

# Anchoring bias in a case of recurrent abdominal pain

Scott Keeney,<sup>1</sup> Alexandra Halalau<sup>2</sup>

<sup>1</sup>Internal Medicine, Beaumont Health System, Royal Oak, Michigan, USA

<sup>2</sup>Beaumont Health System, Oakland University William Beaumont School of Medicine, Rochester, Michigan, USA

## Correspondence to

Dr Scott Keeney,  
Scott.Keeney@beaumont.edu

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## DESCRIPTION

A 43-year-old man with a history of idiopathic acute pancreatitis and pancreatic pseudocysts presented to the hospital with acute abdominal pain, vomiting and hypotension. He had been hospitalised on multiple occasions for recurrent, self-limited abdominal pain accompanied by hypotension. Given his history, the symptoms were typically attributed to acute chronic pancreatitis, despite absence of lipase elevation and pancreatic pathology on cross-sectional imaging. He had received multiple imaging studies and was scheduled for an elective cholecystectomy as biliary colic was a suspected aetiology for his symptoms.

On hospital presentation, lipase level was 12 U/L (10–65 U/L) and a CT of the abdomen demonstrated ascites and intestinal oedema without pancreatic inflammation or calcification (figure 1). Empiric antibiotics were initiated. On further enquiry, the patient described intermittent swelling of his extremities without urticaria for years and provided a picture of his swollen hand during one of these episodes (figure 2). Additionally, family history disclosed that two of his siblings were diagnosed with hereditary angioedema and thus suspicion for this condition was raised. A screening C4 level was <6 mg/dL (12–43 mg/dL) and a C1-inhibitor level was 5 mg/dL (19–37 mg/dL), confirming the diagnosis of type 1 hereditary angioedema due to C1-inhibitor deficiency (C1-INH-HAE). The patient improved with supportive care and was referred to an immunologist.

C1-INH-HAE is a rare genetic disorder with an autosomal dominant inheritance pattern characterised by recurrent attacks of oedema involving various organ systems including the skin, respiratory mucosa and intestinal tract.<sup>1</sup>



**Figure 1** Ascites and intestinal oedema during an attack of hereditary angioedema.



**Figure 2** Diffuse hand swelling during an attack of hereditary angioedema

The pathophysiology of angioedema in this condition is related to loss of C1-inhibitor-mediated control of the plasma kallikrein-kinin (contact) system, the downstream effect being excess bradykinin release and thus vascular permeability and oedema. Additionally, C1-inhibitor controls activation of the complement cascade and its deficiency leads to a permanent reduction in C4 levels. As a result, C4 is the recommended screening test as its level will be low even between attacks.<sup>2</sup> There are two variants of C1-inhibitor: deficiency (type 1) or dysfunction (type 2).<sup>2</sup> If C4 is low and C1-inhibitor is less than 50% of normal, the patient has type 1 C1-INH-HAE. If C4 is low and the C1-inhibitor level is normal or high, the diagnosis of type 2 C1-INH-HAE is made by demonstrating reduced activity of C1-inhibitor.<sup>2</sup> Treatment involves avoidance of triggers (trauma, infectious disease, stress),<sup>1</sup> on demand subcutaneous or intravenous therapies for acute attacks, and prophylactic therapy if attacks are frequent or severe enough.<sup>2</sup>

Hereditary angioedema is an infrequent cause of recurrent abdominal pain. Delay in recognition can result in significant morbidity or mortality, as well as unnecessary imaging studies and abdominal surgery. Given our patient's history of pancreatitis, the possibility of an alternative cause was not entertained despite lack of objective data. He fell victim to the effects of anchoring bias, or the tendency to make a judgment based on previous information.

**Contributors** SK and AH are guarantors of this case report and accept responsibility for the overall content of the case report. SK



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## Learning points

- ▶ Hereditary angioedema without urticaria should be suspected in patients with recurrent, self-limited abdominal pain. Early diagnosis can reduce future unnecessary imaging studies and surgeries and can prevent future attacks.
- ▶ A C4 level is the recommended screening test, and if low the diagnosis is confirmed by a level of C1-inhibitor less than 50% of normal (type 1 C1-INH-HAE) or reduced activity of C1-inhibitor (type 1 C1-INH-HAE).<sup>2</sup>
- ▶ Treatment involves patient education, avoidance of triggers, pharmacotherapy to abort attacks, and discussion of the benefits and risks of preventive medications.<sup>2</sup>

and AH were involved in the idea to submit this case report and are responsible for writing the case report.

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