

# Eosinophilic granulomatosis polyangiitis associated with acute acalculous cholecystitis

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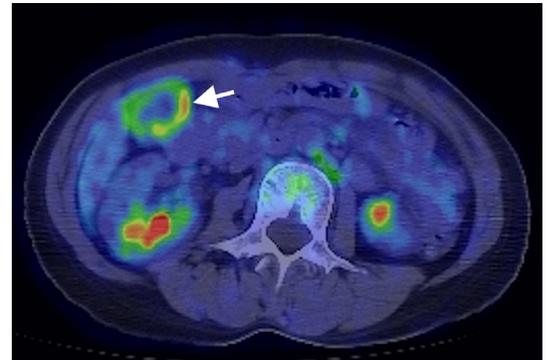
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## DESCRIPTION

A 53-year-old man presented with a 2-week history of progressive skin rash, numbness, arthralgia and myalgia, as well as a 9-month history of cough and wheezing. He was diagnosed with new-onset asthma and underwent inhaled steroid therapy; however, this treatment was ineffective for cough and wheezing. His medical history also included intractable chronic sinusitis, and he denied abdominal pain. Physical examination revealed erythema, purpura and bullae on the extremities. Laboratory test results showed a white blood cell count of 24 700  $\mu$ /L with hypereosinophilia (13 585  $\mu$ /L), lactate dehydrogenase of 300 U/L,  $\gamma$ -glutamyl transpeptidase of 207 U/L, alkaline phosphatase of 596 U/L, C reactive protein of 6.5 mg/dL and an increased myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA) titre. A nerve conduction study showed decreased amplitude with axonal neuropathy. Abdominal ultrasonography revealed gallbladder wall thickening without obvious gallstones (figure 1). Fluorodeoxyglucose positron emission tomography (FDG-PET) revealed abnormal tracer accumulation in the gallbladder wall without any luminal uptake, typically referred to as 'the rim sign' (figure 2), which suggested acute acalculous cholecystitis (AAC). Histopathological evaluation of a skin biopsy specimen showed eosinophilic infiltration and granuloma. Based on the clinical presentation and elevated MPO-ANCA titre, the patient was diagnosed with eosinophilic granulomatosis with polyangiitis (EGPA). The patient's symptoms including those associated with AAC improved after intravenous pulse corticosteroid therapy, and no relapse occurred after the 5-year follow-up.

AAC, defined as an acute necroinflammatory disease of the gall bladder in the absence of cholelithiasis, accounts for approximately 10% of all cases of acute cholecystitis and shows



**Figure 2** Fluorodeoxyglucose positron emission tomography showed abnormal accumulation in the gall bladder wall with no uptake in the lumen (the rim sign, arrow).

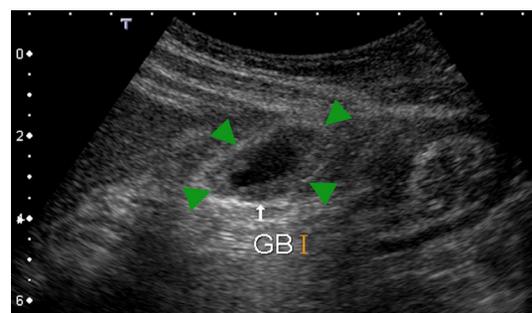
multifactorial pathogenesis.<sup>1</sup> It is strongly associated with a variety of clinical conditions, the most common being diabetes mellitus, malignancies, congestive heart failure and shock.<sup>2</sup> Systemic vasculitides, including EGPA, polyarteritis nodosa, immunoglobulin A vasculitis, cryoglobulinaemic vasculitis and giant cell arteritis may predispose individuals to AAC.<sup>3</sup> Organ hypoperfusion is implicated as a pathogenetic contributor to AAC.<sup>4</sup> Laboratory tests in patients with AAC show non-specific results and include elevated serum bilirubin, alkaline phosphatase and aminotransferases.<sup>5</sup> Radiological features suggestive of AAC include

## Patient's perspective

I was told that acute cholecystitis would require surgical removal, but it was good to know that it improved with medication alone.

## Learning points

- ▶ Acute acalculous cholecystitis (AAC) accounts for approximately 10% of all cases of acute cholecystitis.
- ▶ Systemic vasculitides, including eosinophilic granulomatosis with polyangiitis, may predispose individuals to AAC.
- ▶ AAC associated with rheumatic or autoimmune diseases may possibly show a better response to early immunosuppressive therapy without the need for surgical intervention.



**Figure 1** Abdominal ultrasonography showed thickening of the gallbladder wall (arrowheads) but no obvious gallbladder stones.



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gallbladder wall thickening, the sonographic Murphy's sign and visualisation of pericholecystic fluid.<sup>6</sup> FDG-PET in our patient revealed increased FDG uptake in the gallbladder wall without any luminal uptake, typically referred to as the rim sign.<sup>7</sup> AAC associated with rheumatic or autoimmune diseases may possibly show a better response to early immunosuppressive therapy without the need for surgical intervention.<sup>8</sup>

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