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Case Report

What do we know about intravascular fasciitis affecting inferior vena cava? management and report of a case

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ABSTRACT

Intravascular fasciitis is a rare entity characterized by the proliferation of myofibroblasts and giant cells in relation to small and medium-sized blood vessels. It characterized by a rapid growth and its morphology can simulate other malignant neoplastic processes of more aggressive behavior such as sarcomas. It belongs to a group of benign lesions that affect the subcutaneous tissue and fascia (sometimes to the muscle) sharing characteristics similar to proliferative fasciitis, nodular fasciitis and proliferative myositis. It can sometimes be confused with lesions typical of fibromatosis even. We report a case of a patient with a symptomatic retroperitoneal mass who underwent surgery. The intraoperative findings showed a tumor originating in the inferior vena cava. The histopathological study revealed an intravascular fasciitis.

Keywords: Intravascular fasciitis, Inferior vena cava, Nodular fasciitis, Proliferative myositis, Proliferative fasciitis

INTRODUCTION

Intravascular fasciitis (IF) is a rare, benign lesion characterized by the proliferation of myofibroblasts and giant cells in relation to small and medium-sized blood vessels.[1] It was first described by Patchefsky and Enzinger in 1981 as a variant of nodular fasciitis (NF).[2,3]

NF, like proliferative fasciitis (PF) and proliferative myositis (PM), belong to a group of benign lesions of soft tissues characterized by their rapid growth and that by their morphology can sometimes be confused with other malignant neoplastic processes of more aggressive behavior such as sarcomas.[4]

The FI is characterized by intravascular proliferation (or associated with the wall of arteries and veins of any size) of a large number of fusiform-like myofibroblasts on a hyaline matrix associated with the presence of giant cells.[1,5] They are mostly prominent nucleolus cells and abundant number of mitoses but without the presence of atypia.

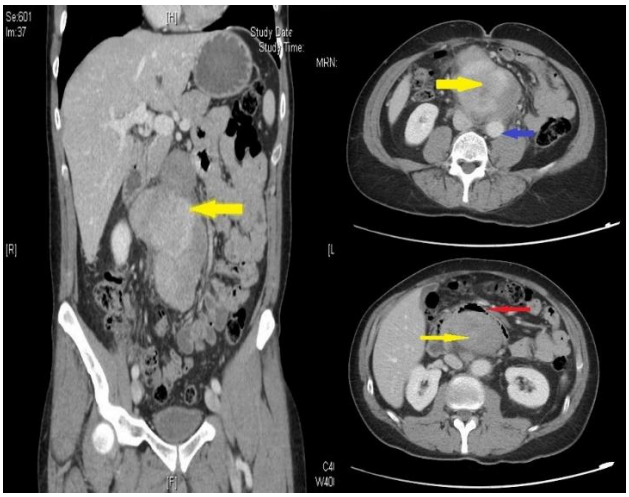
Immunohistochemically, myofibroblasts present positivity for vimentin and specific smooth muscle actin staining, being negative for keratin, S-100 protein, desmin, CD31, CD34 and betacatenin. Giant cells, as in other locations, express CD68.[1,6]

IF is a rare entity that belongs to a group of benign lesions of rapid growth and whose importance lies in the differential diagnosis that they pose with other malignant lesions such as sarcomas or other tumors. Next, we

present a case of inferior infrarenal vena cava IF that debuted as a retroperitoneal mass in our patient.

CASE REPORT

Patient of 45 years without antecedent of interest that refers to abdominal tumor of recent appearance. The physical examination reveals mass effect in mesogastrium, of stone consistency, not painful. Abdominal-pelvic CT showed a retroperitoneal mass of 140x120x100 mm, with a heterogeneous density, with hypervascularized zones and with calcifications in intimate contact with the third duodenal portion, displacing the inferior vena cava, impressing of its infiltration (Figure 1). Gastro-colonoscopy was performed without presenting significant alterations. The blood tests and hormonal study showed no alterations. Complementary abdominal MRI was produced that impressed with dependence on the third / fourth duodenal portion (Figure 2). Under suspicion of a malignant neoplastic process (duodenal GIST, retroperitoneal sarcoma ...) the patient was operated on.



The yellow arrows show the retroperitoneal mass. The red arrow indicates the relationship between the mass and the duodenum and the blue one the inferior vena cava.

Figure 1: Coronal and axial images of CT scan.

A bilateral subcostal incision was made for the opening of the abdominal cavity, mobilization of the duodenal frame by Kocher maneuver and reduction of hepatic angle by means of Catell for access to the retroperitoneal space. A polylobulated tumor mass was observed in intimate contact with the third duodenal portion but without any dependence on it. No pancreatic or intestinal involvement was observed part of the tumor. On the other hand, dependence of the tumor of the anterior aspect of the infra-renal inferior vena cava could be observed (Figure 3). A third and fourth portion duodenal resection with pancreatic preservation was performed with anastomosis of the second portion to the first jejunal loop. The tumor was completely resected and inferior infrarenal vena cava was previously vascular, and the ligature was made distally and proximally without prosthesis. There were no signs of peritoneal dissemination or hepatic involvement.

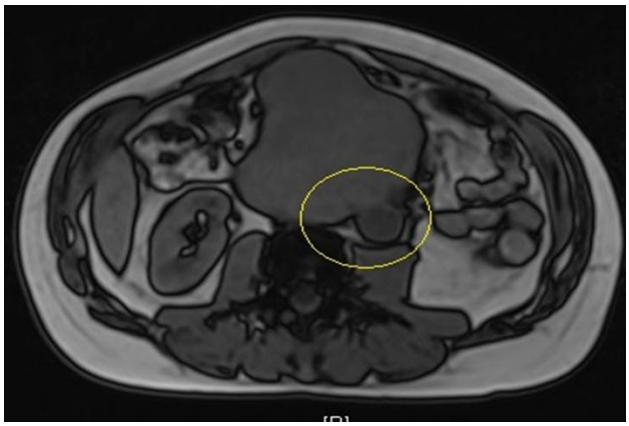


Figure 2: Retroperitoneal mass without plane of separation with inferior vena cava visualized in MRI.

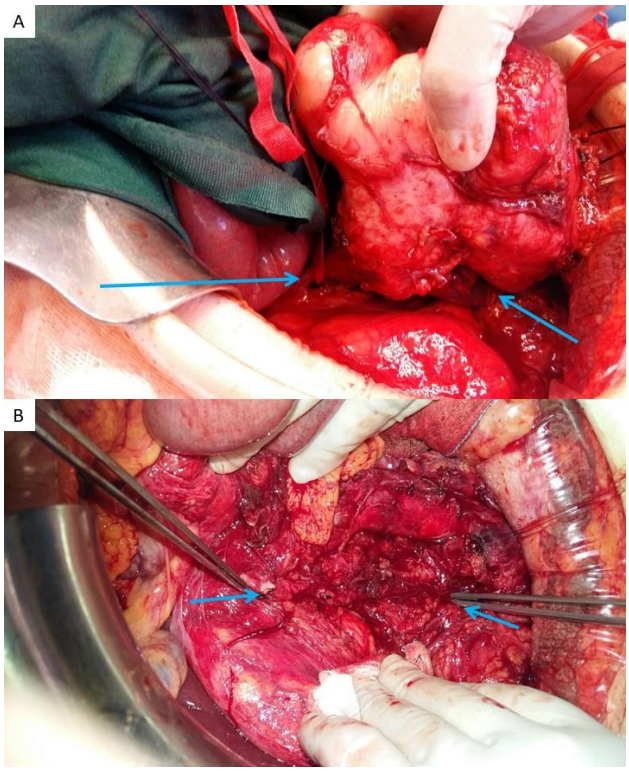


Figure 3: (A) Vascular control of infrarenal inferior vena cava. Origin of the tumor on vena cava noticed. (B) Ligation of inferior vena cava and tumor removed.

The macroscopic study of the surgical specimen revealed a tumor mass, nodular and polylobed of 153 x 85 x 112 mm depending on the anterior wall of the inferior vena cava (Figure 4). The histological analysis showed a mesenchymal tumor with fusocellular proliferation and nodal cells, with numerous mitoses without signs of atypia. Immunohistochemistry was positive for vimentin, actin, aml and calponin being negative for CD68, CD31, CD34, myoglobin, myogenin, D2-40 and betacatenin

compatible with intravascular fasciitis. The Ki67 expressed was about 3%. The resected duodenum showed no significant alterations or local infiltration data (Figure 5).

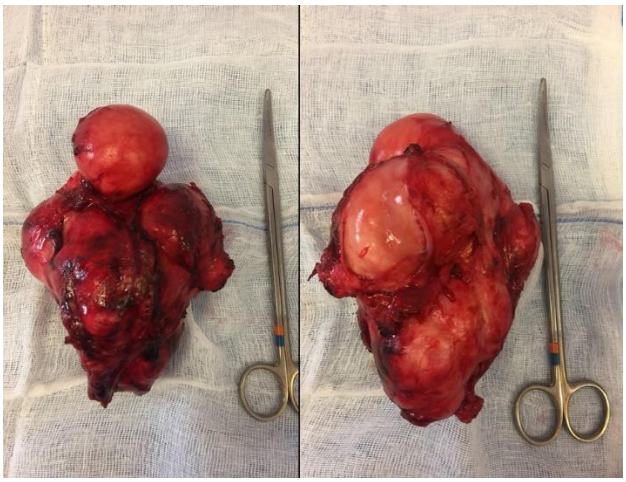


Figure 4: Anterior and posterior aspect of polylobulated mass originating in inferior vena cava.

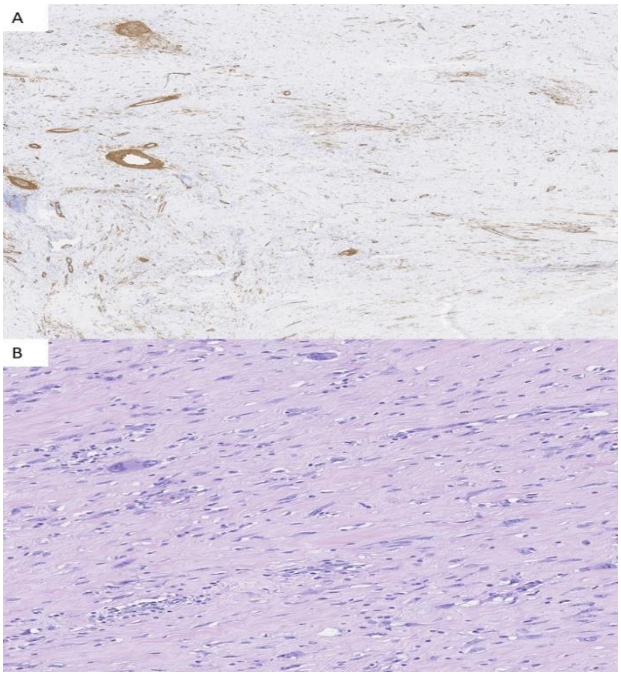


Figure 5: (A) Immunohistochemistry positive for vimentin, actin, aml and calponin. (B) Low expression of Ki67.

After surgery, the patient entered the Intensive Care Unit for surveillance and monitoring. The patient developed an important SIRS consisting of hypotension and tachycardia secondary to postoperative acute pancreatitis. Despite correct volume replacement and correction of hydroelectrolyte alterations, acute pancreatitis evolved to its necrohemorrhagic form, worsening the patient's condition, causing multiorgan failure refractory to treatment and vasoactive and respiratory support, dying on the fifth postoperative day.

DISCUSSION

IF belongs to a group of benign lesions that affect the subcutaneous tissue and fascia (sometimes to the muscle) sharing characteristics similar to PF, PM and NF.[7] IF is considered a rare subtype or variant of NF that affects medium- and small-caliber blood vessels.[2,8] PF, a term defined by Soule in 1962, was initially used to refer to cases of NF.[9,10] In 1975, Chung and Enzinger described 53 cases of PF, separating it from the NF term.[11] PF consists of a benign lesion affecting fat and fascia characterized by proliferation of fibroblasts and giant cells.[7] It is a non-neoplastic inflammatory process, whose cells are rapidly growing with abundant mitosis. PF and PM are clinically and histologically similar[9]. PM is a fast-growing reactive intramuscular lesion characterized by the proliferation of myofibroblasts and giant cells located between muscle fascicles.[12]

It can sometimes be confused with lesions typical of fibromatosis. However, in fibromatosis, the cells are arranged in bundles in a collagen stroma, lacking an inflammatory component, unrelated to blood vessels and lacking extravasated erythrocytes.[6]

Authors such as Dahl have proposed framing PF, NF and PM with the designation of soft tissue pseudoarachosarticular lesions.[12]

NF is histologically similar to PM and PF except that it is usually without giant cells.[5,13] The main difference between NF and IF is the involvement of blood vessels.[3] Patchefsky and Enzinger in 1981 described 17 cases of The IF is characterized by involving muscular arteries and veins suggesting vascular invasion and being confused with malignant lesions like sarcomas. It can affect the intimate, middle or adventitia of the vessel, as well as localizing at the perivascular level of soft tissues as in our patient.[3] Intravascular growth is exceptional.[2]

IF affects men and women equally. It is most commonly located in the upper, lower extremities and head and neck. Its etiology may be due to primary trauma or to an episode of previous venous thrombosis.[1,5,14]

It usually develops as a rapidly growing painless mass that can be made symptomatic by obstructing the lumen of the blood vessel during its growth.[5,15]

IF can originate from small intradermal vessels to large caliber veins. It has been described in arteries and veins of trunk and limbs (digital artery, palmar vein arch, anterior tibial vein, femoral vein) as well as in head and neck vessels, orbit and submucosa of the oral cavity.[2,5,16] Veins are affected more frequently than arteries, but both may be affected.

Arteriography and / or venography may not be sufficient for diagnosis.[5] Ultrasonography may aid in the diagnosis of soft tissue lesions but the combination of CT and MRI has been shown to be more useful.[5,15]

Differential diagnosis of retroperitoneal lesions should be made with other malignant neoplastic processes such as spindle cell carcinoma (positive for cytokeratins AE1 / AE3), melanoma metastases (positive for S-100 protein), gastrointestinal mesenchymal stromal tumors GIST) positive for CD117, solitary fibrous tumor (positive for CD34), liposarcomas, leiomyosarcomas, rhabdomyosarcomas, Edwin's sarcoma, epithelial angiosarcomas (positive for CD34 and CD31) as well as with peritoneal implants suggestive of peritoneal carcinomatosis. Other benign processes such as fibromatosis (positive for beta-catenin), pyogenic granuloma, hamartomatosis, adventitious periquistic disease, hemangioendotheliomas, leiomyomas, myofibroblastic tumor (Alk-1 positive) or Masson's tumor.[5,14,15,17-19]

Fine needle aspiration (FNA) of the lesion, as in cases of PF, may be an acceptable method for diagnosis, although the gold standard is the surgical excision of the lesion on suspicion of malignancy, thus avoiding the possibility of local dissemination during the procedure.[20,21] Some authors propose follow-up of the lesion, based on histology and as long as malignancy has been ruled out.

CONCLUSION

In conclusion, Intravascular fasciitis is a rare benign soft-tissue tumor that affects small and medium-sized veins and arteries. Clinically it presents as a rapidly growing tumor mass that can cause symptoms by compression or in relation to vascular occlusion. The diagnosis of IF is extremely difficult, being a low-incidence entity and presenting as a rapidly growing mass, easily confused with other tumors such as sarcoma of parts soft. The combination of the clinical history of the patient with the physical examination, the results of the imaging tests and the anatomopathological analysis is essential for diagnosis.

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