Overview of gastrointestinal manifestations of vasculitis

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INTRODUCTION

The vasculitides are defined by the presence of leukocytes in vessel walls with reactive damage to mural structures. Symptomatic involvement of the gastrointestinal tract may occur either in isolation or in combination with multiple organs. Vasculitis should be considered in patients when the history, physical exam, laboratory, and/or radiologic data indicate that multiple organ systems are involved. However, the diagnosis may be confirmed only after pathologic examination of excised tissue due to ischemia, infarction, or perforation of the involved portion of the gastrointestinal tract. It is rare to make a diagnosis based on endoscopic biopsy alone because of the superficial nature of the mucosal biopsies obtained at endoscopy. A full-thickness biopsy is necessary to evaluate arterioles.

This topic will review the clinical features and evaluation of patients with gastrointestinal vasculitis. The major vasculitides that may present with gastrointestinal involvement are also described. The diagnosis and treatment of major vasculitides are discussed in detail separately. (See "Overview of and approach to the vasculitides in adults" and "Clinical manifestations and diagnosis of polyarteritis nodosa in adults" and "Gastrointestinal manifestations of systemic lupus erythematosus" and "Granulomatosis with polyangiitis and microscopic polyangiitis: Clinical manifestations and diagnosis" and "Mixed cryoglobulinemia syndrome: Clinical manifestations and diagnosis".)

EPIDEMIOLOGY

Gastrointestinal involvement with vasculitis is most prevalent in patients with immunoglobulin A vasculitis (Henoch-Schönlein purpura), antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis, polyarteritis nodosa (PAN), and Behçet syndrome, although it can be seen with any of the vasculitides [1]. In three series with a total of 351 patients with systemic vasculitis, approximately one-third had gastrointestinal manifestations [2-4].

MAJOR VASCULITIDES WITH GASTROINTESTINAL INVOLVEMENT

The frequency and type of gastrointestinal manifestations vary depending on the specific vasculitis. Symptoms of vasculitis involving the gastrointestinal tract result from ischemia to the affected organ. Although vasculitis is frequently classified by the size of the vessels involved, the symptoms from either large-, medium-, or small-vessel vasculitis can overlap. As such, abdominal pain, nausea, vomiting, diarrhea, and/or gross or occult blood in the stool is common. Gross bleeding is more commonly seen in vasculitis involving the colon compared with that involving the small bowel, but there is considerable overlap of symptoms so it is difficult to determine clinically which portion of the gastrointestinal tract is involved.

The major vasculitides with gastrointestinal involvement are discussed below, using the international Chapel Hill Consensus Conference (CHCC) classification categories. (See "Overview of and approach to the vasculitides in adults".)

Large-vessel vasculitis

Takayasu arteritis — Takayasu arteritis (TAK) is a chronic vasculitis of unknown etiology that predominantly affects the aorta and its primary branches. Women are affected in 80 to 90 percent of cases, with an age of onset that is usually between 10 and 40 years; the prevalence is greatest in Asia. Genetic factors, such as the human leukocyte antigen (HLA)-B*52 allele, and alteration in genes encoding proinflammatory cytokines and mediators of humoral immunity may be involved [5].

Gastrointestinal involvement is rare in TAK, but stenoses or occlusion of the large or medium gastrointestinal arteries can occur, resulting in ischemia of the small or large intestine and, rarely, the spleen or liver [6]. (See "Clinical features and diagnosis of Takayasu arteritis", section on 'Symptoms and signs'.)

Giant cell (temporal) arteritis — Giant cell (temporal) arteritis (GCA) also affects the aorta, with a predilection for the cranial branches of the aortic arch. The incidence of GCA increases with age and peaks between 70 and 80 years. (See "Clinical manifestations of giant cell arteritis", section on 'Epidemiology'.)

Mesenteric vessels are rarely affected in GCA, and differentiation of vasculitis from atherosclerosis is difficult among the age groups who have GCA. Abdominal symptoms can also result from the presence of an abdominal aortic aneurysm or dissection. (See "Clinical manifestations of giant cell arteritis", section on 'Atypical features' and "Clinical manifestations of giant cell arteritis", section on 'Large vessel involvement'.)

Medium-vessel vasculitis

Polyarteritis nodosa — Polyarteritis nodosa (PAN) typically affects the medium-sized muscular arteries, with occasional involvement of small muscular arteries. The main risk factor for PAN is hepatitis B virus (HBV) infection, with higher rates observed in areas endemic for HBV infection or without vaccination programs. (See "Clinical manifestations and diagnosis of polyarteritis nodosa in adults", section on 'Epidemiology'.)

Abdominal pain is present in the majority of patients with PAN, is usually nonspecific, and varies in intensity. The pain is likely caused by transmural necrotizing inflammation of the mesenteric vessels leading to bowel ischemia, which is usually the small bowel [7]. Other gastrointestinal symptoms seen with PAN include nausea, vomiting, diarrhea, hematochezia, and melena. Liver or splenic infarcts as well as occlusion of the hepatic veins (Budd-Chiari syndrome) have also been described due to hepatosplenic vessel involvement [8]. (See "Clinical manifestations and diagnosis of polyarteritis nodosa in adults", section on 'Gastrointestinal disease'.)

Small-vessel vasculitis

ANCA-associated vasculitis — Some of the specific clinical features associated with gastrointestinal manifestations for each of the major clinicopathologic variants of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis are described below.

- **Microscopic polyangiitis** – Gastrointestinal manifestations have been reported in 5 to 30 percent of patients [9,10]. Findings consistent with vasculitis may be observed on small or large bowel biopsy in patients with gastrointestinal involvement, but there are no other unique histopathologic findings. (See "Granulomatosis with polyangiitis and microscopic polyangiitis: Clinical manifestations and diagnosis", section on 'Other manifestations'.)

- **Granulomatosis with polyangiitis** – Gastrointestinal involvement has been observed in 5 to 10 percent of patients with granulomatosis with polyangiitis (GPA) [9,11]. Involvement of the small and large bowel are most common, but any organ in the gastrointestinal tract can be involved. Histologic examination may show granulomatous lesions [12]. In some cases, it can be difficult to distinguish the colitis associated with GPA from that of

inflammatory bowel disease. (See "Granulomatosis with polyangiitis and microscopic polyangiitis: Clinical manifestations and diagnosis", section on 'Other manifestations'.)

- **Eosinophilic granulomatosis with polyangiitis** – In a large cohort study of 383 patients with eosinophilic granulomatosis with polyangiitis (EGPA) followed for over five years, 23 percent developed gastrointestinal tract involvement [13]. Biopsy specimens from erosions in the gastrointestinal tract may show eosinophilic infiltration in the mucosa [14]. Infiltration of eosinophils in the mucosa of the gastrointestinal tract can also result in pain, motility disorders, obstructive symptoms, and diarrhea as seen in eosinophilic gastroenteritis. (See "Clinical features and diagnosis of eosinophilic granulomatosis with polyangiitis (Churg-Strauss)", section on 'Gastrointestinal tract'.)

Immunoglobulin A vasculitis (Henoch-Schönlein purpura) — Immunoglobulin A vasculitis (Henoch-Schönlein purpura) is a small-vessel vasculitis that typically occurs in children, although all ages can be affected. Patients classically exhibit lower-extremity purpura, arthritis, and hematuria. (See "IgA vasculitis (Henoch-Schönlein purpura): Clinical manifestations and diagnosis".)

The gastrointestinal tract is affected in up to one-half of patients with immunoglobulin A vasculitis. Gastrointestinal symptoms include colicky abdominal pain, nausea, vomiting, diarrhea, constipation, and occult or overt intestinal bleeding. The course can wax and wane over several weeks and is often self-limited. (See "IgA vasculitis (Henoch-Schönlein purpura): Clinical manifestations and diagnosis", section on 'Gastrointestinal symptoms'.)

Cryoglobulinemic vasculitis — Cryoglobulinemic vasculitis is most commonly associated with hepatitis C virus (HCV) infection but can also be associated with HBV, human immunodeficiency virus (HIV), autoimmune diseases, and lymphoma, or it can be idiopathic ("essential cryoglobulinemia"). Although gastrointestinal involvement is rare in cryoglobulinemic vasculitis, when it occurs, symptoms can be severe and include abdominal pain, bloody stool, intestinal ischemia, intestinal perforation, acute cholecystitis, and pancreatitis [7,15]. HCV-infected patients may progress to cirrhosis as well. (See "Mixed cryoglobulinemia syndrome: Clinical manifestations and diagnosis", section on 'Clinical manifestations'.)

Variable-vessel vasculitis

Behçet syndrome — Behçet syndrome is a vasculitis of unknown etiology and can affect vessels of any size in the venous and arterial circulation. It is characterized by recurrent oral and genital ulcerations and can be complicated by articular, ocular, cutaneous, gastrointestinal, neurologic, or vascular disease. The frequency of gastrointestinal involvement varies geographically, ranging from 3 percent of patients in a Turkish cohort to approximately 50

percent in a Japanese cohort [16,17]. Gastrointestinal ulcerations can occur in any part of the gastrointestinal tract but are most often seen in the terminal ileum, cecum, and ascending colon. Mucosal ulcerations can be attributed to either neutrophilic infiltrates that can mimic inflammatory bowel disease or intestinal ischemia and infarction due to large-vessel vasculitis (especially the mesenteric vessels). (See "Clinical manifestations and diagnosis of Behçet syndrome", section on 'Gastrointestinal involvement'.)

Single-organ vasculitis — Isolated gastrointestinal single-organ vasculitis has been reported in almost any organ associated with the gastrointestinal tract, including the esophagus, stomach, small and large intestines, gallbladder, and pancreas [7,18,19]. Most patients present with abdominal pain, and in a small case series of 18 patients, 12 (67 percent) presented with an acute abdomen requiring surgical intervention [19]. (See "Overview of and approach to the vasculitides in adults", section on 'Single-organ vasculitis'.)

Vasculitis associated with systemic disease

Rheumatoid vasculitis — Rheumatoid vasculitis can develop in patients with longstanding (usually 10 to 15 years) erosive rheumatoid arthritis (RA). Although autopsy studies suggest it is common, occurring in 25 percent of patients, less than 1 percent of patients with RA develop clinical signs of vasculitis, and this is becoming an increasingly extremely rare condition. Ten to 38 percent of these patients will have gastrointestinal manifestations of the vasculitis, usually mesenteric ischemia [20]. (See "Clinical manifestations and diagnosis of rheumatoid vasculitis".)

Systemic lupus erythematosus — The vasculitis associated with systemic lupus erythematosus (SLE) involves small- and medium-sized vessels and can affect the gastrointestinal tract [21]. Lower abdominal pain secondary to mesenteric vasculitis is generally an insidious symptom that may be intermittent for months prior to the development of an acute abdomen with nausea, vomiting, diarrhea, gastrointestinal bleeding, and fever [22]. Mesenteric vasculitis is uncommon in SLE but typically occurs in patients with multiorgan involvement and active disease. (See "Gastrointestinal manifestations of systemic lupus erythematosus", section on 'Mesenteric vasculitis/ischemia'.)

HIV infection — Vasculitis is rare in the post-highly active antiretroviral therapy (HAART) era of HIV infection, but it can occur and ranges from mild skin manifestations to life-threatening illness that can involve the vasculature of the gastrointestinal tract [23]. Treatment is difficult because the pathophysiology is not understood [23].

CLINICAL PRESENTATION

The predominant clinical presentation of systemic vasculitis involving the gastrointestinal tract results from mesenteric ischemia and/or ischemic colitis, which can lead to infarction. Distinguishing between ischemia and infarction caused by vasculitis can be difficult, just as it is with atherosclerotic or embolic vascular disease (see "Overview of intestinal ischemia in adults" and "Acute mesenteric arterial occlusion" and "Nonocclusive mesenteric ischemia" and "Mesenteric venous thrombosis in adults"). Most patients experience abdominal pain, which may be acute or chronic; the latter presentation is also characteristic of chronic mesenteric ischemia due to a low-flow state and may include abdominal pain after eating and weight loss, in addition to nausea, vomiting, and diarrhea. These patients can also present with small bowel obstruction secondary to strictures, intussusception due to submucosal edema and hemorrhage, or massive gastrointestinal bleeding secondary to, usually, large bowel ischemia and necrosis [24]. (See "Chronic mesenteric ischemia".)

The presentation of patients with ischemic colitis secondary to vasculitis is similar to that of patients with ischemic colitis from other etiologies in that both present with acute onset of mild abdominal pain and tenderness over the affected bowel, commonly on the left side of the abdomen. Mild to moderate amounts of rectal bleeding or bloody diarrhea typically develop within 24 hours of the onset of abdominal pain. (See "Colonic ischemia".)

Patients with vasculitis involving the gastrointestinal tract may present with acute abdominal pain due to acute mesenteric ischemia (acute-on-chronic mesenteric ischemia) from thrombus formation. These patients frequently have pain that is out of proportion with findings on abdominal exam before intestinal infarction occurs. Bowel infarction resulting in perforation and peritonitis are rare complications of chronic intestinal ischemia due to vasculitis, but they can occur during acute ischemic episodes. (See "Overview of intestinal ischemia in adults" and "Acute mesenteric arterial occlusion" and "Nonocclusive mesenteric ischemia" and "Mesenteric venous thrombosis in adults".)

Systemic vasculitis can cause ischemic hepatitis [2-4,9]. Liver test abnormalities are relatively common, but clinically significant liver disease is rare. (See "Ischemic hepatitis, hepatic infarction, and ischemic cholangiopathy" and "Approach to the patient with abnormal liver biochemical and function tests" and "Congestive hepatopathy".)

Other, rarer gastrointestinal manifestations of vasculitis include acute pancreatitis, acute cholecystitis, gastritis, and esophagitis [2-4,9]. (See "Chronic mesenteric ischemia" and "Protein-losing gastroenteropathy" and "Clinical manifestations and diagnosis of acute pancreatitis".)

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of gastrointestinal vasculitis varies based on the clinical presentation. Patients with nonvasculitic disease processes may present with symptoms and findings that closely mimic various vasculitides. The differential diagnosis of colonic ischemia is broad and includes small bowel ischemia, infectious colitis, inflammatory bowel disease, and a myriad of other causes for abdominal pain and lower gastrointestinal bleeding. These are discussed in detail separately. (See "Colonic ischemia", section on 'Differential diagnosis'.)

Some of the major diseases to consider as part of the differential diagnosis of gastrointestinal vasculitis include other systemic rheumatic diseases such as systemic lupus erythematosus (SLE), atherosclerotic disease, drug reactions, and vaso-occlusive processes. Among the most important diseases to exclude are infections and malignancies since the immunosuppressive therapy could worsen these conditions and a delay in diagnosis can be extremely dangerous. While it is beyond the scope of this review to provide a comprehensive list of all possible alternative diagnoses, we present several categories of mimics of vasculitis in a table (table 1).

DIAGNOSTIC EVALUATION

Our approach — A gastrointestinal vasculitis should be suspected in patients with clinical features of gastrointestinal ischemia in the absence of risk factors for atherosclerotic vascular disease. In patients without an established diagnosis of vasculitis, the presence of associated systemic or constitutional symptoms in combination with single and/or multiorgan dysfunction is suggestive of a vasculitis. However, the absence of these features does not exclude gastrointestinal vasculitis. (See "Colonic ischemia".)

Our approach to evaluating the patient with possible gastrointestinal involvement of vasculitis depends on whether the patient has an established diagnosis of vasculitis or whether they are presenting for the first time with gastrointestinal symptoms. For patients who have an established diagnosis, the evaluation is focused on ruling out other causes of gastrointestinal symptoms followed by treatment of the underlying vasculitis. (See "Overview of and approach to the vasculitides in adults", section on 'Diagnostic approach'.)

In patients who do not have a known vasculitis but present with predominantly gastrointestinal symptoms and other findings suspicious for a systemic vasculitis, additional testing will vary depending on the constellation of symptoms. The diagnosis of individual vasculitides is generally guided by the pattern of organ involvement, histopathologic features, and characteristic findings on abdominal imaging studies. The approach to diagnosing different

forms of vasculitis are discussed separately in the appropriate topic reviews. (See "Overview of and approach to the vasculitides in adults", section on 'Diagnostic approach'.)

Laboratory testing — There is no single diagnostic test for any of the vasculitides. Basic laboratory tests are mostly used to help ascertain the extent of organs affected and their degree of involvement. Initial laboratory testing should include complete blood count (CBC), serum creatinine, liver chemistries, serologies for viral hepatitis, serum cryoglobulins, and a urinalysis with urinary sediment. An erythrocyte sedimentation rate (ESR) and/or C-reactive protein (CRP) may also be included in the workup, but they are neither sensitive nor specific for vasculitis and must be interpreted in the clinical context. Blood cultures should be drawn to help exclude infection (eg, infective endocarditis). (See "Overview of and approach to the vasculitides in adults".)

An elevated serum lactate level is helpful in evaluating patients with suspected mesenteric ischemia, although it is not specific and a normal value does not exclude this condition.

Additional, more specific serologic testing such as antineutrophil cytoplasmic antibodies (ANCAs) that may further aid in the diagnosis when the presence of an underlying vasculitis is unknown are discussed elsewhere. (See "Overview of and approach to the vasculitides in adults", section on 'Laboratory tests'.)

Vascular imaging — Computed tomography (CT) angiography of the abdomen with intravenous contrast is the preferred imaging technique for patients with suspected vasculitis-induced mesenteric ischemia. Catheter-based angiography is now primarily used for treatment (angioplasty or stent placement), though some experts believe it may be useful in evaluation of patients in whom small-vessel vasculitis is still suspected after all other noninvasive options have been exhausted. (See "Overview of intestinal ischemia in adults", section on 'Advanced abdominal imaging'.)

Endoscopy — Endoscopy is generally performed to exclude alternative diagnoses. Endoscopic findings in patients with vasculitis of the gastrointestinal tract are usually nonspecific. Biopsies obtained rarely provide histologic confirmation of vasculitis. In one study of 148 patients with a primary vasculitis of the upper gastrointestinal tract, histopathologic findings of vasculitis were detected in only 8 of 124 (5 percent) patients from whom biopsy samples were obtained. Nonetheless, endoscopy is useful in excluding other diseases, and sometimes the gross appearance of the mucosa, especially in the colon, is highly suggestive of ischemia.

Endoscopy should be performed with caution in the evaluation of a patient with suspected gastrointestinal involvement from an acute flare of vasculitis because of the elevated risk of perforation in the setting of ischemia. When indicated, it should be performed with minimal air

insufflation to avoid excessive distention that could lead to colon perforation. Colonoscopic findings in the acute setting frequently include edematous, friable mucosa; erythema; and interspersed pale areas. More severe disease is marked by cyanotic mucosa and scattered hemorrhagic erosions or linear ulcerations. In the chronic phase of ischemic colitis, mucosal atrophy and areas of granulation tissue may be found. Biopsies taken from affected areas may show nonspecific changes such as hemorrhage, crypt destruction, capillary thrombosis, granulation tissue with crypt abscesses, and pseudopolyps. Biopsy of a postischemic stricture is marked by extensive transmural fibrosis and mucosal atrophy. Importantly, endoscopic biopsies are not deep enough to sample involved blood vessels. We do not routinely perform video capsule endoscopy as it can only visualize the small bowel and does not permit tissue sampling or therapeutic intervention [25]. (See "Colonic ischemia".)

Other nonspecific endoscopic findings that may be found in the affected portion of the gastrointestinal tract in patients with vasculitis include erosions, ulcers, petechiae, submucosal hemorrhage, edema, and nodularity. The affected area of the gastrointestinal tract varies based on the type of vasculitis. For example, in patients with Behçet syndrome, discrete ulcerations can be found throughout the gastrointestinal tract but are most often seen in the terminal ileum, cecum, and ascending colon [26]. (See "Clinical manifestations and diagnosis of Behçet syndrome", section on 'Gastrointestinal involvement'.)

MANAGEMENT

There are two major components to the treatment of gastrointestinal vasculitis. The first is treatment of the underlying disease, usually with immunosuppressive agents. The second component of therapy is aimed at the gastrointestinal symptoms. For patients with mesenteric ischemia, control of the vasculitis should lead to symptom resolution. An exception is that ischemic symptoms can occur late and in the absence of active disease in patients with healed arteritis in whom scarring has led to progressive attenuation of the vascular lumen. Surgical intervention is necessary in patients with mesenteric infarction or with intestinal perforation.

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Vasculitis".)

SUMMARY AND RECOMMENDATIONS

- While several vasculitides may be associated with gastrointestinal involvement, it is most prevalent in patients with immunoglobulin A vasculitis (Henoch-Schönlein purpura), antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis, polyarteritis nodosa (PAN), and Behçet syndrome. The frequency and type of gastrointestinal manifestations vary among the vasculitides. (See 'Epidemiology' above and 'Major vasculitides with gastrointestinal involvement' above.)

- The predominant clinical presentation of systemic vasculitis involving the gastrointestinal tract results from mesenteric ischemia and/or ischemic colitis. Most patients experience abdominal pain, which may be acute or chronic; the latter presentation is also characteristic of chronic mesenteric ischemia due to a low-flow state and may include abdominal pain after eating and weight loss, in addition to nausea, vomiting, and diarrhea. (See 'Clinical presentation' above.)

- A gastrointestinal vasculitis should be suspected in patients with clinical features of gastrointestinal ischemia in the absence of risk factors for atherosclerotic vascular disease. In patients without an established diagnosis of vasculitis, the presence of associated systemic or constitutional symptoms in combination with single and/or multiorgan dysfunction is suggestive of a vasculitis. However, the absence of these features does not exclude gastrointestinal vasculitis. (See 'Our approach' above.)

- Our approach to evaluating the patient with possible gastrointestinal involvement of vasculitis depends on whether the patient has an established diagnosis of vasculitis or whether they are presenting for the first time with gastrointestinal symptoms. For patients who have an established diagnosis, the evaluation is focused on ruling out other causes of gastrointestinal symptoms followed by treatment of the underlying vasculitis. (See 'Our approach' above.)

- In patients who do not have a known vasculitis but present with predominantly gastrointestinal symptoms and other findings suspicious for a systemic vasculitis, additional testing will vary depending on the constellation of symptoms. The diagnosis of individual vasculitides is generally guided by the pattern of organ involvement, histopathologic features, and characteristic findings on abdominal imaging studies. The approach to diagnosing different forms of vasculitis are discussed separately in the appropriate topic reviews. (See "Overview of and approach to the vasculitides in adults", section on 'Diagnostic approach'.)

- Pertinent studies in patients with suspected mesenteric ischemia include serum lactate levels, as well as abdominal imaging studies which includes either computed tomography

(CT) angiography of the abdomen with intravenous contrast. (See 'Laboratory testing' above and 'Vascular imaging' above.)

- Endoscopic findings in patients with vasculitis of the gastrointestinal tract are usually nonspecific. Endoscopy should be performed with caution, if required, in the evaluation of a patient with suspected gastrointestinal involvement from an acute flare of vasculitis because of the elevated risk of perforation in the setting of an edematous, ischemic colon. (See 'Endoscopy' above.)

- The two principal components of the treatment of intestinal vasculitis are treatment of the underlying disease, usually with immunosuppressive agents, and therapy aimed at the gastrointestinal symptoms. For most patients with mesenteric ischemia, timely control of the vasculitis should lead to symptom resolution. Surgical intervention is necessary in patients with mesenteric infarction or with intestinal perforation. (See 'Management' above.)

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GRAPHICS

Major categories of mimics of vasculitis

Infectious causes (eg, endocarditis, HBV, HCV, HIV)
Atherosclerosis
Thromboembolic disease
Congenital causes (eg, aortic coarctation, middle aortic syndrome)
Hereditary disorders (eg, Marfan syndrome, Ehlers-Danlos syndrome)
Fibromuscular dysplasia
Hypercoagulable states (eg, APS, TTP)
Vasospastic disorders (eg, RCVS, drug exposures)
Other multisystem inflammatory disorders (eg, sarcoidosis, Susac syndrome)
Malignancy (eg, lymphoma, leukemia)
Iatrogenic (eg, postradiation therapy)
IgG4-related disease

HBV: hepatitis B virus; HCV: hepatitis C virus; HIV: human immunodeficiency virus; APS: antiphospholipid syndrome; TTP: thrombotic thrombocytopenic purpura; RCVS: reversible cerebral vasoconstriction syndrome; IgG4: immunoglobulin G4.

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