Case Report

A rare case of extraperitoneal gastrointestinal stromal tumor arising from kidney

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ABSTRACT

Mesenchymal tumors of the gastrointestinal tract which arise from the interstitial cells of Cajal and express C-Kit protein or CD117 on immunohistochemistry are known as gastrointestinal stromal tumors (GISTs). Extraperitoneal GISTs (EGISTs) are rare tumors arising from the mesentery, omentum, or retroperitoneum. We report a case of a 52-year-old male who presented with a huge abdominal lump arising from the right renal capsule that was found to be EGIST on histopathological examination and immunohistochemistry.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract.[1] They arise from the interstitial cells of Cajal which express C-Kit (CD117) protein, a tyrosine kinase growth factor receptor which is expressed in more than 95% of cases of GISTs and is the most common immunohistochemical marker. Stomach, small intestine, colon, rectum, and esophagus are the most common site of GISTs.

Tumors similar to GISTs may arise from structures outside the gastrointestinal tract such as omentum, mesentery, and retroperitoneum and are called extraperitoneal-GISTs (EGISTs).[2]

We present a case of a primary EGIST arising from the right renal capsule which wasmanaged surgically.

CASE REPORT

A 52-year-old male presented to us with progressive abdominal discomfort and swelling of 3 months' duration along with loss of appetite and weight. On examination, a huge nontender firm mass (20 cm × 15 cm) occupying the whole of the right side of the abdomen and crossing the midline up to the left midclavicular line was palpable.

Ultrasound showed a large retroperitoneal lesion of size 18.5 cm × 15 cm of heterogeneous echogenicity and encasing the right kidney with loss of fat planes with the right kidney.

Contrast-enhanced computed tomography of abdomen was done which showed a 17 cm × 17 cm × 20 cm heterogeneous right retroperitoneal lesion with peripheral enhancement. Central necrosis was noted with loss of fat planes with right renal cortex postero-inferiorly. The right kidney showed normal contrast enhancement and excretion. The right proximal and mid ureter were deviated medially with loss of intervening fat planes. The lesion was causing compression and superior displacement of the right lobe of liver and anterior and left lateral displacement of pancreas and bowel loops. There was also stretching and medial displacement of the inferior vena cava (IVC), aorta, and other vessels. The IVC was significantly compressed [Figure 1].[3,4] There

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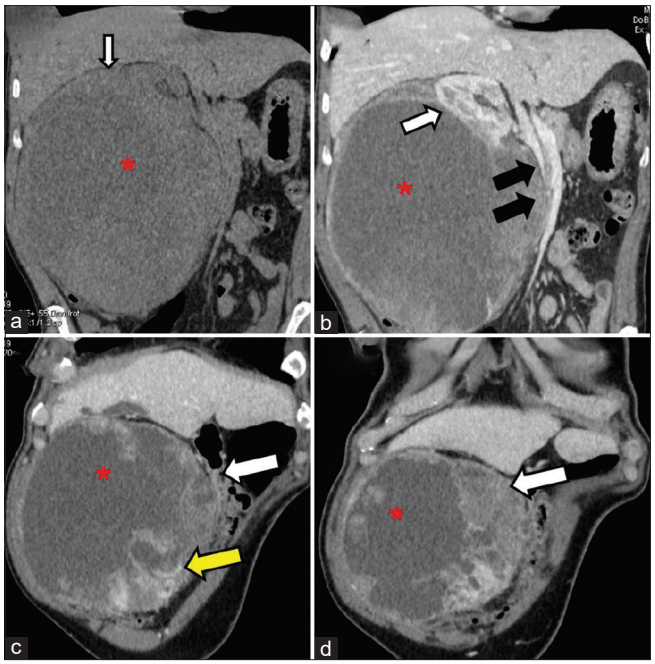

Figure 1: (a) Plain computed tomography scan shows the mass abutting the inferior surface of the liver and occupying the entire right subhepatic with necrosis (*). (b) The mass encasing and displacing the right kidney (white arrow) and splaying the adjacent renal vein and inferior vena cava (black bold arrows) with necrosis (*). (c) Displacement of ascending colon and hepatic flexure (white arrow). The mass shows necrosis (*) and peripheral enhancing nodular soft tissue (yellow arrow). (d) Peripheral soft-tissue component (white arrow) on the delayed and equilibrium phase of the dynamic computed tomography study with central liquefaction (*)

was no evidence of any other mass lesion in the remaining retroperitoneum or the GI tract.

In view of radiological finding of involvement of the right kidney, a clinical diagnosis of right renal malignancy (leiomyosarcoma) was made and exploratory laparotomy for excision of the mass through a modified chevron incision performed.

Exploratory laparotomy revealed a huge swelling of variegated consistency with no evidence of metastasis to liver and no retroperitoneal lymph nodes. Caecum and ascending colon were adherent to the mass and pushed anteriorly and to the left across the midline. IVC was splayed and adherent to the mass posteromedially. The right kidney was not visible being entirely engulfed by the mass with the right renal vessels entering the mass. Posteriorly, the mass was adherent to the psoas fascia. Frozen section from the medial (left) margin was found to be negative for malignancy. Since the right kidney was engulfed by the mass, the mass was removed *en bloc* along with the psoas fascia and weighed 5.5 Kg. Additional slices of tissue from the margin of resection at the ascending colon were taken and send for the frozen section which was negative. The course during hospital stay was uneventful and the patient was discharged on day 3 postoperatively.

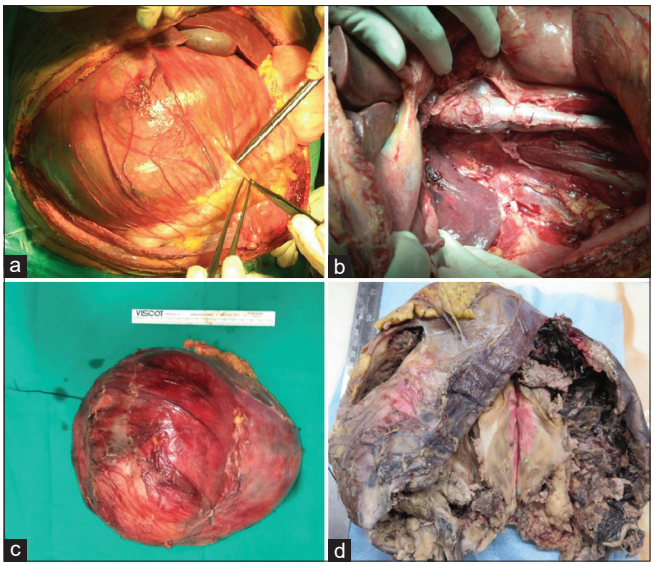

Figure 2: (a) Intraoperative picture showing the cecum and ascending colon pushed anteromedially by the mass. (b) Tumor bed after the resection of the mass showing inferior vena cava left renal vein and right psoas muscle denuded of its fascia. (c) Resected tumor. (d) Specimen of the retroperitoneal tumor composed of solid areas, necrosis and hemorrhage. It shows outer and inner aspect of tumor and the encased kidney with multiple areas of capsular invasion with focal infiltration of cortex at lower pole

Histopathology revealed an encapsulated retroperitoneal tumor composed of solid areas, necrosis and hemorrhage and was completely enclosing the right kidney with tumor cells showing diffuse C-Kit immunostaining [Figures 2 and 3]. The patient was started on imatinib mesylate and was doing well till 4 months after surgery on follow-up but was lost to follow-up after that.

DISCUSSION

Kidney, adrenal gland, retroperitoneal lymph nodes, and other soft tissues are the primary site of origin of retroperitoneal neoplasm.[5] In our patient, the large tumor mass was completely encasing the right kidney and appeared to be originating from the renal capsule with focal invasion of the renal parenchyma at the lower pole. In view of the clinical presentation of a large mass of a relatively short duration and apparently arising from the kidney, a differential diagnosis of renal sarcoma, sarcomatoid renal tumor, and renal cell carcinoma was entertained. The presence of c-kit (CD 117) established the diagnosis of GIST in our case.

Only few cases of EGIST have been documented in literature.[6] In view of the rare occurrence of GIST arising from retroperitoneum, it was a diagnostic challenge. Some experts use imatinib mesylate for treatment of such tumors but there is no consensus regarding its use.

Primary stromal tumors of the kidney have been reported in transplant recipients and as coincidental pathology along with other renal tumors. This is the first case of EGIST in a nontransplant kidney to be reported as far as we could search.

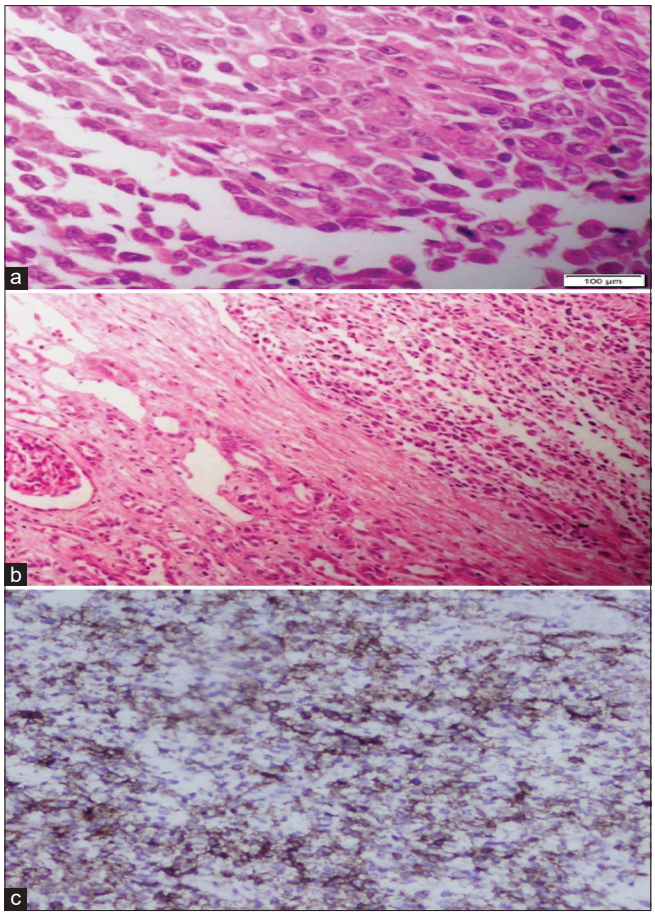

Figure 3: (a) Sheets of epithelioid cells with condensed eosinophilic cytoplasm, well-defined cell border, round to spindled nuclei with prominent nucleoli. (b) Sheets of epithelioid cells with plump spindle cells adjacent to renal parenchyma represented by a glomerulus and tubules. (c) Tumor cells show c-kit immunostaining

CONCLUSION

Primary stromal tumors of retroperitoneum are rare, and an EGIST arising from a native kidney has not been reported. Surgical removal of tumor along with involved organs is the treatment of choice. The data of reported cases in the literature need to be reviewed for better understanding of etiopathogenesis, clinical behavior, and treatment of this disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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