mesenteric ischemia. Hypercoagulopathy disorders, such as deficiencies of antithrombin III, protein S or C, and Leiden’s mutation are the risk factors for mesenteric venous thrombosis [1].

The mesenteric veins and their tributaries can become occluded from causes other than venous thrombosis, such as Behçet’s syndrome [2], systemic lupus erytematosus [3] and with drugs [4].

To describe ischemic injury resulting from phlebitis or venulitis affecting either the bowel or mesentery, without any evidence of coexisting arterial inflammatory involvement or an obvious predisposing cause, recently has been used the term of “mesenteric inflammatory veno-occlusive disease” [5].

The etiology, incidence and clinical manifestations remain unknown because the disease is rarely suspected.

Material and methods
A 63 year man was admitted in Emergency County Hospital of Craiova with acute abdomen. Physical examination revealed a distended abdomen with diminished sound and rebound tenderness. Laboratory analysis detected an elevated WBC count (11 800/mm³), urea 43 mg%, creatinine 0.98 mg%, Hb 11.7 g/dl, glycemia 89 mg%, Quick t. 100%, Howell t. 134 sec., Na⁺ 132 mEq/l, K⁺ 3.7 mEq/l, TGO 17 iu, TGP 19 iu, normal international normalized ratio (INR), normal protein C activity and protein S activity. In emergency was made abdominal radiography (Figure 1) that showed dilated thickened large bowel loop.

The patient underwent exploratory laparotomy. Ischemic nonviable sigmoid was easily identified, and we performed sigmoidectomy, temporary colostomy. He was discharged from hospital after 12 days.

The sigmoid and mesosigmoid obtained from sigmoidectomy was fixed in a 10% formalin solution for three weeks. After fixation, sagittal sections of 1 cm thickness were made through the sigmoid and mesosigmoid and 12 specimens from different parts of the lesion were processed into paraffin blocks using standard histological procedures.

Sections of 5 µm were stained with Hematoxylin–Eosin, Gömori’s method for reticulin and Orcein for elastin. The examination was made at a Nikon microscope.

Results
A plain film of the abdomen shows dilatation of large bowel and, in particular, of the ascendant and transverse colon with widening of the haustral folds and irregularity of the bowel wall and the absence of air distally to the rectum (Figure 1).

The ileo-cecal valve was competent, and thus, there was no small bowel air. The uniform wall thickening who reflects submucosal edema and hemorrhage was present on plain films of the abdomen, but was not emphasized the presence of gas in the bowel wall (“pneumatosis intestinalis”).
After exploratory laparotomy the characteristic pattern of injury consists of superficial epithelial necrosis of the sigmoid, associated with an inflammatory pseudomembrane. We performed sigmoidectomy, temporary colostomy.

Operative pieces was 30 cm long section of sigmoid, dark, wine-red in color, swollen and edematous wall. At both extremities, the sigmoidian wall had a normal color. Macroscopically there was mesosigmoidal congestion and the sigmoid wall appeared edematous, hemorrhagic and thickened with evidence of infarction (Figure 2). Upon the sectioned pieces there was also transmural hemorrhagic infarction (Figure 3).

We found on opening, hemorrhagic content with blood clots inside the sigmoidian lumen. The thromboses were located above all in the smallest and proximal mesosigmoidal venous ramifications, but not in the collector branches and major mesosigmoidian venous vessels.

A careful histological examination of the resected specimens reveled transmural sigmoid and mesosigmoid edema, hemorrhage and abundant inflammatory infiltrate composed of polymorphs, eosinophils, lymphocytes and bizarre fibroblasts. The pathognomonic lesion was a necrotizing venulitis in the submucosa and mesosigmoid (Figure 4). A histologic investigation shows a variable inflammatory infiltrate of multiple veins of colonic wall and the mesosigmoid as well as thrombotic vein occlusion in different stages without involvement of the arteries. The necrotizing venulitis was characterized by fibrin deposition and polymorph infiltration in the mesosigmoid veins. Mesosigmoid veins showed focal thrombotic occlusions of recent onset (Figure 5).

The arterial circulation was unaffected. The concentric or eccentric myointimal hyperplasia of the veins and occlusive phlebosclerosis were absent.

The intima of the mesosigmoid and sigmoidal mural veins was focally thickened by a marked increase in cells and matrix between the endothelium and internal elastic lamina.

Microscopically, sigmoidal mural vein consist of dilated, distorted, thin-walled vessel, mostly lined only by endothelium and, by a small amount of smooth muscle. These findings in this case were consistent with mesenteric inflammatory veno-occlusive disease.

Within the submucosa was a heterogeneous population of cells including lymphocytes, plasma cells, macrophages, eosinophiles, fibroblasts and mast cells embedded within a dense connective tissue. The mucosa was separated into three layers: the muscularis mucosa, the lamina propria and intestinal epithelium. Colonic mucosa consisted of tightly packed crypts lined primarily by goblet cells with intervening flat epithelial surfaces covered by absorptive cells. Cells were generated in the crypts and extruded from the flat intercrypt region on the surface.

A thin layer of smooth muscle makes up the muscularis mucosa. Below this is the connective tissue of the submucosa. There were no crypt abscesses, granulomas alteration of the crypt architecture, or immune complex deposits.

**Discussions**

In 1976 was described the first case of isolated mesenteric venous inflammation and associated thrombosis leading to mesenteric ischemia [3]. Various terminologies have been used to describe these pathological findings, including enterocolic lymphocytic phlebitis [4], intramural mesenteric venulitis [6], necrotizing and giant cell granulomatous phlebitis [7], idiopathic myointimal hyperplasia [8], and most recently mesenteric inflammatory veno-occlusive disease [5].

Although few cases have been reported, their histological findings were similar to those of our patient. Myointimal hyperplasia of the mesenteric veins, which is usually associated with chronic mesenteric inflammatory veno-occlusive disease and represent organized thrombus [5], was absent and there was no evidence of either arterial inflammation or occlusion.

Because most patients diagnosed with venous thrombosis are subsequently started on anticoagulant therapy, it is important to distinguish patients with mesenteric inflammatory veno-occlusive disease from mesenteric venous thrombosis. Acute mesenteric venous thrombosis may be idiopathic, but is usually is associated with other disease entities. The underlying conditions most frequently associated with mesenteric venous thrombosis include coagulopathies such as polycytemia vera, sickle cell disease, antithrombin III, protein S and protein C deficiency. Other predisposing include portal hypertension, infra-abdominal sepsis, blunt and operative trauma, and renal transplantation. Use of oral contraceptives associated with mesenteric venous thrombosis is recognized with increasing frequency. In our patient, the blood test for hypercoagulability was negative and the histologic features of isolated focal thrombotic occlusion and distension of inflamed sigmoidian vein was more consistent with a diagnosis of mesenteric inflammatory veno-occlusive disease and not found in mesenteric venous thrombosis. However, thrombosis is commonly seen as a part of the inflammatory phenomenon in mesenteric inflammatory veno-occlusive disease, but it represents a secondary change. It is possible for thrombosis to eclipse the underlying inflammatory component, leading to an incorrect diagnosis [5].

The terms “ischemic colitis” and “colon infarction” are suitable for only parts of the spectrum of colonic ischemia, for the milder reversible ischemic episodes are not inflammatory but hemorrhagic and coagulation necrosis, true infarction is often absent. There are two major forms of colonic ischemia: gangrenous (transmural) and non-gangrenous colitis (disease contained within the colonic wall). Gangrenous ischemic colitis is caused by obstruction of the major mesenteric vessels. In non-gangrenous ischemic bowel disease the hypo-oxygenation is caused by hypoperfusion of the gut wall microcirculation. Hypoperfusion is most commonly caused by vascular disease (collagen disease, vasculitis, diabetes, atherosclerosis) or by increased viscosity of the blood in sickle cell disease or polycythemia vera.
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Figure 1 – Abdominal radiography showed dilated thickened large bowel loop

Figure 2 – Operative piece: macroscopic aspects with hemorrhagic necrosis of the mucosa, submucosa, muscular layer, under serosa, retraction, infiltration of the mesosigmoid

Figure 3 – Sectioned operative piece emphasize the hemorrhagic necrosis of the mucosa, submucosa and muscular layer of the sigmoid

Figure 4 – Hemorrhagic necroses of the mucosa, submucosa with the presence of the polymorph infiltrate (HE stain, ob ×4)

Figure 5 – In center of the image focal thrombosis occlusion of the mesenteric vein. Necrotising venulitis characterized by fibrin deposition and polymorph infiltration. The artery was unaffected by the veno-occlusive process (HE stain, ob ×20)
Our patient does not have a history of collagen disease, sepsis, myocardial infarct, congestive heart failure, or other inflammatory bowel disease. Mesenteric inflammatory veno-occlusive disease must be distinguished from other types of vasculitis affecting the gastrointestinal tract (linked by intestinal phlebitis or venulitis to drugs, systemic lupus erythematosus, Behçet’s syndrome), but predominantly affect the arterial circulation [5].

The etiology of mesenteric inflammatory veno-occlusive disease remains unclear. Veno-occlusive disease can also affect the liver and lungs with similar histological features [9, 10], and a relationship with food contamination, toxins and drugs has been implicated. Have also been reported, three case of mesenteric inflammatory veno-occlusive disease associated with the rutoside [11], a drug used to treat varicose veins, as have cases of mesenteric inflammatory veno-occlusive disease associated with anti-phospholipid syndrome [12] and with primary cytomegalovirus infection [13].

Based on our review of the literature, mesenteric inflammatory veno-occlusive disease can occur at any age and there is a slight male preponderance. It appears to affect a colon in more than 50% of patients, although it has also been reported to affect small bowel, gallbladder and omentum [5].

In the series reported by Hu JC et al., the small bowel was commonly affected, which also included the first reported case of mesenteric inflammatory veno-occlusive disease affecting the appendix [14].

In concordance with our patient, many patients present with a history of prodromal symptoms of a last few days. No characteristic endoscopic or imaging findings have been noted in this prodomal period and majority of patients suffer acute abdominal symptoms necessitating exploratory laparotomy. Ultrasound and colonoscopy and neither of these investigations proved helpful in making of diagnosis. Abdominal radiography in our case showed dilated thickened large bowel loop. Indications for immediate surgery are a necrotizing venulitis leading to peritoneal signs: segmental resection. In our case we practiced sigmoidectomy, and the colitis of Behçet’s syndrome

The indication of prophylactic anticoagulant therapy is not necessary as in case of mesenteric venous thrombosis, but the patients should receive prophylactic anticoagulant therapy in the perioperative period.

Conclusions

The incidence and etiology of mesenteric inflammatory veno-occlusive disease remain unknown, because it is rarely suspected and often under-reported. Mesenteric inflammatory veno-occlusive disease is still often misdiagnosed as mesenteric venous thrombosis, probably because of its low incidence, with possible dangerous complications resulting from subsequent anticoagulant therapy.

The anticoagulants are not necessary, but in the perioperative stage the patient should receive these as a prophylactic therapy.

References


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