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Hereditary angioedema with recurrent abdominal pain and ascites

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Abbreviations used

C1 INH: C1 esterase inhibitor
HAE: Hereditary angioedema
nl: Normal value

Hereditary angioedema (HAE) is a disorder characterized by decreased levels or function of complement C1 esterase inhibitor (C1 INH) and recurrent episodes of soft tissue swelling.^{1,2} Gastrointestinal involvement may cause severe abdominal pain, suggesting acute abdomen.³⁻⁵ We report a patient with HAE who was seen on two occasions with severe abdominal pain. Determination of the cause of this pain was made more difficult by the presence of significant ascites as determined by abdominal ultrasonography. On both occasions the patient was treated medically, and the ascites and other abdominal symptoms resolved within 3 days. Ascites has not, to our knowledge, been previously described with intestinal involvement in HAE.

CASE REPORT

A 19-year-old woman presented with known HAE. At the time of her initial diagnosis, the values were C1 INH less than 6 mg/dl (normal value [nl], 5 to 30 mg/dl) and 9% of activity for the functional assay of inhibitor activity (0% to 40% is presumptive evidence of HAE). At the time of admission to the hospital, she had a 1-day history of abdominal pain, fever, and protracted vomiting. The patient did not have any evidence of angioedema elsewhere.

Medical history was significant for multiple episodes of angioedema associated with minor trauma. Her family history was significant for HAE in a sibling and a paternal uncle. The patient was taking stanozolol 2 mg/day and oral contraceptive pills.

On examination, the patient appeared to have severe abdominal pain. Her temperature was 100° F (oral), heart rate was 100 beats/min, respiratory rate was 16 breaths/min, and blood pressure was 90/50 mm Hg. Results of examination of head, eyes, ears, nose, and throat and of the heart and lungs were unremarkable. The abdomen was soft with diffuse tenderness, and bowel sounds were present. Results of rectal and pelvic examinations were unremarkable. Significant laboratory values were as follows: hemoglobin, 14 gm/dl; hematocrit, 43.9%; white blood cell count, 14,900 cells/m³. Differential was 60% neutrophils, 14% bands, 18% lymphocytes, and 1% monocytes; C4 was less than 4 mg/dl (nl, 5 to 30 mg/dl). Serum glutamate oxaloacetate transaminase value was 44 IU/L (nl, 1 to 30 IU/L), and serum glutamate pyruvate transaminase value was 63 IU/L (nl, 1 to 30 IU/L). Results of chest x-ray were negative, and abdominal

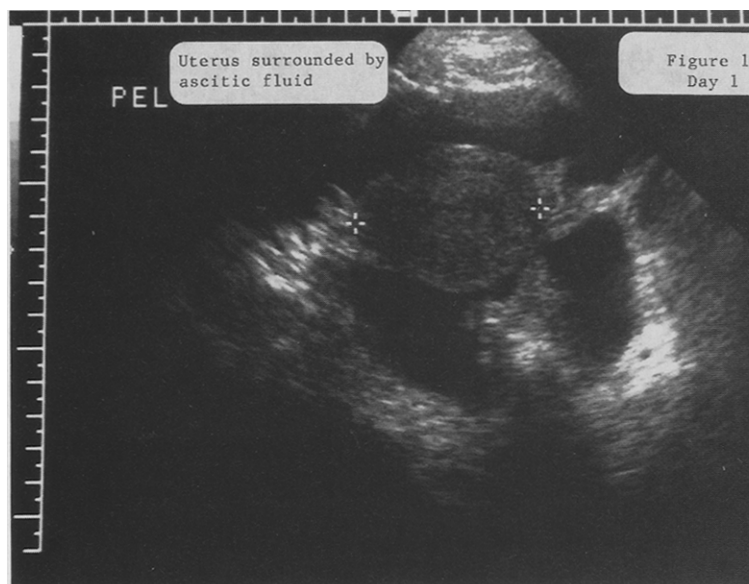


FIG. 1. Uterus surrounded by ascitic fluid (day 1).

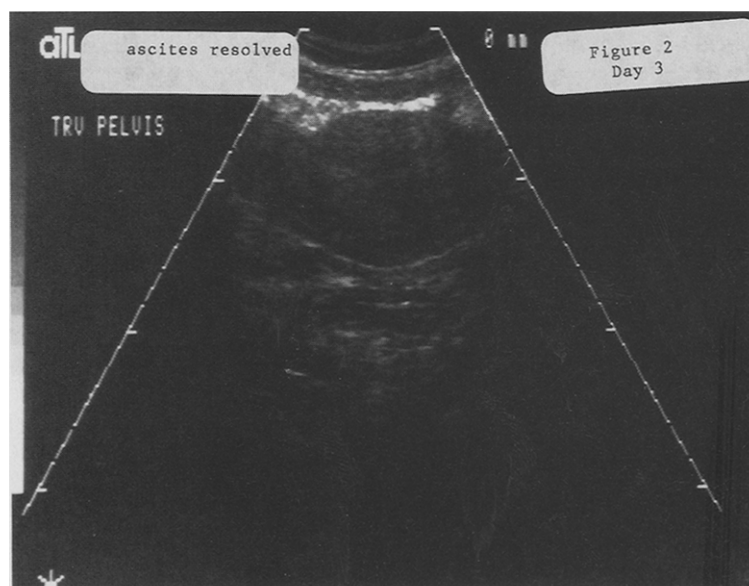


FIG. 2. Ascites resolved (day 3).

radiographs revealed ascites but no evidence of obstruction or ileus.

Ultrasonography of the abdomen (Fig. 1, day 1) revealed a large amount of free abdominal fluid around the liver and in the pelvis (estimated to be 350 to 500 ml in volume).

Urinalysis showed 2+ protein, and results were positive for ketones and nitrates with 0 to 2 white blood cells and 1+ bacteria. Paracentesis was performed, but no fluid was aspirated.

Despite considerable enthusiasm for an exploratory laparotomy, the patient was treated with intravenous fluids, diphenhydramine, and methylprednisolone. Her

stanozolol dose was also increased to 6 mg/day. Her symptoms decreased several hours after initiation of therapy. Repeat ultrasonography on the third hospital day showed complete resolution of ascites (Fig. 2).

She was discharged while receiving 2 mg of stanozolol per day but continued to take oral contraceptive pills. Two weeks later, she had a similar episode of abdominal pain with similar laboratory results and repeat ultrasound findings of ascites. Her repeat C4 value was less than 4 mg/dl. She was treated as before and was discharged while receiving 6 mg of stanozolol per day; oral contraceptive pills were discontinued. The patient has been free of

symptoms for the ensuing 12 months, and her C1 INH level has risen to within normal range.

DISCUSSION

HAE is manifested by episodic attacks of non-pruritic, nonpitting angioedema of the extremities, face, and upper airway.¹ The attacks may occur spontaneously or may be precipitated by trauma, anxiety, or emotion. Increased frequency of HAE attacks has been reported in women taking oral contraceptives.⁵

Involvement of the gastrointestinal tract in HAE may produce abdominal pain, nausea, vomiting, and diarrhea as a result of obstruction produced by bowel wall edema.^{6,7} Patients experiencing intestinal involvement may be seen with findings consistent with an acute abdomen.

The patient discussed in this case report was seen with two episodes of HAE mimicking an acute abdomen and underwent surgical evaluation. The findings of abdominal tenderness, fever, leukocytosis, and a left shift in the differential white blood cell count led to serious consideration of exploratory abdominal surgery. Management was made more difficult by ultrasonographic studies of the abdomen showing significant ascites, which to our knowledge has not been previously reported in HAE. Disappearance of the ascitic fluid in 3 days, concomitant with the resolution of abdominal symptoms, and its reappearance during a later episode suggest that the ascites was a manifestation of the HAE. Failure to recognize this concom-

itant finding in patients with HAE may lead to unwarranted surgical procedures because the nature of abdominal symptoms may be further obscured by the ascites, and exploratory laparotomy may seem advisable. We report this case so that clinicians will not be confused by the finding of ascites on ultrasonographic studies of the abdomen during an acute abdominal attack in patients with HAE. In some patients ascites may accompany other manifestations of abdominal HAE.

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Urinary eosinophil-derived neurotoxin in Churg-Strauss syndrome

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