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Retroperitoneal leiomyosarcoma originating from the inferior vena cava: A case report and literature review

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Abstract. Leiomyosarcoma of inferior vena cava is scarce retroperitoneal tumor with a poor prognosis. It is usually a misdiagnosed condition due to unspecific clinical course. Imaging is a sensitive tool in establishing the accurate preoperative diagnosis and determining the extent of the tumor. However, tumors may mimic other retroperitoneal conditions making the diagnosis challenging. Herein, we encountered a rare case of leiomyosarcoma of the inferior vena cava in a 69-year-old woman for whom the primary diagnosis was different from the pathological one and the tumor appeared in preoperative imaging as a gastrointestinal tumor. The presence intraoperatively of intimate attachments with the inferior vena cava wall raised the suspicion of vascular origin. The tumor was removed in bloc with a segment of inferior vena cava. Pathological findings led to a further diagnosis of the tumor as a leiomyosarcoma originating from the inferior vena cava. Complete surgical resection is the cornerstone of treatment. The prognosis depends directly on free tumor margins and early diagnosis. Knowing this rare entity is mandatory to assess the correct diagnosis, achieve adequate therapeutic management and improve patient's outcome.

Key words: leiomyosarcoma, inferior vena cava, retroperitoneal neoplasms, vascular neoplasms, case reports, diagnostic imaging, surgical oncology

Introduction

Leiomyosarcoma of the inferior vena cava (LMSIVC) is a rare mesenchymal tumor originating from the smooth muscle cells of the tunica media of the inferior vena cava (IVC) wall (1). It represents 2% of all leiomyosarcoma (LMS) and 0.7 % of retroperitoneal tumors (2). Only a few hundred cases have been reported in the literature, since Perl's first report in 1871 (3). Due to the no-specific clinical course, the diagnosis is mostly delayed (4). Imaging plays a crucial role in establishing the preoperative diagnosis and assessing its extension (3). Despite the advancement of imaging techniques, tumors with extensive extravascular growth patterns may overlap with other retroperitoneal tumors or abdominal diseases, which leads to a diagnostic dilemma (5). We present a rare case of LMSIVC, in a 69-year-old woman, for whom the initial suspected diagnosis was different from the pathological one, and the tumor appeared on preoperative imaging as a gastrointestinal stromal tumor. We aim to highlight the challenges of assessing this rare entity and we discuss the clinico-pathological features as well as the prognosis and potential therapeutic approaches.

Case presentation

A 69-year-old woman, presented with vague and paroxysmal abdominal pain for 4 months.

Physical examination revealed a large and fixed mass in the right hypochondrium. Computed tomography (CT) showed a right lobulated retroperitoneal mass measuring 25x17x11 cm. The enhancement aspect was strong and heterogeneous. Areas of necrosis were noted, without calcifications or fatty components. The tumor caused a mass effect on the duodenum, the pancreas, the right kidney, the vena cava and the renal vein (Figure 1A, B). A gastrointestinal stromal tumor was suggested according to the opinion of the radiologist team, and a CT-guided biopsy was performed. Histological examination revealed dense tumor proliferation arranged in intersecting fascicles of spindle cells (Figure 1C). These cells exhibit elongated and blunt-ended nuclei showing moderate to pronounced nuclear pleomorphism and a moderate amount of pale eosinophilic cytoplasm. Mitotic activity was high with the presence of atypical mitosis (Figure 1D). The stroma was highly vascularized. A focal palisaded arrangement was noted. Fibrosis and myxoid zones were seen. On immunohistochemical study, tumor cells were strongly positive for SMA (Figure 1E) and h-caldesmone (Figure 1F) and negative for CD34, MDM2, Dog1 and S-100. These findings were consistent with the diagnosis of LMS. A laparotomy was performed, and the tumor was closely adherent to the IVC, which was difficult to dissect from the vascular wall. This suggested a vascular origin of the tumor. The tumor was removed in bloc with a segment of IVC and the right kidney. On gross pathological examination, the tumor was well circumscribed with a tan to gray cut surface, measuring 25 cm in the great diameter.

It displayed necrosis and hemorrhage changes. The tumor infiltrated the IVC wall and perirenal fat (Figure 2A). Histological examination of the surgical specimen confirmed biopsy findings and showed a highly cellular proliferation of malignant smooth muscle cells, disposed in interlacing bundles. The mitotic rate was of 21/10 high power fields and the necrosis occupied less than 50% of the tumor volume. The tumor emerged directly from the tunica media of IVC.

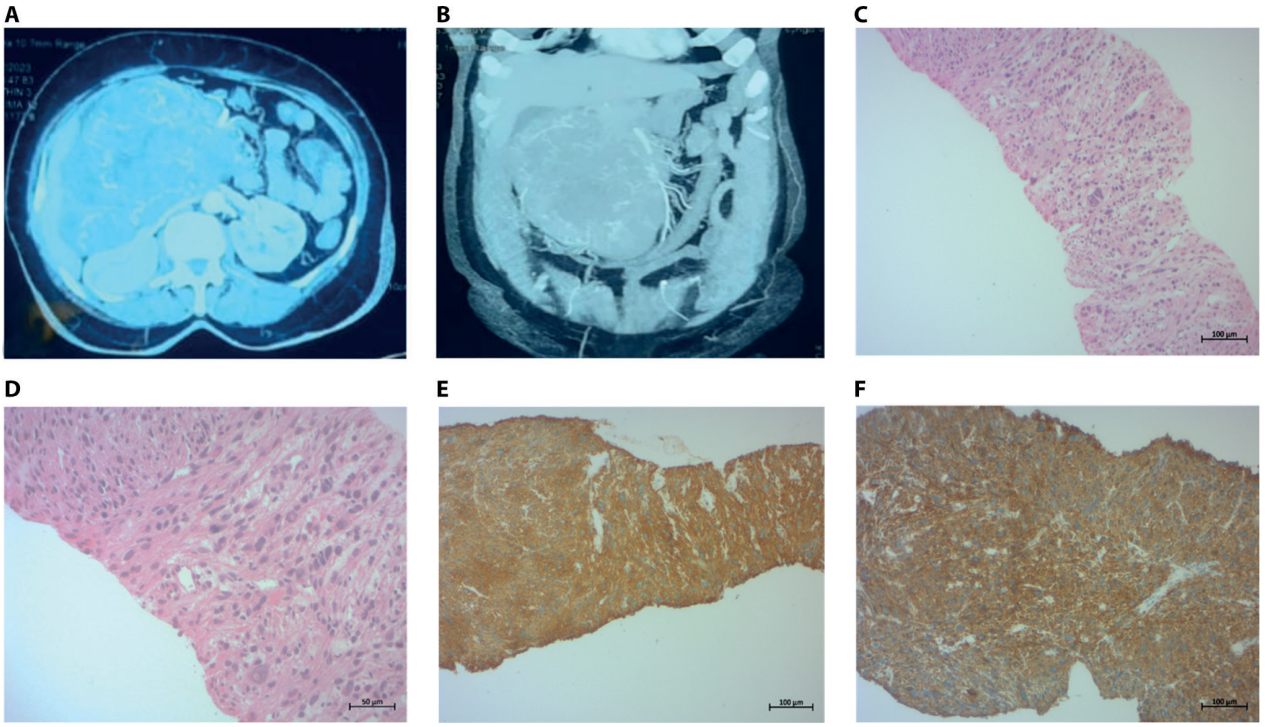

Figure 1. - Axial (A) and sagittal (B) computed tomography images show large heterogeneous retroperitoneal mass compressing the right kidney, the duodenum, the renal vein and the vena cava. (C) Biopsy specimen showing malignant tumor proliferation of spindle cells arranged in intersecting fascicles (HEx25) (D) Spindle tumor cells exhibiting eosinophilic cytoplasm and plump nuclei with nuclear pleomorphism (HEx200) (E) Tumor cells stain positive for SMA (x100) (F) Tumor cells stain positive for h-caldesmone (x100)

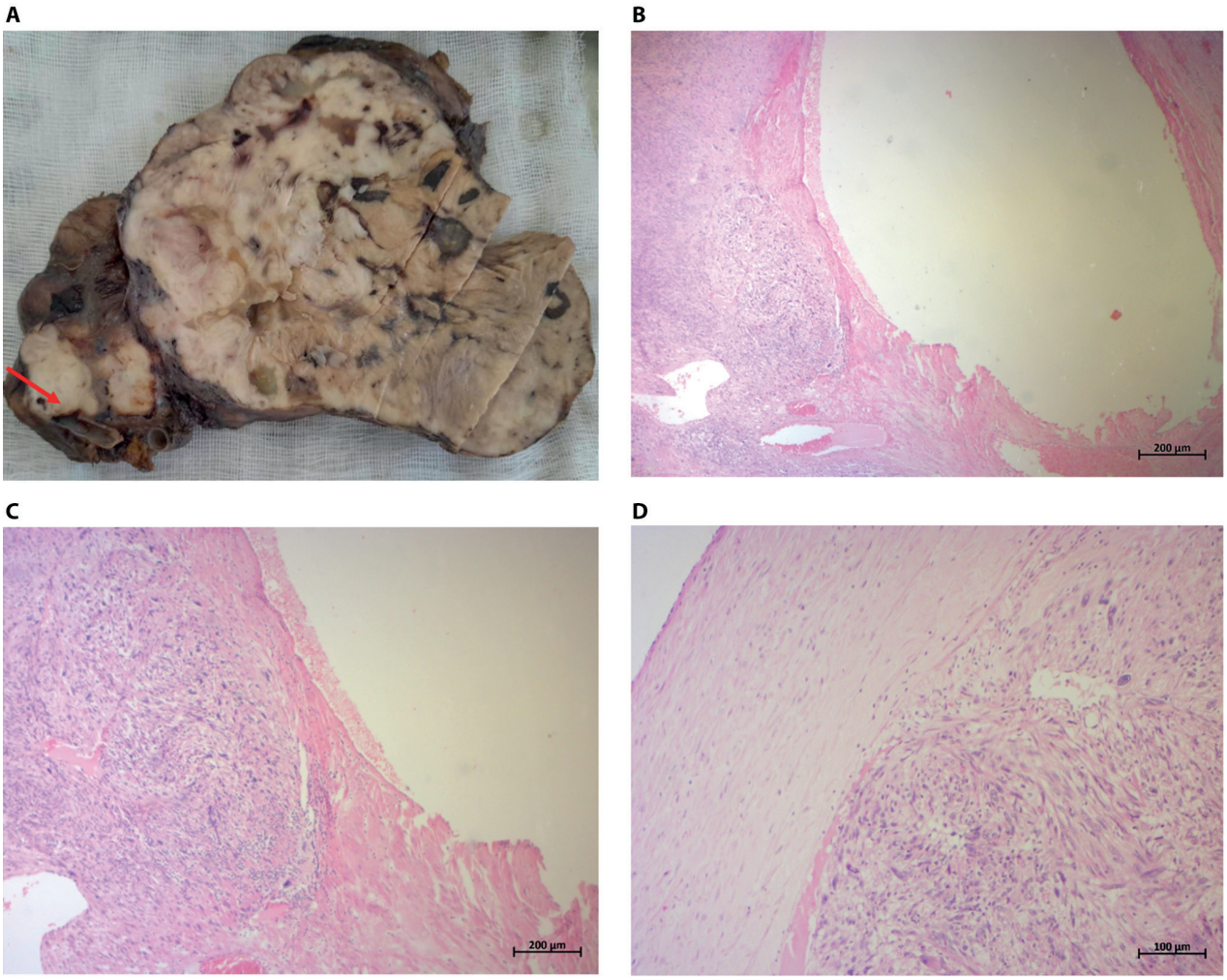

Figure 2. - (A) Gross photograph revealed lobulated, tan to gray tumor with foci of necrosis and hemorrhage; Note the invasion of the inferior vena cava segment (red arrow) Tumour arising from the media of the vena cava wall (B, HEx25) (C, HEx50) (D) High power view showing malignant spindle cells proliferation compatible with biopsy features (HEx100)

Macroscopic and microscopic findings supported the diagnosis of LMS originating in IVC (Figure 2 B, C, D). The tumor grade was 2 according to the Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC). The tumor invaded the perirenal fat and the specimen of the right kidney was free of tumor. The tumor margins were positive. The patient was referred for adjuvant treatment.

Discussion

Vascular LMS is a rare mesenchymal tumor, accounting for 1% of all soft tissue sarcoma (6,7). The IVC is the most affected vessel among vascular LMS representing 60% of cases, followed by renal veins (8). LMSIVC is a rare malignant condition with scarce reports. It can grow in extraluminal, intraluminal or mixed patterns. Our case is consistent with the extraluminal type, which is the most reported one (62% of cases) (1). LMSIVC affects more commonly women in the fifth or sixth decade (9). It is a slow-growing retroperitoneal tumor that causes late symptoms. Similar to our case, the clinical features are usually no specific due to compression of adjacent organs, which makes the diagnosis challenging and established at advanced stages (6). Symptoms vary depending on the affected segment along the IVC and can be classified into three

types. The first type is when the tumor develops below the renal veins (36% of cases) and manifests as an abdominal mass or an edema of the lower limbs and deep venous thrombosis. The second type is when the middle-segment is involved and presents as nephrotic syndrome (44% of cases). Finally, the third type is the rarest (20% of cases) and includes cases that affect the emergence of the hepatic vein, which usually leads to Budd-Chiari syndrome (4,6). In our case, the tumor matched the first type and patient presented with a mass in the right hypochondrium. Radiologic imaging plays an important role in determining the preoperative diagnosis as well as the extent of involvement and the possibility of resection (6). CT and magnetic resonance imaging are two strong tools to delineate the emergence from the IVC and the relationship with retroperitoneal structures (5). This tumor is characterized by a solid lobulated appearance with a mean size of 10 cm. It usually displays heterogeneous enhancement owing to necrosis and hemorrhage changes. Calcifications are rare (8,9). However, tumors with large extravascular development are difficult to be distinguish from tumors that invade the IVC, which makes a challenging diagnosis. Renal, hepatic and adrenal tumors are the most frequent mimics (5). Gastrointestinal stromal tumors, intimal sarcoma and other primary retroperitoneal tumors such as liposarcoma, nerve sheath tumors and undifferentiated pleomorphic sarcoma may also overlap with LMSIVC (1,2,5). Thereby, CT-guided biopsy and pathological examination are required to achieve the final diagnosis (8). As reported in our case, the mass was initially suspected as a gastrointestinal tumor in preoperative imaging, and the definitive diagnosis of LMSIVC was established by pathological analysis. Macroscopically, LMS appears as a fleshy mass of gray to white to tan color. Larger tumors often exhibit hemorrhage, necrosis or cystic areas. The tumor margin usually appears well circumscribed; however infiltrative tumors are also seen (8,9). The histological appearance is characterized by spindle-shaped cells, set in intersecting fascicles and exhibiting plump nuclei and pale to brightly eosinophilic cytoplasm. Moderate nuclear pleomorphism with frequent mitosis including atypical ones are usual (6). A storiform or palisaded arrangement can be seen. The tumor is highly cellular, although fibrosis and myxoid change may occur. Epithelioid morphology is extremely rare. Coagulative necrosis is present in larger masses (8). Some tumors are poorly differentiated and show anaplastic and pleomorphic features, which can be considered as pleomorphic dedifferentiated LMS. Tumor grade is usually high, similar to other soft tissue LMS (9). By immunohistochemistry, this tumor stains positive for myogenic markers (SMA, desmin, and h-Caldesmon) in 70% of cases. None of these markers is specific to smooth muscle cells, and the positivity for two myogenic markers is supportive. The myogenic stain is usually weaker in pleomorphic areas. Additionally, other markers such as Keratin, EMA and CD34 may be focally positive (6,10). Complete surgical resection remains the gold standard treatment of the LMSIVC (6,7). The role of adjuvant therapy is still controversial, given the resistance of this tumor to both chemotherapy and radiotherapy. These treatments may be useful in metastatic or recurrent disease, high-grade tumors and positive surgical margins (2,10). LMSIVC has a poor prognosis with a survival rate around 30% to 50% at 5 years, even after radical surgical removal (10). Local recurrence is a frequent finding during follow-up (57% of cases) and distant metastases are common, involving mainly the liver, lungs, brain and peritoneum (8,10). The prognosis depends directly on early diagnosis, size of the tumor at diagnosis and negative margins (4). Thus, we should consider the LMSIVC in front of no specific abdominal symptom in a middle-aged woman to assess an appropriate diagnosis at early stage and properly plan the therapeutic modalities.

Conclusion

LMSIVC is a rare and aggressive tumor. The diagnosis remains challenging and difficult to set in the preoperative course, due to the absence of a specific clinical symptom. Although imaging is mandatory to reveal an accurate diagnosis and determine the extent of disease, tumors may mimic other retroperitoneal tumors and thereby pathological analysis is required. Knowing this rare condition is crucial to set an early diagnosis allowing sufficient surgical management and higher survival rate.

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