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Leiomyosarcoma of renal vein: A case report

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ABSTRACT

Leiomyosarcoma of the renal vein is a rare tumor of complex diagnosis. We present a case of renal vein leiomyosarcoma in a patient presented with left flank pain. The patient underwent radical nephrectomy with tumour excision.

Introduction

There are approximately 30 cases of leiomyosarcoma of the renal vein reported in literature. They affect primarily the inferior vena cava in more than 50% of the cases [1]. We present a case of primary leiomyosarcoma of the left renal vein.

Case report

65 year oldfemale, came to outpatient department with complaints of left sided flank pain of more than 1 year duration. Physical examination was normal. An abdominal ultrasound showed a tumor near the left renal hilum in close contact with the renal vein. Contrast enhanced CT showed a well lobulated iso-dense heterogeneously enhancing lesion of 4.9 × 3 cm anterior to left renal vein. Lesion was not separately visualised from renal vein (Fig. 1). She underwent Left radical nephrectomy with excision of the mass en bloc (Fig. 2). Histopathology report showed a neoplasm of 5 × 3.8 × 3.2 cm arising from left renal vein. Section showed neoplasm arising from tunica media of renal vein. Cells are spindly with moderate eosinophilic cytoplasm with elongated blunt ending vesicular nuclei showing nuclear pleomorphism in areas. Some cells show multinucleation. Mitotic figures were 5–6/10 hpf (Fig. 3). Renal vein cut margin is free of neoplasm. Kidney renal artery and ureter were free. One hilar lymph node was present which showed follicular hyperplasia only. Metastatic workup done was negative. Immunohistochemistry showed tumor cells getting stained positive with Desmin, Vimentin, H-Caldesmon and SMA (Smooth Muscle Actin) (Fig. 3).

Discussion

Leiomyosarcoma of the inferior vena cava was first described in 1871, and its diagnosis and treatment is still challenging.[1] Leiomyosarcoma is an uncommon soft tissue tumor, generally occurring in myometrium, respiratory tract and retroperitoneal organs. It seldom originates in vascular structures, and the inferior vena cava accounts for more than 50% of the such cases.[2] These tumors are seen in women over 30 years in 85% of the cases.[2] More frequently they are left-sided (64%). Symptoms are nonspecific, such as mild lumbar and abdominal pain, and wasting. Hematuria and palpable mass are rare.[2]

Until the 80's, approximately 50% of the cases were autopsy findings. Presently they are incidentally found in routine studies.[2] Clinical context and ultrasound and computed tomography studies are nonspecific and do not allow an adequate differential diagnosis with other retroperitoneal solid tumors.[3]

Leiomyosarcoma spread occurs primarily by local extension. Through hematogenous spread it affects liver (25%), lungs (63%), bones (19%) and, less frequently, lymph nodes.[3] At diagnosis, approximately half of the cases present metastatic disease or are nonresectable due to local invasion, presenting therefore a poor prognosis.[3]

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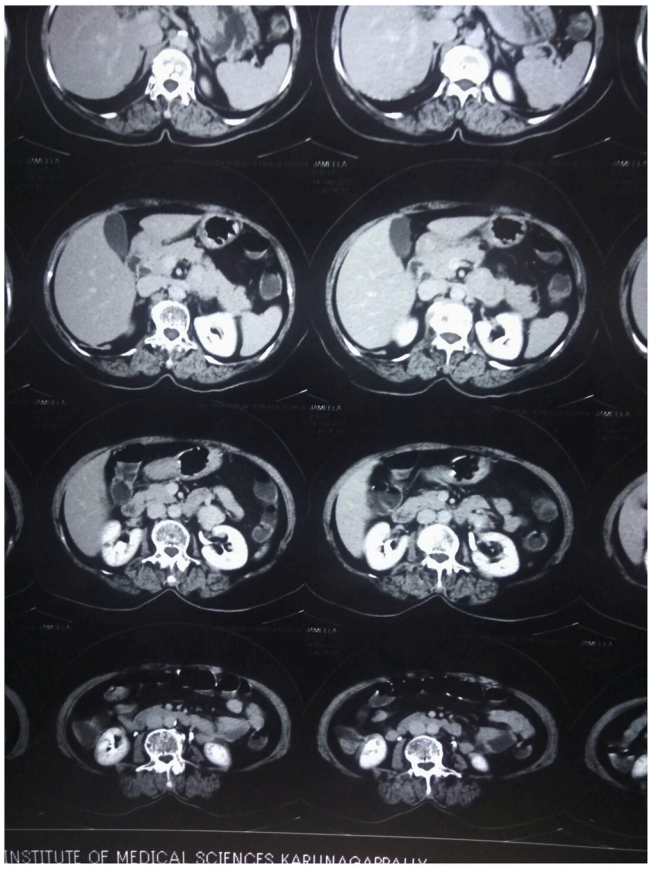

Fig. 1. CT picture showing the tumour close to the left renal hilum attached to the left renal vein.

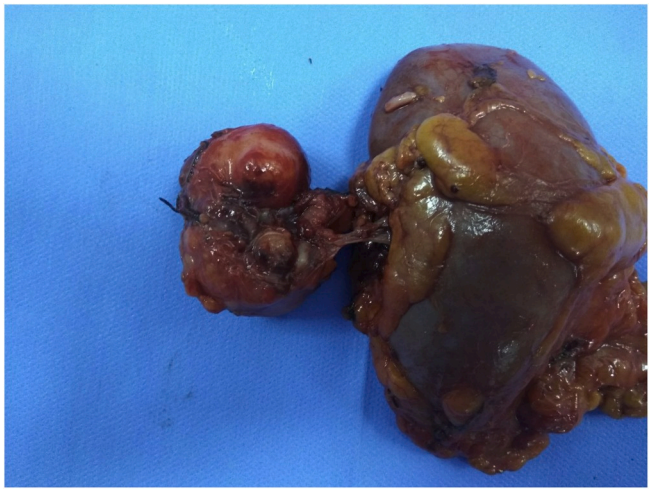

Fig. 2. Specimen showing the left kidney along with the mass arising from the renal vein.

The best treatment for leiomyosarcoma is surgery with total tumor removal. This is the option that offers the best chances of local control and 5 years survival. Studies performed at Memorial Sloan Kettering, New York, showed that the major prognostic factor is total surgical resection. When it is complete, 5 years survival free of disease is of approximately 60%, v/s. just 30–35% when it is partial. Once total removal is performed, major prognostic factor becomes histological grade, with 5 years free of disease survival of 90–95% for low grade tumors, and of 30–35% for high grade tumors.[4]

Radiotherapy and adjuvant chemotherapy have limited effects due to toxicity on contiguous structures. Adjuvant therapy is generally used to high grade tumors, with partial resection.[3]

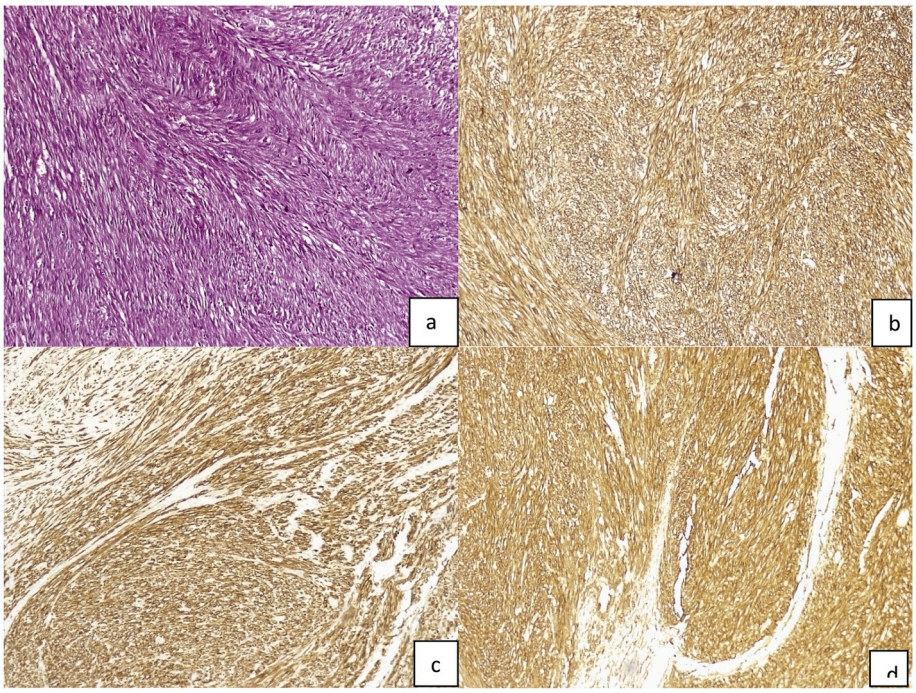

Fig. 3. a) Haematoxylin and eosin staining of the tumour and immunohistochemistry showing strong positivity with b) Desmin, c) H- Caldesmon and d) Smooth Muscle Actin (SMA).

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