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ULTRASONOGRAPHIC IMAGES OF SPONTANEOUS INTRAMURAL HEMATOMAS OF THE INTESTINAL WALL IN TWO PATIENTS WITH CONGENITAL BLEEDING TENDENCY

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Gastrointestinal bleeding is a well-known event in patients suffering from congenital hemostatic disorders. In hereditary hemorrhagic telangiectasia it is the most frequent form of bleeding after epistaxis;[1] in hemophilia the reported incidence varied from 10 to 25%.[2] In contrast, spontaneous non traumatic hematoma of the digestive apparatus is a rare complication,[3,4] characterized by pain mimicking abdominal tumor,[5] gastric or esophageal obstruction,[6] or acute obstructive disease of the intestinal tract.[7] Massive or occult gastrointestinal blood loss is generally observed. Invasive and/or expensive procedures, such as endoscopy, CT-scan and or MRI, are often required to make a correct diagnosis.

Ultrasonography is rarely used to search for hematomas of the intestinal wall, and no imagings are available in congenital bleeding disorders. Therefore the aim of this report was to show the ultrasonographic features of this complication observed in two patients and relate them to the clinical course of these patients.

BS, a 47-year-old man, affected by severe hemophilia A (factor VIII < 1U/dL) with low titer of inhibitor (0.9 Bethesda Unit), was admitted to our Department for abdominal pain and subocclusive state mimicking appendicitis.

Hemoglobin was 14.3 g/dL and blood was present in his stools. Radiologic evaluation of the abdomen evidenced dilated loops of the small intestine. Ultrasound examination of the right lower quadrant showed a thickening of the intestinal wall with intraluminal fluid (Figure 1). The favorable clinical outcome obtained through treatment with factor VIII concentrates which resulted in complete resolution of ultrasonographic and radiologic findings after a few weeks prompted us to make a diagnosis of spontaneous

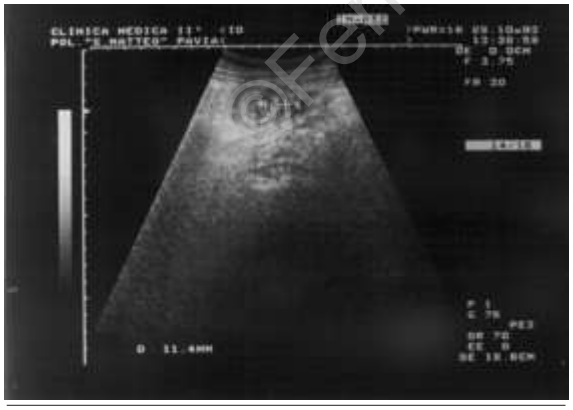

Figure 1. BS: ultrasound imaging of the small intestinal loop: thickening of the wall (11.4 mm) with narrowing of the lumen and the presence of fluid.

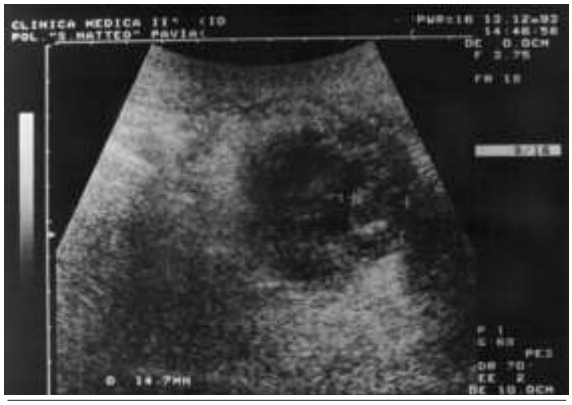

Figure 2. CF: ultrasound imaging performed in correspondence with the palpable mass in the left lower quadrant: conspicuous thickening of the intestinal wall (14.7 mm) characterized by a non homogeneous aspect and the presence of fluid in the lumen.

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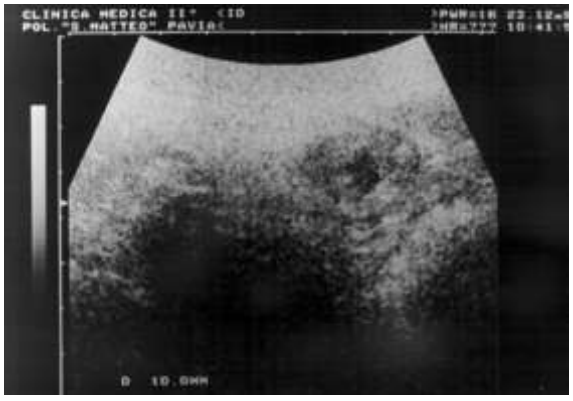

Figure 3. CF: ultrasound imaging of the same area after 10 days: significant reduction in the thickness of the intestinal wall (10 mm).

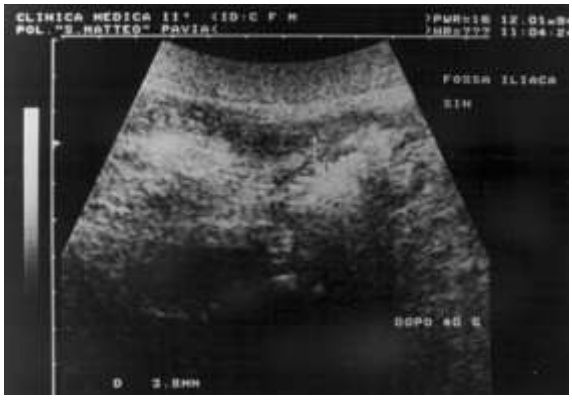

Figure 4. CF: ultrasound imaging of the same area after 40 days: normalization of the thickness of the intestinal wall (3.8 mm).

intramural hematoma of the terminal ileum in a hemophilic patient.

CF, a 55-year-old man affected by hereditary hemorrhagic telangiectasia complicated by sideropenic anemia due to recurrent epistaxes, was admitted to our Department for colicky left lower quadrant pain and a palpable mass in this area. Blood was present in his stools. Ultrasound examination revealed a non homogeneous thickening of the intestinal wall in correspondence with the palpable mass and the presence of fluid in the bowel lumen (Figure 2). Barium enema showed stenosis and rigidity along 9 cm of the sigmoid colon, compatible with a neoplastic formation. Colonoscopy evidenced a soft edematous stenosis suggestive of intramural hematoma of the intestinal wall, which was confirmed by histologic examination. Serial ultrasonographies demonstrated a progressive reduction in the intestinal wall involvement, ending in a complete resolution in about 40 days (Figures 3 and 4).

In patients with congenital hemorrhagic disorders, spontaneous hematomas of the intestinal wall, although rare, often show emergency pictures that are not easy to diagnose, and may require techniques such as CT scan and MRI, which are expensive, difficult to organize in a short period of time, and for these reasons not indicated for evaluating clinical course.

Endoscopic examination is generally adequate for identifying the site of bleeding in the upper or lower digestive tract when a profuse loss of blood is present.[2] Nevertheless, in congenital coagulopathies this invasive diagnostic approach implies replacement therapy with clotting factor concentrates, in order to avoid further hemorrhagic complications, and it is not strictly indicated in the monitoring of the clinical course of the bleeding. Furthermore, in hemophilic patients with inhibitors or in acquired hemophilia effective hemostatic therapy is often difficult.[8]

Ultrasonographic evaluation of the intestinal wall may be not sufficient to establish a diagnosis of intramural hematoma in all cases, but it allows the clinician to suspect this complication and follow its evolution.

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