A 45-year-old patient with no previous diagnosis presents at the emergency room because of gastrointestinal bleeding. Hemogram showed 8.6 mg/dl hemoglobin, 80,000 platelets, and signs of disseminated intravascular coagulation with D-dimers at 2000. During physical examination, we found penis varicosities and asymmetric leg hypertrophy (Fig. 1). Colonoscopy and angiography showed massive mesenteric angiomatosis (Fig. 2). An exploratory laparotomy confirmed the previously described findings, showing a massive mesenteric angiomatosis that included the internal and external iliac arteries and veins, which were unresectable (Fig. 3). The patient died a few months after diagnosis from massive intestinal hemorrhage. A diagnosis with Klippel-Trenaunay-Weber syndrome with mesenteric angiomatosis and an associated disseminated intravascular coagulation syndrome that led to angiomatosis (Kassabach-Merrit syndrome) was made.

**DISCUSSION**

Klippel-Trenaunay syndrome is a rare congenital mesodermal alteration with hemangiomas, varicosities, and asymmetric leg and arm hypertrophy secondary to congenital anomalous vascularization. It was originally described, in 1900, by Klippel and Trenaunay. In 1907, Weber added to the syndrome arteriovenous fistulas, a rare alteration (1). Alterations in the development of the mesoblastic germinal layer affect the angioblastic, lymphoblastic, and osteoblastic structures, thus determining a limitless number of associated malformations (2-4). This syndrome is a rare clinical pathological condition characterized by the presence of vascular alterations at different levels (5). These include hemangiomas, phlebectasies and varicosities (6). Intracranial, gastrointestinal, urinary or bronchial angiomatosis are rare, particularly when they are generalized. In legs and arms occasionally arteriovenous fistulas (7), lymphangiomatous abnormalities, and atrophy are observed. Also, macrodactyly (8), syndactyly, polydactyly, and oligodactyly can be observed. Some patients present with
rectal hemorrhage from colonic varicosities, and chronic anemia may also be present (5). Lower gastrointestinal bleeding must be treated by endoscopic hemostasis. Surgical resection of the affected segment must be reserved for failed therapies and cases of massive angiodysplasia (9). The present patient not only had features of this syndrome, but angiomatosis extended throughout the entire gastrointestinal tract. Moreover, this patient presented feature of the Kasabach-Merrit syndrome secondary to the massive underlying angiomatosis. Diagnosis can be reached visually when alterations are evident. However, patients will have to be evaluated using a strategy of noninvasive imaging procedures (1) such as ultrasonography, Doppler, CT, and MRI. A phlebography would be indicated to evaluate surgical treatment, and an arteriography when an arteriovenous fistula is suspected. Angio-MRI with gadolinium allows a greater precision in the diagnosis. Regarding differential diagnosis, vascular malformations may be primary manifestations of other sporadic or hereditary disorders that should not be mistaken for Klippel-Trenaunay syndrome—Maffucci, Gorham, Bannayan, Riley-Smith, Solomon, Proteus, etc. syndromes (10). Mortality of this syndrome is about 1%, with morbidity due to complications such as bleeding and platelet sequestration, internal hemorrhage, thrombophlebitis, aseptic cellulitis, heart failure in case of multiple or great size fistulas, bacteremia, osteomyelitis, and propensity to fractures (11). The genetic base of this disease is under continuous study. In 2004, Tian et al. (12) described two genetic defects associated to the angiogenic protein, VG5Q. The E133K mutation, found only in 4% of patients, leads this peptide to be “hyperactivate”, and, consequently, resulting in the stimulation of angiogenesis in a more aggressive form than that of normal protein VG5Q. In any case the genetic origin of this mutation remains uncertain, and only one chromosomal translocation has been found in a patient.

REFERENCES