Acute acalculous cholecystitis: an unusual presenting feature of Churg–Strauss vasculitis

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First described in 1951, the incidence of Churg–Strauss vasculitis (CSV) is 2.5 per 100 000 people per year. It is characterized by pulmonary and systemic necrotizing vasculitis, vascular or extravascular granuloma, eosinophilia and tissue infiltration by eosinophils, and nonspecific elevation in the immunoglobulin E level. It is also found in those with asthma and allergic rhinitis or sinonasal polyposis. Coarse granular and perinuclear antineutrophil cytoplasmic antibodies can be detected in 38%–50% of patients. Pathologically, CSV is characterized by small-vessel angiitis and extravascular necrotizing granulomas, usually containing eosinophilic infiltrates.

We describe an unusual presentation of CSV: acute acalculous cholecystitis.

Case report

A 38-year-old woman presented with a 24-hour history of persistent epigastric pain. She had experienced similar attacks over the past month, so her family physician arranged for abdominal ultrasonography, which demonstrated thickening of the gallbladder wall and possible sludge. The patient had a history of allergic rhinosinusitis and poorly controlled asthma, so she was treated for presumed gastroesophageal reflux as a contributing factor to her asthma.

On physical examination, the patient had respiratory wheezing bilaterally, epigastric pain and a positive Murphy sign. Laboratory test results showed an elevated leukocyte count (18.5×10^9/L) with marked eosinophilia (7.9×10^9/L), normal liver enzymes and an elevated serum lipase level (80 U/L). The diagnosis was acute cholecystitis. The patient received antibiotics, but because she had no clinical improvement over the initial 24 hours we sent her to the operating room for a laparoscopic cholecystectomy. The gallbladder appeared inflamed, consistent with the preoperative diagnosis. The procedure was uncomplicated, and the patient went home 2 days postoperatively.

The patient returned to the emergency department on postoperative day 4 with nausea, diffuse epigastric and chest pain and a prominent cough. Her leukocyte count was 22.2×10^9/L, predominantly eosinophils (12.0×10^9/L). Liver enzyme levels were normal, but the serum lipase was again elevated (115 U/L). An ultrasound and a computed tomography (CT) scan of the abdomen showed a small amount of fluid in the gallbladder fossa but no evidence of a collection that would arouse concern for an abscess or bile leak. The serum troponin T level was elevated (0.11 mg/L) but the serum creatine kinase level was normal (61 U/L). The electrocardiogram showed T-wave inversion in the inferior and lateral leads. A general internal medical consultation led to echocardiography followed by urgent cardiac catheterization, which demonstrated normal coronary arteries and no abnormalities of wall motion, findings that led to a presumptive diagnosis of myocarditis. After consultation with the allergy and immunology service, serologic testing revealed an acute inflammatory process with an elevated C-reactive protein and erythrocyte sedimentation rate (17.5 mg/L and 59 mm/h, respectively), a marked elevation in her immunoglobulin E level (510×103 U/L), but antinuclear antibodies, anti-double stranded DNA, coarse granular and perinuclear antineutrophil cytoplasmic antibodies were not detectable, and her extractable nuclear antigen panel was negative. Pathological examination of the gallbladder specimen indicated eosinophilic inflammation with an associated small-vessel vasculitis (Fig. 1) but no gallstones or sludge.

We diagnosed CSV based on marked peripheral eosinophilia, previously known atopy with sinusitis and poorly controlled asthma, biopsy-proven small-vessel eosinophilic vasculitis and a systemic vasculitis with myocarditis. The patient received steroids parenterally. A CT scan of the sinuses demonstrated abnormalities consistent with CSV. A chest radiograph showed no evidence of pulmonary infiltrates, but these had been present on earlier investigations by her family physician. Her eosinophil count has remained suppressed with corticosteroid therapy. She has had no recurrence of her chest or abdominal pain, and her asthma has been asymptomatic.

Discussion

Though pulmonary symptoms are the most common clinical features of CSV, other systems involved include dermatologic, neurologic, cardiac, renal and gastrointestinal. Involvement of these systems can result in symptoms related

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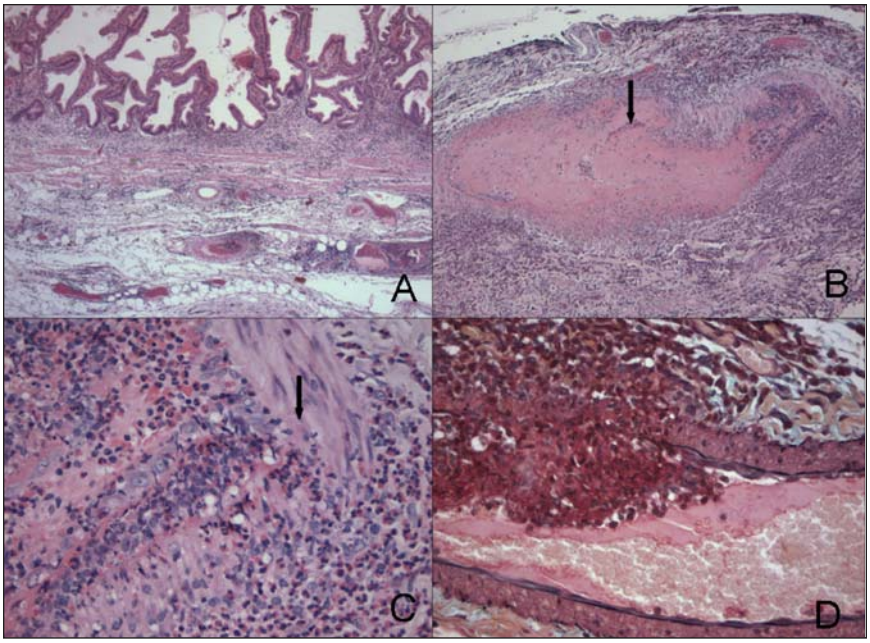

FIG. 1. The excised gallbladder specimen. (A) Full-thickness gallbladder section shows dense inflammation in the wall (hematoxylin–eosin, original magnification × 20). (B) Deep muscular artery is partially destroyed by inflammation and contains thrombus (arrow) (hematoxylin–eosin, original magnification × 40). (C) Mixed inflammation with a predominance of eosinophils infiltrating the artery wall (arrow) (hematoxylin–eosin, original magnification × 400). (D) Inflammation destroying muscle and black elastic fibres of the artery (elastic trichrome stain, original magnification × 200).

to peripheral neuropathy, myocarditis, glomerulonephritis and palpable purpuric lesions of the skin. Cardiac manifestations tend to be the major cause of death, accounting for up to 48%.[1]

Gastrointestinal manifestations of CSV include gastroenteritis, ileal or colonic ulcers with subsequent bleeding, ischemia and perforation.[2] Acute cholecystitis has been described through rare reports in the literature, as either calculous or acalculous.[3–5] In our patient, eosinophilic infiltration of the gallbladder wall and granuloma formation around arterioles in the absence of gallstones made for the diagnosis of cholecystitis.

The American College of Rheumatology recognizes CSV if 4 of 6 criteria are met, including asthma, eosinophilia (> 10% on differential), mononeuropathy, transient pulmonary infiltrates, paranasal sinusitis, and a biopsy specimen containing a blood vessel with extravascular eosinophils. Our patient exhibited all features except for mononeuropathy.

This case highlights a common surgical presentation resulting from a much less common immunologic process. General surgeons should be aware that gastrointestinal manifestations of CSV can occur, and marked peripheral eosinophilia should alert the clinician to the possibility of this rare disorder.

Competing interests: None declared.

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