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Leiomyosarcoma of the Inferior Vena Cava Mimicking a Tumor of Surrounding Structures: Report of Two Cases

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Abstract

We report two cases of leiomyosarcoma diagnosed after a further analysis of the neoplasm. In the first case, a 61 years-old woman with a specific symptoms showing to the CT scan a soft tissue mass firstly suspected to grow from the caudate lobe of the liver with invasion of the Inferior Vena Cava (IVC). Laparotomy revealed a mass arising from the caval confluence with the left renal vein, in contact with caudate lobe. A tangential resection of vena cava was performed. In the second case, a 66 years-old man with Crohn disease reported asthenia and articular pain. MRI scan showed a hyperactive vascular mass between duodenum and head of pancreas making contact with the IVC, similar to a duodenal GIST. Surgical exploration revealed a solid mass originating from anterior wall of the IVC, with intravascular invasion. A partial venous resection with prosthetic patch reconstruction was performed. In both cases histology confirmed a cavalleiomyosarcoma, an extremely rare tumor. It involves predominantly the segment of the vein between the renal and hepatic veins. Surgical resection is the only choice for long term cure. The role of neoadjuvant and adjuvant therapy is still unclear.

Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is a rare clinical entity, whose current knowledge about management is based on case reports and small case series [1-3]. It accounts of approximately for 5 to 15% of all retroperitoneal tumors [4] and, because of its slow growing patterns, diagnosis is usually made lately. Since the first case report of segmental resection of leiomyosarcoma of the IVC was reported in 1928 [5], surgical resection remains the only therapeutic approach associated to prolonged survival [3]. The main goal of surgical resection is to achieve a complete excision of the tumor, to prevent recurrence and to preserve venous return [2,3,6]. The role of neoadjuvant and adjuvant therapies is not clear, as the rarity of this tumor does not allow comparison of large series [7-9]. We report two cases of leiomyosarcoma arising from the suprarenal tract of the IVC. Localization and suspected infiltration of adjacent organs made difficult the preoperative. Their origin from IVC was detected only intraoperatively.

Case Reports

Case #1

A 61-year-old woman presented with a 5-year history of recurrent right upper quadrant pain associated to occasional vomiting and diarrhea and 1-month history of bilateral leg swelling. She denied of appetite or weight loss. Physical examination revealed right upper quadrant pain with deep palpation, but no mass was palpable. Her medical history accounted hypertension, autoimmune thyroiditis and psoriatic arthritis. No surgical history was reported. A recent esophago-gastro-duodenoscopy did not reveal any lesion in the stomach and duodenum. A CT scan of the abdomen revealed a heterogeneous soft tissue mass (5 x 3.2cm) in close contact with the hepatic caudate lobe, posterior wall of the portal vein and the confluence of the IVC and the Left Renal Vein (LRV), without clear cleavage plans with all of them (Figure 1A). The diameter of the IVC and the LRV was significantly reduced, but no intraluminal thrombus was detected (Figure 1B). The radiological origin of the lesion was not detectable. No ascetic fluid was present. Routine laboratory studies revealed normal liver function test results (Alanine aminotransferase [ALT] 13 U/l; Aspartate aminotransferase [AST] 23 U/l; Albumin 4 g/dl; Alkaline Phosphatase [ALP] 69 U/l) and hem coagulation parameters within normal limits. Her white blood cells count was normal and her serum tumor markers (alpha-fetoprotein, Ca 125, Ca 19-9, Ca 72-4, CEA) were within normal range. Her chromogranin A and gastrin serum levels were both elevated (120.3 ng/ml and 3786 pg/ml, respectively). A neuroendocrine tumor of the caudate lobe of the liver infiltrating the confluence of IVC and LRV was firstly suspected, but the PET-CT scan with somatostatin analogue was negative.

Exploratory laparotomy was carried on having the more suitable setting for any type of surgical treatment that might be necessary. This surgical procedure revealed a solid mass arising from the confluence of the IVC and LRV (Figure 1C). No infiltration of the posterior wall of the portal vein, the hepatic caudate lobe and the second portion of the duodenum was detected. A tangential resection of ¾ of the IVC circumference and ligation of the LRV was performed. A patch angioplasty of the IVC was then completed with a PTFE patch fixed with non-absorbable 5/0 stitches (Figure 1D). On macroscopic evaluation, the mass measured 5×3×3.5 cm and infiltrated the IVC wall thickness. The neoplasm was composed of spindle cells organized in irregular interwoven fasciculi. Nuclear pleomorphism and anaplasia was detected. Immunohistochemistry revealed positive reactivity for smooth muscle markers (desmin, SMA, HHF35) and a high (>20%) proliferative index (Ki 67). The histological and immunohistochemically aspects were consistent with the diagnosis of IVC leiomyosarcoma. This led to a poor indication for adjuvant chemotherapy treatment [10]. Furthermore, patient refused this

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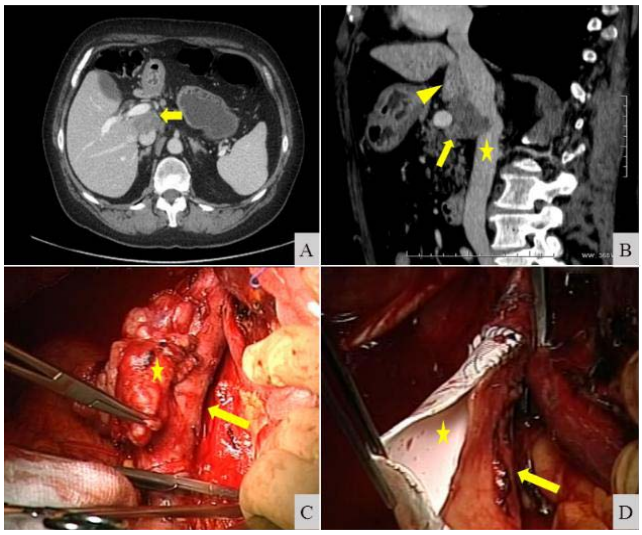

Figure 1: Clinical case #1: **A)** CT scan of soft tissue mass (arrow) of undetectable origin, adhering/infiltrating the caudate lobe of the liver, the inferior vena cava and the posterior wall of the portal vein; **B)** CT scan of soft tissue mass (arrow) between the inferior vena cava (star) and caudate lobe (arrow head); **C)** Intraoperative view of leiomyosarcoma (star) of inferior vena cava (arrow); **D)** Intraoperative view of inferior vena cava (arrow) reconstruction with a PTFE vascular prosthetic patch (star).

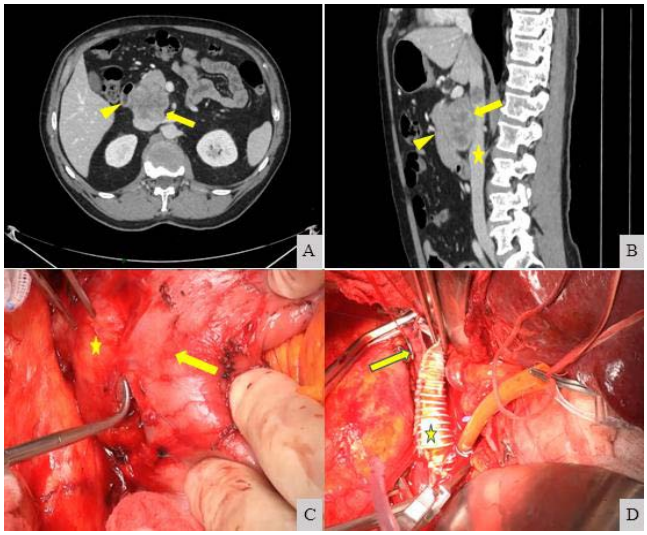

Figure 2: Clinical case #2: **A)** CT scan of soft tissue mass (arrow) near duodenal right wall (arrow head); **B)** CT scan of soft tissue mass (arrow) in contact with duodenum (arrow head) and inferior vena cava (star); **C)** Intraoperative view of leiomyosarcoma (arrow) adhering duodenal right wall (arrow head); **D)** Intraoperative view of inferior vena cava (arrow head) reconstruction with a PTFE vascular prosthetic patch (star).

treatment in relation to a reduced prognosis envisaged by the immunohistochemically pattern [11]. The postoperative course was uneventful, and 10 months after surgery the patient was alive and doing well, without signs of local or distant recurrence, and with complete resolution of the bilateral leg swelling.

Case #2

A 65-years-old man with Crohn Disease undergone regular radiologic follow up, performed an abdominal MRI scan that revealed a solid tissue mass (5x5.7cm) between duodenum and pancreatic head, with a close contact with the IVC determining intra-caval thrombosis. These finding was indicative of duodenal GIST. He referred an increase of articular pain associated with asthenia. At a clinical observation it was not observed abdominal soreness or palpable mass. Routine laboratory test indicated normal hepatic parameters (ALT 13 UI/l; Albumin 3.9 g/dl; ALP 69 UI/l) and normal white cells count.

The CT scan confirmed the duodenal origin of the neo formation (Figure 2A) associated to an infiltration of IVC with neoplastic thrombosis (Figure 2B). No ascetic fluid was present.

With a subsequent echo endoscopy, the mass seemed to come from the duodenal wall with a completely esophitic development, determining a large contact with IVC. During this procedure, a trans-gastric ago biopsy was carried out. Only the following histological examination made a diagnosis of leiomyosarcoma with the following immunophenotype: desmin +; smooth muscle actin +; caldesmon +; DOG1 -; CD117 -; MDM2 -. In consideration of histological finding, the patient underwent a surgical procedure to remove leiomyosarcoma. A first exploration of the retroperitoneal cavity demonstrated the origin of the mass from the anterior wall of the IVC, adhering duodenal lateral wall (Figure 2C). A venous extracorporeal circulation was carried out connecting the right iliac vein and the left subclavian vein. After a palpatory examination to recognize the intravascular limits of neoplasm, the IVC was clamped between renal veins and suprahepatic veins to isolate the affected portion of vessel. Subsequently, a resection of the anterior wall of IVC en bloc with retroperitoneal mass and neoplastic thrombus was performed. The vein was reconstructed with a PTFE vascular prosthesis patch fixed with non-adsorbable 5/0 stitches (Figure 2D).

On microscopic evaluation, neoplasm was composed of spindle cells with necrosis in about 50% of total mass and with 19 mithosis per field, compatible with Grade 2 Leiomyosarcoma. In the postoperative phase, the patient did not present complications. Subsequently adjuvant radiotherapy and chemotherapy (with anthracyclines and dacarbazine) were performed. At a follow-up of 12 months after the surgery, the patient was alive, without any signs of recurrence.

Discussion

Leiomyosarcoma of the IVC is extremely rare, accounting for approximately 0.5% of adult soft tissue sarcomas [12]. Up to now, fewer than 400 cases of IVC leiomyosarcomas have been reported in literature. They usually occur in the sixth decade [13] and the diagnosis is often delayed [14]. Up to 10.5% of the IVCl eiomyosarcomas are detected accidentally, while in 33% of cases they are found at autopsy [13,15].

Tumor involvement is classified into 3 groups according to the level in the IVC segment: segment I, infrarenal; segment II, suprarenal not including the main suprahepatic veins; and segment III, suprahepatic with possible intracardiac extension [15,16]. The retrohepatic segment of the IVC is the most common site of involvement [13,16].

We reported two cases of IVC leiomyosarcomas, whose diagnosis

was made difficult by their position and by the impossibility to detect the origin of the tumor. The rarity of leiomyosarcomas and their tendency to infiltrate adjacent structures usually lead to misdiagnosis. Moreover, not all radiologists manage to recognize radiological characteristics of leiomyosarcoma, although some studies outlined its typical features [15]. Therefore, the recognition of leiomyosarcomasis often made intraoperatively or confirmed by histopathological analysis. In our cases, both events occurred, underlining the importance of the surgical approach for both therapeutic and diagnostic purposes [16]. Neither patient was undergone removal of surrounding structures. This does not happen in all the cases described in the literature [17]. IVC leiomyosarcomas arise from the vessel smooth muscle and may have intraluminal or extra luminal growth pattern, involving nearby structures in most cases [18].

Radical surgical resection, as in other retroperitoneal sarcomas [19], offers patients potential for long term survival and is the only chance for long term cure [20,21]. Resection of nearby structures for radical tumor excision is usually required, and complex resections can be performed safely with possibility of cure [22,23]. Laparoscopic excision of this kind of tumor is possible in exophytic tumor, especially in non-compromising IVC lumen tumors [24].

The actuarial 5 and 10-year survival reported is 49.4% and 39.5%, respectively, with a cancer free actuarial survival at 5 and 10 years of 31.4% and 7.4%, respectively [13].

The role of neoadjuvant and adjuvant therapy is still unclear [25]. No evidence of any benefit in reducing recurrences has been reported from radiation therapy and chemotherapy [26]. Kieffer et al. [15] reported radical resection followed by adjuvant chemotherapy as the optimal therapeutic strategy for leiomyosarcomas of the IVC without metastasis at the time of initial diagnosis. Surgery, alone or in combination with chemotherapy and possibly radiation therapy, is generally not curative, but it constitutes the only hope of prolonged survival [27].

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