INTRODUCTION

Spleen diseases are rare, being splenic abscesses, splenic cysts, benign tumors (hemangioma, lymphangioma and others), and malignant tumors (lymphomas, metastases and others)\(^1\). Primary benign tumors of the spleen are extremely rare and account for less than 0.007% of all tumors identified upon surgery and autopsy\(^2\). Splenic lymphangiomas (SL) are benign cystic tumors resulting from congenital malformations of the lymphatic system that appear as single or multiple lesions of the spleen\(^3\)\(^4\)\(^5\). SL mainly affect children and rarely manifest after 20 years of age\(^6\). The main clinical manifestations are abdominal pain, vomiting and a palpable mass, although these tumors are asymptomatic in most patients\(^2\)\(^7\)\(^8\). The treatment is eminently surgical. With the advent of minimally invasive surgery and after the first videolaparoscopic splenectomy performed by Delaitre and Maignien in 1991, various surgeons have adapted and improved this technique for the treatment of spleen diseases, especially for patients with hematological disease such as immune thrombocytopenic purpura\(^2\)\(^9\).

CASE REPORT

A 59-year-old white woman was referred to the specialty outpatient clinic of the Department of Hematology, University Hospital, Faculty of Medicine, Federal University of Triângulo Mineiro (HCFM-UFTM), with a one-year history of diffuse abdominal cramps mainly in the epigastric region and left hypochondrium that did not improve or worsen. The patient reported no urinary and intestinal abnormalities or weight loss. Her personal history revealed that the patient had been a heavy smoker for 40 years and had undergone gastric antrectomy with Billroth II reconstruction for an ulcer 19 years ago, in addition to two previous cesarean sections. Physical examination showed diffuse pain upon superficial and deep palpation of the abdomen, with the spleen being palpable 3 cm below the left costal margin. The laboratory tests were normal. Abdominal US showed an enlarged spleen with a heterogenous echotexture containing echogenic nodules and cystic areas. An abdominal CT with oral and intravenous contrast was requested, which revealed an enlarged spleen of diffusely heterogeneous density, as well as multiple hypodense and coalescent nodular images measuring on average 2 cm in diameter and enhancement after contrast injection (Figure 1). In view of these findings total laparoscopic splenectomy was indicated.

Surgery was performed under general anesthesia and the patient was placed in right lateral semidecubitus after introduction of a bladder and nasogastric tube. A 10 mm trocar was inserted through the umbilical scar (camera) and CO2 was insufflated at a pressure of 10-12 mmHg. Four other trocars (two 5 mm and two 10 mm) were inserted in the left upper quadrant of the abdomen. Inspection of the cavity revealed an enlarged spleen and nodules, in addition to abdominal wall and diaphragmatic adhesions. After inspection of the cavity for the identification of accessory splenic tissue, the short gastric vessels were ligated and clipped and the splenic vein and artery were double ligated with nonabsorbable suture. After lysis of adhesions, the organ was removed into a sterile plastic bag through a Pfannenstiel incision. The results of the anatomopathological and immunohistochemical tests (tumor positive for CD34 and CD31 antibodies and negative for CD68, AE1/AE3 and factor VIII) confirmed the diagnosis of SL (Figure 3). The spleen weighed 265 g and measured 14.0 x 8.5 x 6.0 cm. The patient showed good postoperative evolution and was discharged two days after surgery. After one year of outpatient follow-up, she continues to be asymptomatic and in good health.
The clinical presentation of intra-abdominal lymphangioma is variable. In children, this condition tends to show a shorter and acute duration of symptoms. In adults, the clinical symptoms tend to be mild and nonspecific, with the disease progressing over many months or years before its diagnosis due to the slow growth of the tumor. In a series of seven cases, Komatsuda et al. showed a close relationship between the occurrence of symptoms and the size of the spleen. The main clinical manifestations of SL are pain in the left upper quadrant, abdominal distension, loss of appetite, nausea, vomiting, and a palpable mass. However, cases presenting no symptoms have also been reported. Physical examination can be normal or reveal a palpable mass in the left upper quadrant. Routine laboratory tests and simple chest and abdominal X-rays generally show no abnormalities. The differential diagnosis is extensive and includes lymphoma (a more common malignant tumor), infarction, septic embolism, metastases (melanoma, breast, ovarian and lung cancer), and splenic cysts. In the present case, the patient was symptomatic and a mass was palpable in the abdomen. However, the age of the patient was not within the range in which SL is more prevalent.

US usually shows hypoechoic spaces that contain internal echoes. Low-density, well-delimited subcapsular cysts with thin walls that may contain mural calcifications are generally visualized by CT, suggesting a diagnosis of cystic lymphangioma. A “Swiss cheese” appearance of the spleen has been considered to be pathognomonic. The treatment of choice for intra-abdominal lymphangiomas, including SL, is complete surgical resection since other treatment modalities such as aspiration, drainage and irradiation have shown unsatisfactory results. Regarding SL, some investigators opt for conservative treatment in the case of small and asymptomatic lesions detected incidentally, reserving splenectomy for large, multiple or symptomatic lesions. However, surgical resection is recommended immediately after establishment of the diagnosis since a growing incidence of complications is observed over time, such as infection, hemorrhage, intestinal obstruction and tumor growth (that may prevent complete removal). The prognosis of intra-abdominal lymphangioma after resection is favorable. Recurrence is the main complication, which is demonstrated in 9.5% of patients, frequently after incomplete resection.

Video laparoscopic splenectomy can be performed with the patient in the supine (or modified lithotomy) position or placed in right lateral decubitus. Three or five port sites are used. According to the literature, the second approach is preferred since the right lateral decubitus position increases exposure, improves dissection and possibly reduces the number of trocars. The use of a laparoscopic vascular stapler, electrothermal bipolar vessel sealer (Ligasure®) and ultrasonic knife (Ultracision®) are options to facilitate the surgical steps of hemostasis, but these devices are expensive. In the present case, surgery was performed with the patient in the supine and right semidecubitus position. Five trocars (three 10-mm and two 5-mm trocars) were inserted and an ultrasonic knife was used.

**REFERENCES**