Case Report

Primary renal angiosarcoma

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ABSTRACT

Primary angiosarcoma of the kidney is a rare tumor with only a few case reports in the literature. Management is not standardized and the prognosis is poor. However, clinicians need to be aware of this uncommon entity.

Key words: Angiosarcoma, primary, renal

INTRODUCTION

Angiosarcomas occur commonly in the skin, liver, spleen, bone, and breast and may metastasize to the kidney. However, primary renal angiosarcoma is an extremely rare entity with only around forty case reports in the literature. The treatment protocols are ill defined and prognosis seems to be universally poor.[1] To the best of our knowledge, this is the first case report from India.

CASE REPORT

A 67-year-old gentleman presented with dull aching left loin pain since the last two months. He also had malaise, loss of appetite and significant loss of weight. There was no hematuria, headache, hemoptysis or bone pain. There was no history of previous exposure to radiation or agents like thorotrast and vinyl chloride but he had a sixty pack year history of smoking. Physical examination revealed pallor and a large ballotable mass in the left flank.

On evaluation, his hemoglobin was 5.7 gm% and creatinine was 1.72 mg%. Liver function tests and chest X-ray were normal. Contrast-enhanced computed tomogram revealed a 13 × 12 × 10 cm, predominantly hypodense left upper and inter-polar renal mass with few areas of enhancement, abutting the posterior abdominal wall, psoas and spleen [Figure 1]. After optimization, he underwent open radical nephrectomy through a subcostal transperitoneal approach. Intraoperatively, the left renal mass was seen to involve the entire kidney and infiltrate the splenic capsule, diaphragm and the psoas muscle.

Grossly, the tumor measured 13 × 10 × 7 cm. Sectioning revealed a hemorrhagic surface admixed with areas of necrosis. The posterior aspect showed a raw area with breach of capsule and attachment to remnants of diaphragm. The tumor was not seen to infiltrate the renal sinus or hilum [Figure 2].

Microscopy showed renal parenchyma infiltrated by an extensively hemorrhagic and focally necrotic tumor [Figure 3]. Scanty viable tumor, composed of clusters of spindle to polygonal-shaped cells with moderately pleomorphic and mitotically active nuclei was seen. Some tumor cells were seen to line small spaces containing RBCs [Figure 4]. On immunohistochemistry, the tumor cells were diffusely positive for CD31, CD34 and vimentin and were negative for pancytokeratin and EMA [Figures 5 and 6]. These features are consistent with angiosarcoma. Ki proliferative index was high (>80%).

He had an uneventful postoperative recovery and was started on paclitaxel-based chemotherapy. Paclitaxel was given in a dose of 115 mg intravenously, weekly for 3 weeks, followed by one week rest. The next cycle was started on day 28 and a total of 3 cycles are planned.

DISCUSSION

Angiosarcomas usually arise in the skin and superficial soft tissues, from the endothelium of the blood vessels

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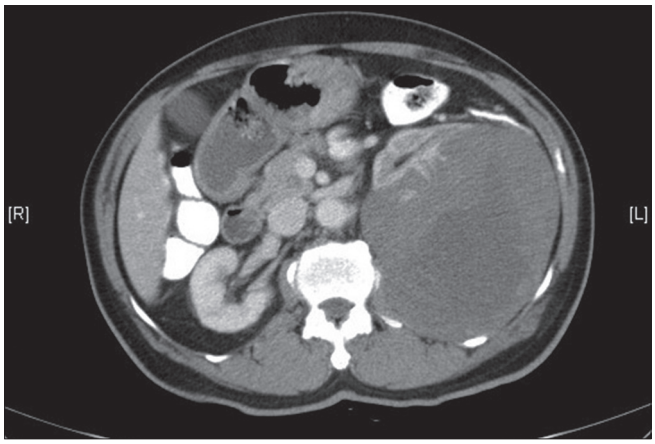

Figure 1: CECT predominantly hypodense left renal mass with few areas of enhancement, abutting the posterior abdominal wall, psoas and spleen

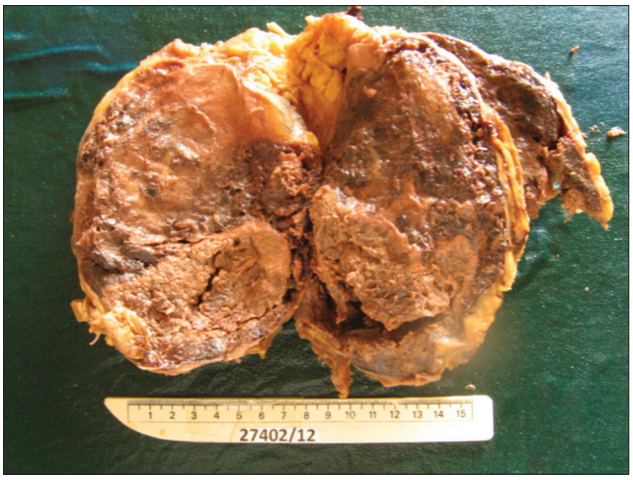

Figure 2: Gross: Hemorrhagic cut surface admixed with areas of necrosis

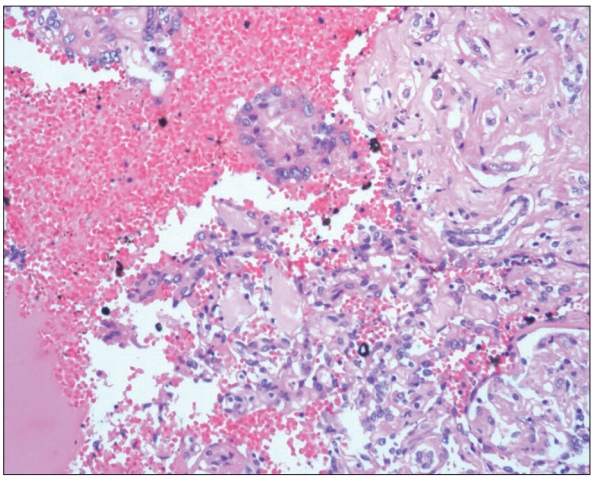

Figure 3: Microscopy: renal parenchyma infiltrated by an extensively hemorrhagic and focally necrotic tumor (H and E, ×200)

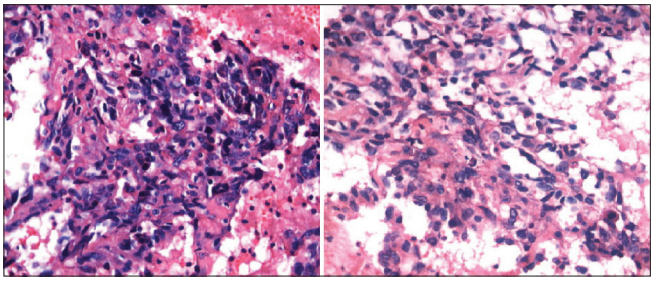

Figure 4: Microscopy: Scanty viable tumor, composed of clusters of spindle-shaped cells with moderately pleomorphic and mitotically active nuclei. Some tumor cells seen to line small spaces containing RBCs

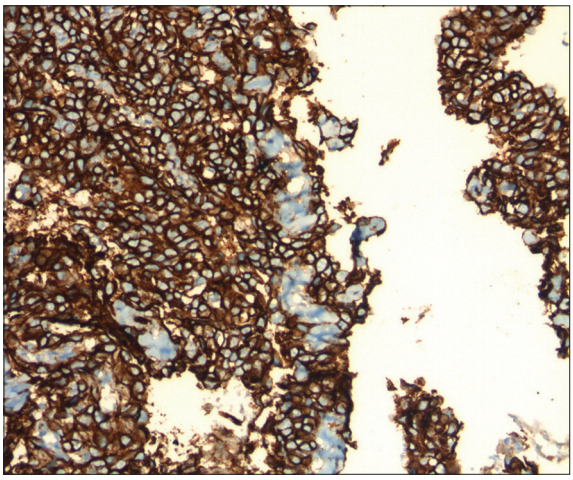

Figure 5: Immunohistochemistry: CD31+

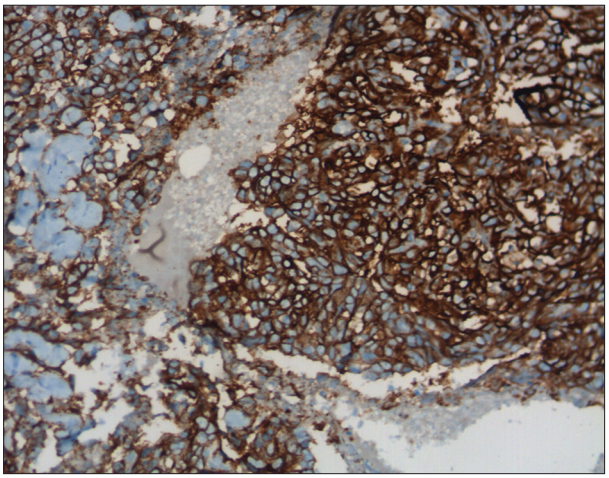

Figure 6: Immunohistochemistry: CD34+

and lymphatics. Renal metastasis is more common than the primary involvement of the kidney. Primary renal angiosarcomas usually occur in the sixth–seventh decade of life and men are affected much more often than women.[1] Certain risk factors are implicated in angiosarcomas, which include exposure to radiation or agents like arsenic, vinyl chloride and thorotrast. Presentation may include flank pain, hematuria, abdominal mass, weight loss and asthenia.[2]

These are highly aggressive tumors with a very dismal prognosis. Early metastasis is common and very few patients survive beyond one year from the time of diagnosis. Clinical and radiological assessment can only suggest a malignant renal mass and diagnosis is possible only after

histopathological examination of the nephrectomy specimen and confirmation with positive immunohistochemical staining for CD-31, CD-34 and factor VIII.[3]

Tumor size has been recommended as a tool to prognosticate primary renal angiosarcomas, with < 5 cm being suggested as a cut off for better prognosis.[4] However, the numbers are too few to suggest definitive outcomes.

Following surgery, adjuvant therapy in the form of radiation and chemotherapy has been suggested based upon results from angiosarcomas in other body parts. Taxanes and ifosfamide-based chemotherapy regimens have been proposed.[2,5] However, rarity of this entity precludes the standardization of the management protocols.

CONCLUSION

Primary renal angiosarcoma is an extremely rare entity. Literature is sparse and though radical nephrectomy is the initial treatment, further management issues are unresolved for this highly aggressive tumor. Urologists need to be aware of this entity and a multidisciplinary approach is required to define adequate treatment protocols.

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