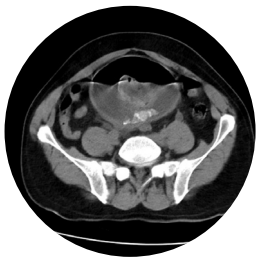

Leiomyosarcoma of the inferior vena cava. What to look for in the images? Presentation of a case

Leiomiosarcoma de la vena cava inferior. ¿Qué buscar en las imágenes? Revisión a propósito de un caso

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Summary

Inferior vena cava leiomyosarcoma (IVC) is a low-growing, malignant mesenchymal tumor that arises in the smooth muscle cells of the tunica media of the vascular wall and has a generally poor prognosis. Images play a crucial role in the diagnostic approach and in surgical planning, therefore, recognizing the most frequent findings is necessary in radiological practice. We describe a case that outlines the most typical findings in different diagnostic modalities including tomography, magnetic resonance imaging and PET-CT, with the aim of recognizing the characteristics that can lead to an earlier diagnosis and therefore to favor patient survival.

Resumen

El leiomiosarcoma de la vena cava inferior (VCI) es un tumor mesenquimal maligno, poco frecuente, de bajo crecimiento, que surge en las células de músculo liso de la túnica media de la pared vascular y que tiene en general un mal pronóstico. Las imágenes juegan un papel crucial en el abordaje diagnóstico y en el planeamiento quirúrgico, por lo que reconocer los hallazgos más frecuentes es necesario en la práctica radiológica. A continuación, se describe un caso que ilustra los aspectos más representativos de esta enfermedad en diferentes modalidades diagnósticas, incluyendo tomografía, resonancia magnética y PET-CT, con el objetivo de hacer un reconocimiento de las características que puedan llevar a un diagnóstico temprano y, por lo tanto, a favorecer la sobrevida de los pacientes.

Presentation of the case

A 38-year-old female patient, with no significant pathological history. She presented with a clinical picture of one month of evolution consisting of edema of the right lower limb, dyspnea at rest, emesis and increased abdominal perimeter. A venous Doppler ultrasound of the lower limbs revealed thrombosis of the common femoral vein and the right greater saphenous vein. Angiotomography ruled out associated pulmonary thromboembolism and extension studies were initiated to look for possible causes of the thrombotic event.

The thoracoabdominal tomography (CT) with intravenous contrast medium showed occupation and dilatation of the inferior vena cava (IVC) due to an expansive lesion, of heterogeneous density and without calcifications (Figure 1a). Images with contrast medium in portal phase showed peripheral enhancement with some foci of central enhancement (Figure 1b), extending from the confluence of the renal veins to the intrathoracic portion (Figure 1c), with a maximum diameter of 6 cm and length of 15 cm.

For better characterization of the lesion, magnetic resonance imaging (MRI) was performed, which confirmed the presence and extension of the mass in the IVC. In the T2-weighted sequences the mass had an intermediate signal with some high signal foci (Figure 2). In the T1-weighted images with precontrast fat suppression the signal was low homogeneous and with contrast medium heterogeneous enhancement was observed, without extraluminal infiltration (figure 3). Diffusion showed some high signal foci with a corresponding drop in signal intensity on the apparent diffusion coefficient (ADC) map, consistent with areas of diffusion restriction (Figure 4). There was no evidence of metastatic disease, invasion of adjacent structures or prominent venous collateral circulation.

PET-CT demonstrated an intravascular solid lesion in the intrahepatic IVC with heterogeneously increased metabolism, with an SUVmax of 5.17; it did not exceed the vessel contours and was also associated with increased metabolism in the vascular walls. No metastases were identified (Figure 5).

Given the location of the mass and its imaging characteristics, a diagnosis of IVC leiomyosarcoma

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was made. The patient underwent endovascular biopsy of the lesion, which revealed a smooth muscle tumor, and by histological and immunohistochemical analysis a definitive diagnosis of low-grade IVC leiomyosarcoma was made (Figure 6).

With this result, the multidisciplinary team decided to perform a complete surgical resection and reconstruction of the IVC with a cadaveric donor aorta graft (Figure 7). However, due to postoperative complications, the patient died on the third day after surgery.

Discussion

Vascular leiomyosarcomas account for 1-2 % of all leiomyosarcomas (1). They are rare, low-growing, low-growing, malignant mesenchymal tumors arising in the smooth muscle cells of the tunica media of the vascular wall (2).

They involve five times more veins than arteries and in total make up one in every 100,000 malignant tumors (3). More than 50 % occur in the inferior vena cava (IVC), which makes them the most common primary tumor of this structure (4). The other locations that follow in order are the thoracic aorta and the pulmonary artery (5). The veins most frequently affected after the IVC are the renal, greater saphenous, pulmonary, femoral and popliteal veins (6).

Since the first description by Pearl in 1871 and the first report of surgical resection by Melchior in 1928, about 600 cases have been documented in the literature. This disease commonly presents between the fifth and seventh decades of life, with female predominance and a female:male ratio of 3:1 (3, 7, 8).

There are three patterns of tumor growth: extraluminal (outside the vessel), intraluminal (inside the vessel) and mixed, and of these, extraluminal growth is the most common form (9). In the case reviewed here it was an intraluminal growth. Leiomyosarcomas are also classified according to their location, dividing the IVC into three segments or levels: segment 1 or lower (from the iliac veins to the renal veins, 34 % of cases), segment 2 or middle (from the renal veins to the hepatic veins, 42 % of cases and better prognosis) and segment 3 or upper (from the hepatic veins to the right atrium, 24 % of cases) (2, 3). In 10-17 % of patients, the entire length of the vena cava is affected (4).

Symptoms are related to the segment of the IVC affected and vary according to the dimensions and growth pattern. Abdominal pain in the right lower quadrant, back or flank is associated with segment 1. Lower extremity edema, pain in the right upper quadrant of the abdomen, renovascular hypertension or renal vein thrombosis are associated with segment 2, and multiple clinical manifestations of Budd-Chiari syndrome are related to segment 3 (10). Tumor extension along the IVC into the right atrium may cause cardiac symptoms (2, 11).

Images play a fundamental role in the characterization of the tumor and its diagnosis, serve as a guide for biopsy, help in surgical planning, facilitate the detection of metastasis and follow-up to evaluate tumor relapse. IVC leiomyosarcoma is a tumor that can present without symptoms and its identification in the images can be an incidental finding, so it is essential to be familiar with its characteristics, given that long-term survival depends on early diagnosis and the performance of extensive surgery (12).

Several studies have shown that CT can provide sufficient information to make an accurate analysis and determine the extent of the tumor (13). It is a sensitive tool for diagnosis and follow-up. IVC leiomyosarcomas are usually large tumors, larger than 10 cm, lobulated, heterogeneous due to hemorrhage and necrosis, tend to be hypovascular and may present peripheral relapse (14), as in the case of this patient. Tumor attenuation depends on the degree of tumor necrosis. Areas of high attenuation are uncommon, but can be seen in areas of recent hemorrhage (12). The typical appearance includes dilatation of the vein and an endoluminal mass isodense to the liver with irregular enhancement and total or almost complete venous obstruction (4).

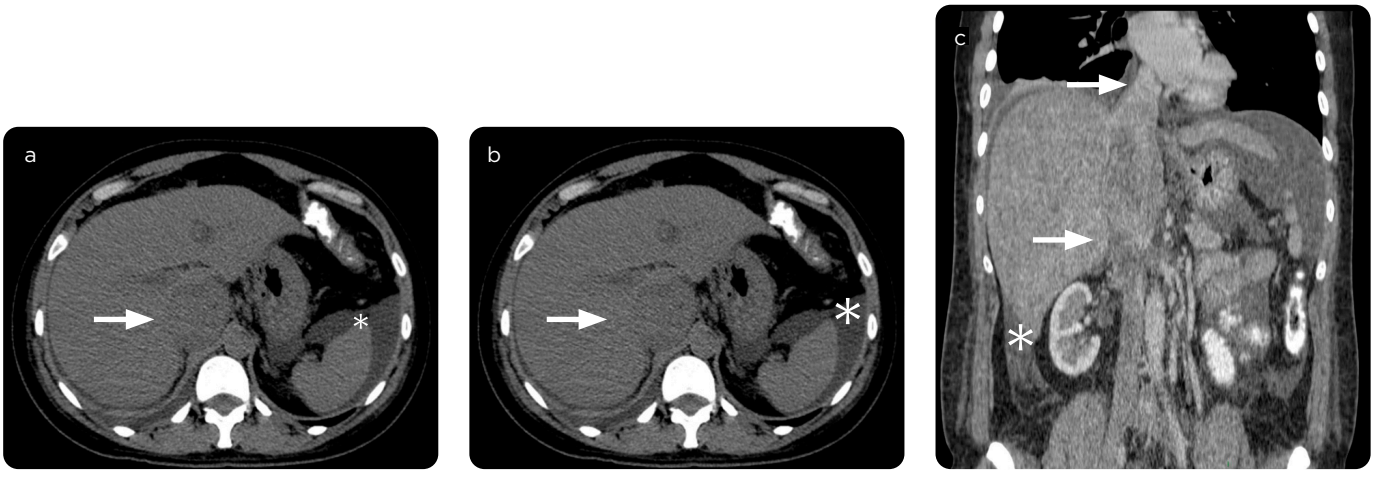

Figure 1. a) Simple axial CT: lesion in the IVC in its intrahepatic portion, without calcifications, slightly heterogeneous (arrow). Ascites is also observed (asterisk). b) Axial CT with contrast medium in portal phase: dilatation of the IVC in the intrahepatic portion. There is complete loss of the lumen, which is occupied by a mass that shows a slight enhancement of peripheral predominance (arrow). Ascites (asterisk). c) CT with contrast medium coronal reconstruction: IVC dilated by intraluminal tumor extending from the confluence of the renal veins to the intrathoracic portion, prior to its arrival to the right atrium (arrow). Right pleural effusion and ascites (asterisk).

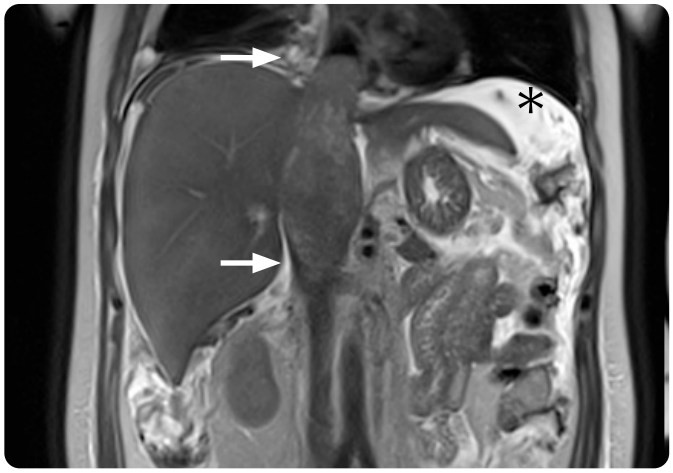

Figure 2. Coronal T2-weighted sequence. IVC with an imperceptible lumen, which is occupied by a mass with extension from the confluence of the renal veins to the intrathoracic portion. Intermediate signal with some areas of high signal intensity (arrows). Ascites (asterisk).

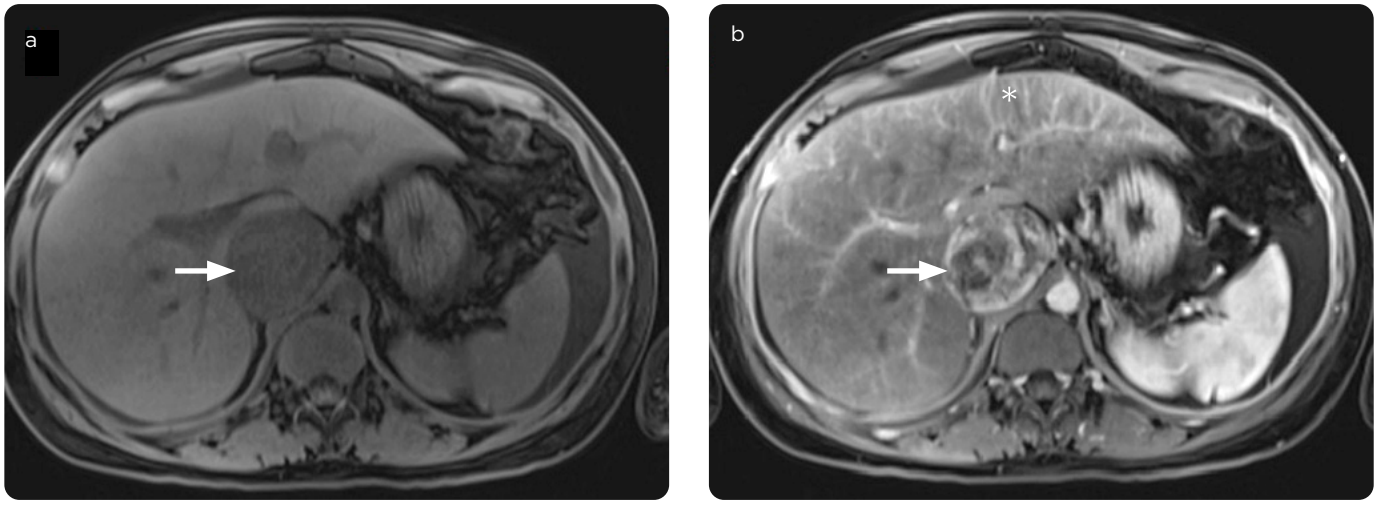

Figure 3. a) Axial T1 fat suppression. Expansive lesion in the IVC in the intrahepatic portion, predominantly hypointense (arrow). b) Axial T1 fat suppression postcontrast in portal phase: heterogeneous enhancement of the mass is identified, especially in its peripheral portion (arrow). The lumen of the IVC is imperceptible. There is hepatic perfusion disorder secondary to decreased venous flow (asterisk).

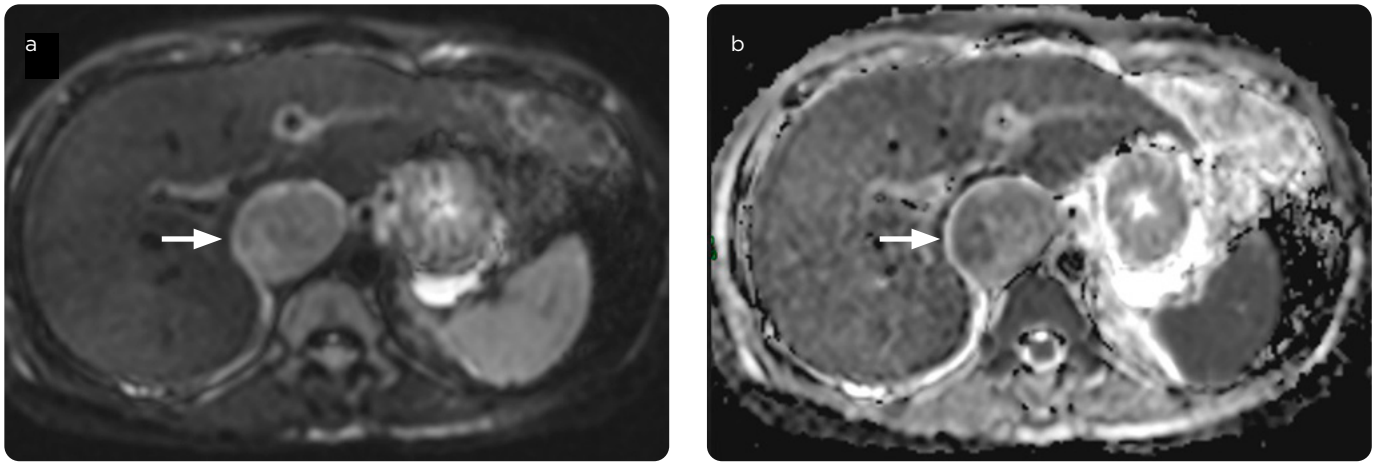

Figure 4. Diffusion sequence and ADC map. Expansive lesion is again identified in the IVC in the intrahepatic portion (arrow) with some high-signal foci in the diffusion that present a drop in signal intensity in the ADC map in relation to restriction zones.

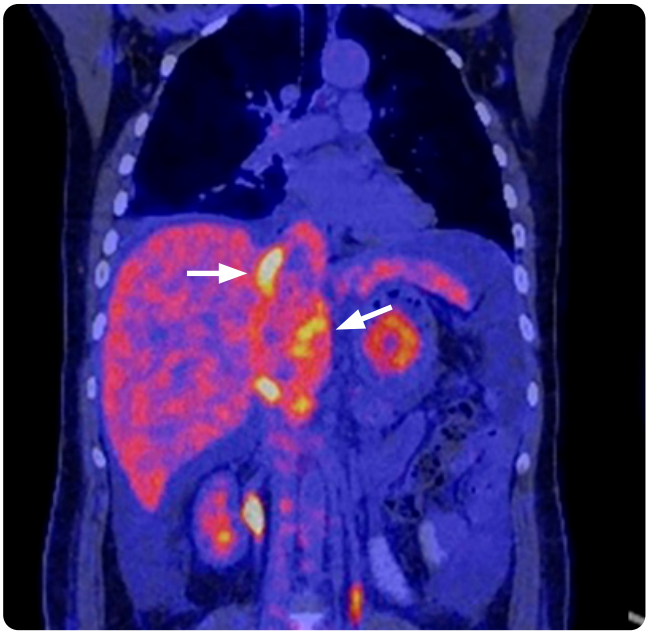

Figure 5. Intravascular solid lesion in the intrahepatic IVC with increased metabolism greater than that of the liver, not exceeding the contours of the vessel (arrows). No metastasis.

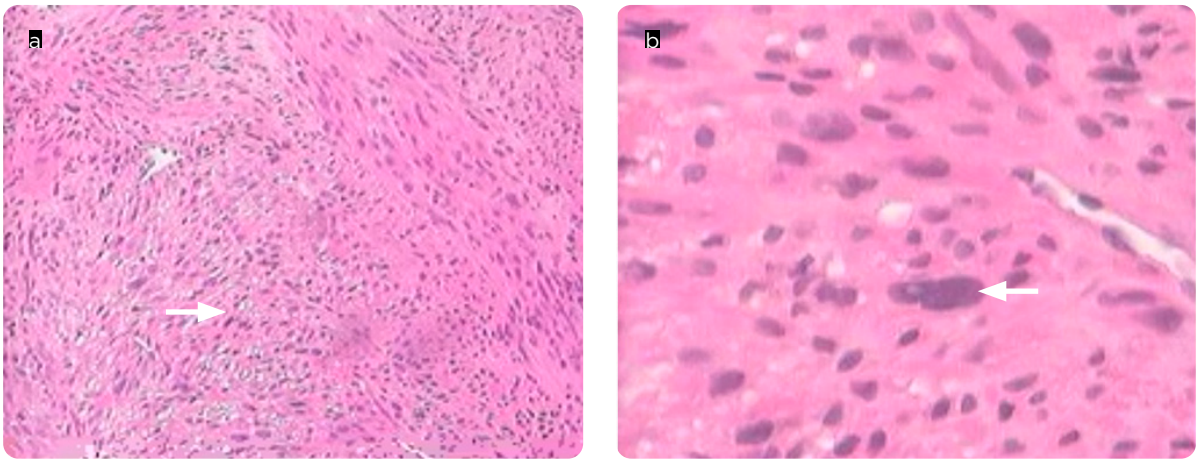

Figure 6. Hematoxylin-eosin staining: proliferation of spindle cells with nuclear atypia and mitosis (arrows), compatible with well-differentiated leiomyosarcoma.

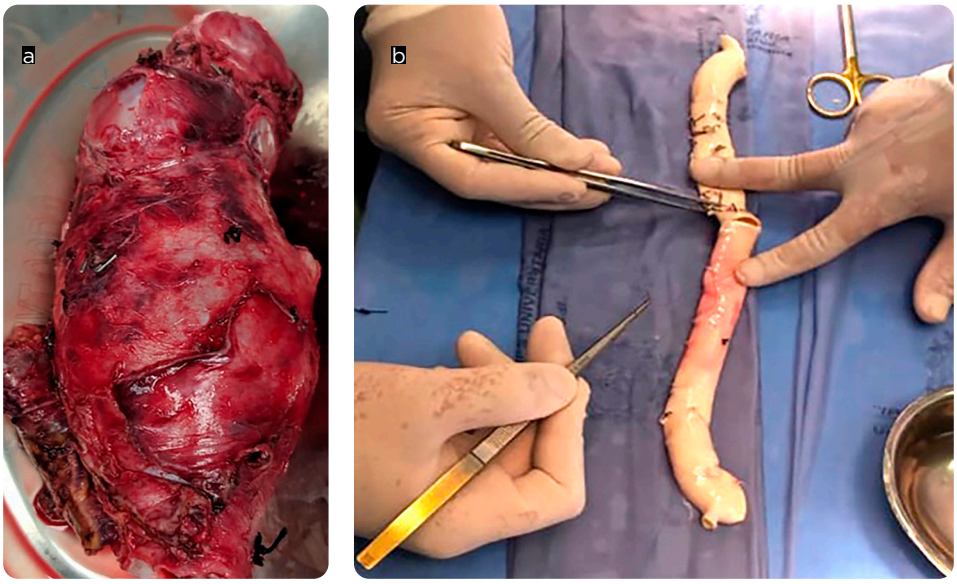

Figure 7. Tumor specimen and implanted graft. a) Completely resected leiomyosarcoma of the IVC. b) Aortic graft from cadaveric donor that was implanted in the patient for reconstruction of the IVC.

Usually, they do not have calcifications and are not associated with regional or distant lymphadenopathy. At the time of presentation, almost half of all patients have metastases in the liver and lungs (12).

On the other hand, magnetic resonance imaging (MRI) accurately shows the extension of the tumor and achieves an earlier detection of its origin and its relationship with adjacent structures. This is due to the fact that it has a better contrast resolution for the evaluation of soft tissues in comparison with CT (15). The signal intensity characteristics vary depending on the degree of necrosis and cystic degeneration within the tumor (2). Generally, in T1-weighted images it has a homogeneous intermediate signal and in T2-weighted sequences a heterogeneous intermediate signal. Intraluminal tumor growth can cause obstruction and dilatation of the IVC with formation of venous collaterals (2, 4, 12).

Although some CT features can help differentiate intraluminal tumor from soft thrombus -such as a filling defect occupying the entire vessel diameter, expansion of the involved vascular structure or extraluminal extension in the case of tumor-, MRI is also very useful for this purpose. Commonly, soft thrombus is isointense to muscle in T2 sequences and does not enhance with contrast medium, while leiomyosarcoma is iso or hyperintense to muscle in the T2 sequence and may present peripheral enhancement with contrast medium. Additionally, because of increased cell density and altered nucleus-to-cytoplasm ratio, tumors may be diffusion restricted with decreased ADC, whereas thrombus is not. In some studies, soft thrombi have been shown to have higher ADC values, which may also aid in differentiation (2, 13, 16). In the patient in this case there were areas of diffusion restriction in the intraluminal tumor.

PET-CT is another tool that allows differentiating with precision the tumor from a soft thrombus, since the tumor will present greater FDG uptake given its increased metabolism (10), a finding that was evidenced in the PET-CT images of the case presented. It also plays an important role in defining the extension of the tumor and the presence of metastasis, factors which in turn will help in the staging and planning of adequate treatment.

Occasionally, IVC leiomyosarcoma with extravascular development can be difficult to differentiate from retroperitoneal tumors that compress or invade the IVC. A useful sign documented by Well et al. is that leiomyosarcomas have an imperceptible cava lumen in approximately 75 % of cases and that this sign is usually not identified in other diagnoses; such a finding is illustrated in Figures 1b, 2, and 3 of the case described here. If the IVC is compressed towards the periphery of the mass adopting a crescent configuration, this finding may suggest that the mass does not originate in the IVC (11).

The differential diagnosis of IVC leiomyosarcoma is that the tumor is narrow when the presentation is intraluminal, given that dilatation and loss of the IVC lumen is almost pathognomonic (11). It is important to differentiate it from a soft thrombus, as discussed in the previous sections. There are other primary cardiac tumors with extension to the IVC that may resemble it, such as angiosarcoma or cardiac lymphoma. Angiosarcoma is the most common primary malignant cardiac tumor, most occur in the right atrium and affect the pericardium, and to a lesser extent involve the retroperitoneum and specifically the IVC, usually manifesting with local lymphadenopathy, which is not observed in leiomyosarcoma and this, in addition to cardiac involvement, can be useful in its differentiation. On the other hand, cardiac lymphoma with extension to the IVC tends to be a homogeneous mass, which is not common in leiomyosarcoma, and cardiac involvement will also be seen in all cases (2).

Another differential diagnosis is leiomyomatosis, a benign condition caused by smooth muscle proliferation of leiomyomas in the uterus, with vermiform extensions that disseminate to the iliac veins and then to the IVC; on imaging leiomyomas can be seen in the uterus and involvement ascending from the iliac veins, unlike what appears in IVC leiomyosarcoma (2).

In the extraluminal type it is a little more difficult to distinguish and is often confused with an adjacent organ tumor that directly invades the IVC. In these cases the organ involved and the characteristics of the tumor should be evaluated; for example, in the case of a retroperitoneal liposarcoma, areas of soft tissue and fat attenuation may be evident (11, 15, 17).

The ideal treatment of IVC leiomyosarcoma consists of en bloc radial resection with or without adjuvant chemotherapy, with the aim of achieving negative margins. The type of surgery is performed according to the segmental location. When they are located in the infrarenal portion and there are abundant collaterals, resection and ligation of the vena cava can be performed; however, when there are few collaterals patients may develop debilitating lower limb edema, so the presence or absence of collaterals may change this type of approach. In these cases and when the tumor is of suprarenal location, complete resection is achieved through the placement of a graft or reconstruction of the IVC (11, 18), as was observed in the case presented.

Surgical approach with curative intent is not always possible. In these cases, the goal of treatment is to prolong survival, control symptoms and prevent progression. For this purpose, a combination of radiotherapy and chemotherapy is performed. Traditional first-line therapy includes gemcitabine and docetaxel (15, 19).

In most patients there is a poor prognosis, with 5 and 10-year survival rates after surgical resection of 31.4 % and 7.4 %, respectively; with a 5-year survival rate close to 0 % when there is incomplete resection, and inoperable cases that only received chemotherapy with or without associated radiotherapy. Even when complete resection is considered to have been achieved, local recurrence is common and is observed in 40-77 % of cases (4, 19, 20). In the case documented here, the patient's death was due to complications related to the surgical procedure. Post-surgical complications include renal failure, especially when there is extension to the renal vein, edema of the lower limbs, graft thrombosis and hemoperitoneum, which can be secondary to dehiscence in the anastomoses (21, 22).

Conclusion

We describe a case of intraluminal IVC leiomyosarcoma that underwent complete resection and reconstruction of the IVC with cadaveric donor aortic graft. This case is very representative and illustrates the main findings in the different diagnostic modalities. Both CT and MRI showed that the increase in the diameter of the IVC and the loss of its lumen are key in the diagnosis. Likewise, the scarce and peripheral enhancement, diffusion restriction and FDG uptake in PET-CT were other tools that facilitated the diagnostic imaging, which, in turn, were very useful for surgical planning. The above shows that the role of the radiologist is fundamental in the approach to this type of patients, so it is essential to be familiar with the most relevant aspects of imaging.

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