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Case Report

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INFERIORVENA CAVA LEIOMYOSARCOMA: CASE REPORT

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Abstract

The inferior vena cava leiomyosarcoma (IVC) is a rareneoplasm. It originates from the smooth muscle cells of the IVC tunica media. The tumor's clinic depends basically on its location and size. The best treatment, with impact on survival, is complete surgical resection.

Key Words: leiomyosarcoma, vena cava, neoplasia.

INTRODUCTION

Sarcomas are malignant tumors that originate from the mesenchymal tissue (1,2). The inferior vena cava leiomyosarcoma (IVC) is a rareneoplasm. Up to now, approximately 300 cases have been reported since the first autopsy diagnosis (3,4). This tumor originates from the smooth muscle cells of the IVC tunica media. It is known that this tumor occurs preferably in female adults in a ratio of 4: 1 with a mean age of 54.4 years (ranging from 15 to 83 years) (1,2,5).

Tumor clinic depends basically on its location and size (1,2). But its growth is insidious with very specific symptoms such as abdominal pain, weight loss, abdominal tumors, fever, weakness, anorexia, vomiting, night sweats and eventually with Budd-Chiari syndrome (2,4,6,7).

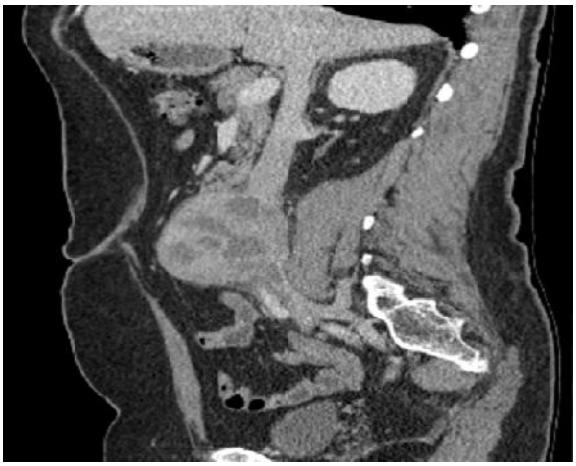
The best treatment, with impact on survival, is complete surgical resection of the tumor without compromising the surgical margins (2,4,6). Treatment options following resection of IVC leiomyosarcomas include primary repair, ligature or reconstruction with prosthesis and depend on the location of the tumor and invasion of adjacent organs and vessels, especially of the renal veins (5)

This article reports a rare case that was addressed at the Vascular Surgery Service of the Samuel Libânio Clinic Hospital in Pouso Alegre, MG, which should be included in the differential diagnosis of retroperitoneal masses, since its surgical planning has an impact on patient survival

CASE REPORT

A female patient, aged 54, was admitted to the Samuel Libânio Clinic Hospital, with colic type abdominal pain that started one year ago with worsening in the last six months associated with tenesmus and hematochezia. He also reported hyporexia and lost ten pounds in two months. Physical examination revealed palpable abdominal mass in mesogastrium, painful to mobilization, inelastic, approximately five centimeters in diameter. No laboratory abnormalities were found in laboratory tests. Full-length tomography of the abdomen in the portal / venous phase, showing expansive formation with contrast enhancement, with areas of liquefaction of the lower third of the inferior vena cava, 2 cm above the iliac bifurcation and with a large contact surface With the distal aorta and right common iliac artery measuring about 8.07 x 6.69 x 7.64 (LL x AP x CC); In addition to intraluminal tumor content extending to the level of the bifurcation.

Figure 1 Computed tomography, sagittal section, shows solid expansive formation in the right paramedian retroperitoneal region, in close contact with the lateral wall of the aorta and the right common iliac artery



DISCUSSION

Vascular leiomyosarcomas are rare neoplasms and may occur in the digestive system, respiratory system, skin, vessels of the myometrium, or in any other structure that has smooth muscles (1,2). They correspond to 2% of the leiomyosarcomas, affecting five times more, the veins (2, 4). Based on the location, the leiomyosarcoma of the IVC is classified into three categories. Segment I (lower): below the renal vessels (IVC infra renal); Segment II (medium): between the origin of the renal vessels to the hepatic vein (Retrohepatic IVC); Segment III (superior): between the origin

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of the hepatic veins to the right atrium (suprahepatic IVC) (6, 7). Most tumors originate in the middle segment (50.8%) and lower segment (44.2%).

VCI leiomyosarcomas are slow-growing and, because of their retroperitoneal location, often present with nonspecific, late and insidious symptoms, such as abdominal pain, weight loss, palpable abdominal mass, fever, weakness, anorexia, vomiting, night sweats and dyspnea (4,5,7,8). Tumors of the lower segment tend to present as lower right quadrant mass, lower back pain, lower limb edema and, rarely, deep vein thrombosis due to obstruction of IVC (4,7,8).

In cases of tumors located in the middle segment, the patient usually complains of pain in the upper quadrant and weakness (4). Renal veins may be involved, resulting in renovascular hypertension and, if occlusion occurs, the patient may have nephrotic syndrome (4,7,8).

Tumors in the upper segment may cause venous and suprahepatic thrombosis, leading to different degrees of Budd-Chiari syndrome (hepatomegaly, jaundice and ascites) due to obstruction of the hepatic venous flow (2,4,7). The main diagnostic tests are: ultrasound, computed tomography, angiotomography and nuclear magnetic resonance (8). The IVC leiomyosarcoma has a reserved prognosis (9).

According to the international papers, disease-free survival and the overall survival rate after surgery with free margins and wide resection were 31.4% and 7.4%, respectively (2).

The prognosis is better for tumors involving the mid segment of the inferior vena cava compared to the upper segment (7). The presence of abdominal pain (which occurs earlier in the middle segment due to the rich innervation of the adjacent organs), indicates a better prognosis, as well as the absence of palpable mass and radical tumor resection (2,6,8).

Regarding the surgical treatment, complete excision with free margin of tumor is ideal. However, when this is not possible, palliative debulking should be offered and complementary treatment performed (2,7). The survival rates of patients undergoing complete resection were 49.4% at five years and 29.5% at 10 years, while disease-free survival was 31.7% at five years and 7.4% at 10 Years (6). The role of chemotherapy and radiotherapy for retroperitoneal sarcomas is unclear (7).

CONCLUSION

There are few reports oflower vena cava leiomyosarcoma and the present work, in its limited coverage in the literature, contributes to the knowledge about the subject.

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