

Recurrent Retroperitoneal Angioedema



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We present 2 similar cases seen at our emergency department (ED). Both were women, patient 1 (P1) was 72 years old and patient 2 (P2) was 57 years old. Both experienced severe cramping lumbar-abdominal pain with no fever, diarrhea, constipation, or dysuria, and both had unremarkable abdominal physical examinations. The patients had no personal or family history of gastrointestinal disease. P1 had arterial hypertension on angiotensin-converting enzyme (ACE)-inhibitor therapy and previous breast cancer without evidence of recurrence. At first ED admission, P2 had moderate C-reactive protein and lipase elevation (2.5 upper normal value). Blood tests including total serum IgG levels, erythrocyte sedimentation rate, antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, and serum protein electrophoresis were unremarkable. In both patients, symptoms resolved in 36 hours after taking painkillers, and the patients were discharged. Both patients returned to the ED within a week of the initial presentation having recurrence of the symptoms. P2 had transient CRP and lipase elevation (1527 to 213 UI/L in 12 hours). Abdominal contrast-enhanced computed tomography (CT) scans in both patients showed no signs of intestinal perforation/occlusion, lesions of abdominal organs, pathologic masses, or ascites. With minimal differences, CT scans showed heterogeneous hypodense “tissue” behind the pancreas, in the intercavo-aortic and celiac areas, and in the para- and perirenal area (also behind the renal fascia bilaterally) extending to the iliac-femoral axis and pelvis. The pictures were interpreted as a pseudohypertrophy of the retroperitoneum caused by abundant edema of the adipose tissue (Figures 1 and 2). This pattern is different from that observed in fibrosis, which usually causes organ encasement, especially to the ureters. Neither patient had imaging suggestive of pancreas involvement. Based on the radiologic finding suggestive of neoplasia, the patients were hospitalized. As in the previous episode, symptoms resolved in 36 hours without specific intervention. A CT scan repeated within 3 weeks showed complete resolution of the retroperitoneal edema (Figures 3 and 4). Nuclear magnetic



FIGURE 1. Abdominal CT scan (coronal view) showing pseudohypertrophy of the retroperitoneum caused by abundant edema of the adipose tissue.

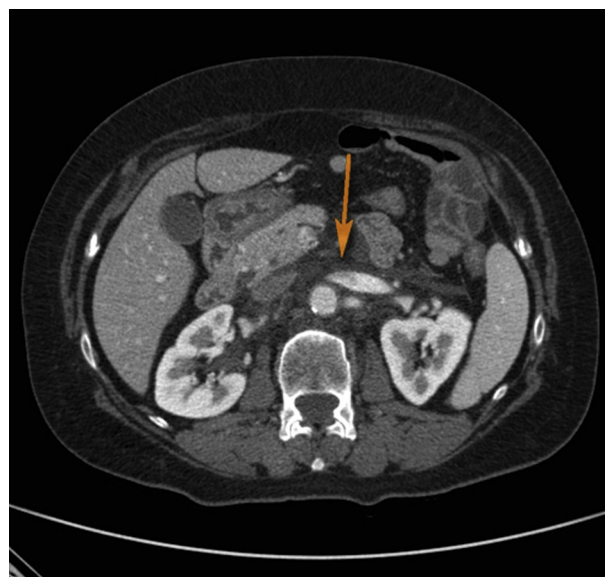


FIGURE 2. Abdominal CT scan (axial view) showing pseudohypertrophy of the retroperitoneum caused by abundant edema of the adipose tissue.

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FIGURE 3. Control abdominal CT scan (coronal view) performed within 3 weeks, showing complete resolution of the retroperitoneal edema.

resonance confirmed the negative results, thus definitively eliminating the concern about neoplasia. The colonoscopy, gynecological examination, and orthopedic evaluation did not reveal significant abnormalities. Conditions underlying angioedema (such as congestive heart failure, cirrhosis, fibroids, or pancreatitis) were ruled out.

Both patients had normal C1-inhibitor function and no mutations in factor XII.

After the second ED admission, ACE-inhibitor was withdrawn in P1, but she experienced 9 additional attacks and P2 had 3 recurrences. Clinical course and radiologic findings were very similar at each episode. During intercritical periods, no abnormalities were detected.

Mast-cell-targeted treatment with antihistamines during acute attacks was not effective. Assuming a bradykinin-mediated edema, icatibant¹ was administered without relief of symptoms.

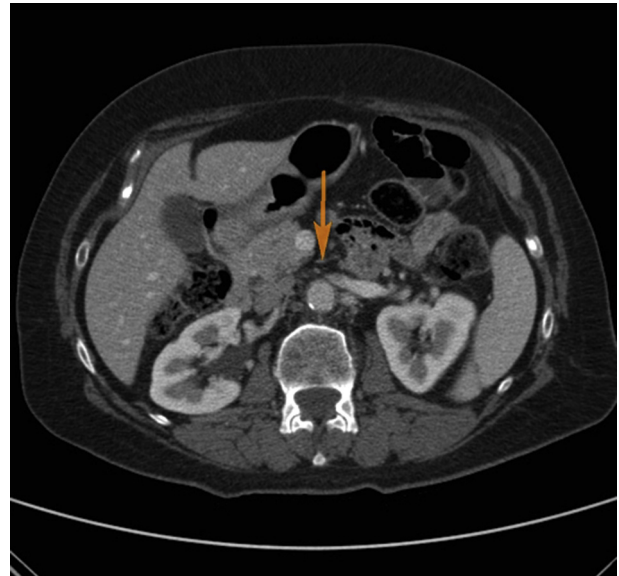


FIGURE 4. Control abdominal CT scan (axial view) performed within 3 weeks, showing complete resolution of the retroperitoneal edema.

This is the first report of isolated retroperitoneal edema with no thickening of omentum/intestinal walls or ascites as described in a previous case report.² In the presence of severe abdominal pain mimicking an acute surgical emergency with no abnormalities suggesting specific pathologic conditions, recurrent retroperitoneal angioedema should be considered. Definitive diagnosis is made *a posteriori*. The edema mediator remains undetermined. The absence of response to icatibant and antihistamines does not definitively exclude bradykinin nor mast cell-mediated edema. The benign course suggests using a symptomatic approach and surveillance of complications.

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