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CASE REPORT

Leiomyosarcoma of level II inferior vena cava—an original solution for bilateral renal vein reconstruction

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Abstract
Introduction: The leiomyosarcoma of the inferior vena cava (IVC) is a rare clinical entity, although it represents the most common malignant tumor of the venous system. Level II IVC tumors (supra-renal) are the most frequent and those who have a better prognosis for the development of symptoms earlier.
Case report: The authors report a case of IVC leiomyosarcoma in a 59-year-old woman, presenting with DVT of the right lower limb, subsequent to prolongued nonspecific abdominal pain. Computed tomography revealed a large retroperitoneal neoformation, centered to IVC, which extended above the renal veins (the left one patent and the right one involved in the mass). The patient underwent *en block* resection of the tumor and reconstruction of the renal veins: construction of a new IVC bifurcation at the supra-renal level with a bifurcated PTFE graft, followed by graft extension to both renal veins using externally-supported 8 mm PTFE grafts. Histology revealed a high-grade leiomyosarcoma. The postoperative period was complicated by a type 2 MI and retroperitoneal hematoma, with occlusion of the right graft branch and partial infarction of the right kidney. The patient underwent surgery again and proceeded to partial resection of the thrombosed graft branch. The patient was discharged home under anticoagulation and is clinically well without edema of the lower limbs, normal renal function, and has begun adjunctive therapy.

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Conclusion: The prognosis of these tumors is poor, with a high recurrence rate. An aggressive surgical approach combined with adjuvant therapy may not be curative, but is the best strategy to prolong survival.

PALAVRAS-CHAVE
Veia cava inferior;
Nível II;
Leiomiosarcoma;
Resseção em bloco;
Reconstrução vascular;
Terapêutica adjuvante

Leiomiossarcoma do nível II da Veia Cava Inferior – uma solução original para a reconstrução das veias renais

Resumo
Introdução: O leiomiossarcoma da veia cava inferior (VCI) é uma entidade clinica rara, embora represente o tumor maligno mais comum do sistema venoso. Os tumores do segmento II da VCI (supra-renal) são os mais frequentes e aqueles que apresentam melhor prognóstico pelo desenvolvimento de sintomatologia mais precocemente.
Caso clínico: Os autores apresentam um caso de leiomiossarcoma da VCI numa doente de 59 anos, que se manifestou por TVP do membro inferior direito, subsequente a quadro de dor abdominal inespecífico arrastado. A tomografia computorizada revelou volumosa neoformação retroperitoneal, centrada à VCI, que se estendia acima das veias renais (a esquerda permeável e a direita envolvida na massa). A doente foi submetida a ressecção em bloco do tumor e reconstrução das veias renais: construção de neo-bifurcação da VCI com prótese de PTFE bifurcada seguido de reconstrução de ambas as veias renais com próteses aneladas (PTFE-8). A histologia revelou tratar-se de um leiomiossarcoma de alto grau. O pós-operatório foi complicado por EAM tipo 2 e por hematoma retroperitoneal com oclusão do ramo protésico direito e enfarte parcial do rim direito. Foi reintervencionada, tendo-se procedido à ressecção parcial do ramo protésico trombosado. A doente teve alta anticoagulada e encontra-se clinicamente bem, sem edema dos membros inferiores, com função renal normal, e iniciou terapêutiva adjuvante.
Conclusão: O prognóstico destes tumores é reservado, com elevada taxa de recorrência. Uma estratégia cirúrgica agressiva combinada com terapêutica adjuvante pode não ser curativa, mas constitui a melhor estratégia para prolongar a sobrevida.

Introduction

The IVC leiomyosarcoma is a rare clinical entity, with only 300 cases described in the literature and most of them refer to individual case reports or small series.[1–5] Represents the tumor that most often affects the venous system and is associated with a poor prognosis.[1,3,4]

Most patients are asymptomatic, for it is a slow growth tumor and only manifests symptoms in advanced stages of the disease.[4,6–8] Its incidence is more common in women around the sixth decade of life.[1] Abdominal pain is the most common symptom; edema of the lower limbs, back pain, weight loss, fever, palpable abdominal mass and, rarely, Budd–Chiari syndrome can also be part of the clinical presentation spectrum.[5,9–11]

The treatment of this tumor is still controversial. In this paper, we present a case report of a patient submitted to a complex vascular construction.

Case report

A 59-year-old woman presented to her attending physician with epigastric and lower back right pain, associated with constipation and weight of the lower limbs. The abdominal and renal ultrasound scan was normal. Six months later, she developed pain and asymmetrical edema of the right lower limb, with tenderness of leg muscle mass and positive Homan's sign. She maintained pain in the epigastric and right upper quadrant on deep palpation and no palpable masses were felt. The patient did not have fever, chest pain, gastrointestinal, urinary or respiratory changes, as well as recent weight loss. She was admitted to a local hospital with the diagnosis of iliofemoral deep vein thrombosis of the right lower limb.

Her past medical and surgical history included essential hypertension, past caesarean section and laparoscopic cholecystectomy; no known risk factors for venous thrombosis or relevant family history were present.

The venous Color-flow Duplex Scan revealed thrombus extension to the juxta-renal IVC and a nodular mass was observed at this level. An abdominal and pelvic CT scan was performed, revealing the presence of a solid heterogeneous mass in the topography of the duodenal arch, without clear cleavage plane with the duodenum, the uncinate process of the pancreas and the IVC. The examination was completed by ecoendoscopy, which was suggestive of GIST, and ultrasound-guided biopsy of the lesion, which was