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Case Report

Urology Section

When Anomalies Meet Tumour: A Case of Leiomyomas in a Horseshoe Kidney

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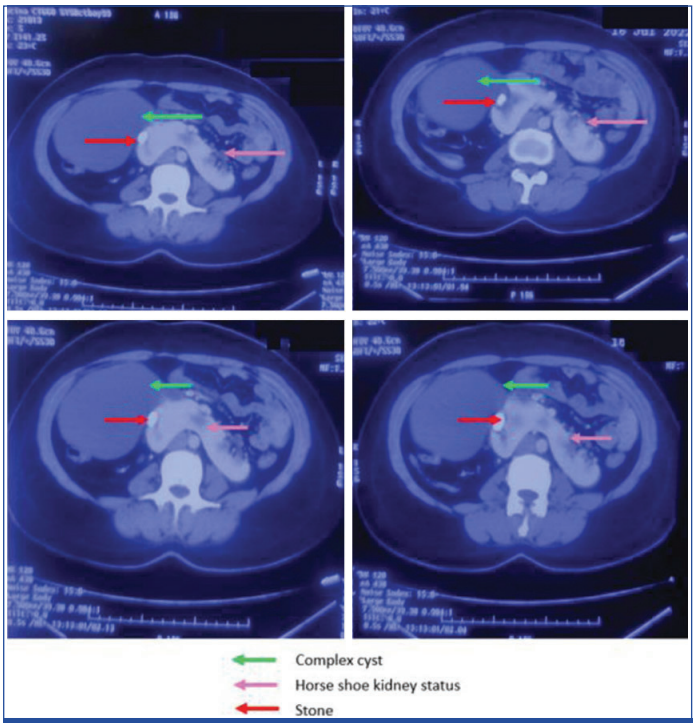
ABSTRACT

The horseshoe kidney is one of the most common congenital renal fusion anomalies of the genitourinary system. It may be associated with other diseases chiefly involving the gastrointestinal tract, skeletal, cardiovascular and central nervous system. With cases of renal tumours of varied types, the occurrence of smooth muscle tumours is rare. The patient may be asymptomatic or present with abdominal/flank pain, palpable mass or haematuria. The increased use of imaging has led to incidental findings of these leiomyomas. The below-presented case is a rare presentation of renal leiomyoma in a 39-year-old female with horseshoe kidneys. The diagnosis was confirmed on immunohistochemistry data. This unique association of renal leiomyoma with horseshoe kidneys is, as per our knowledge, the first reported case, suggesting clinicians consider such a rare differential diagnosis in renal pathology. Knowledge of such rare associations focuses on designing definite treatment protocols for minimising radical nephrectomy.

Keywords: Leiomyosarcoma, Radical nephrectomy, Smooth muscle tumours

CASE REPORT

A 39-year-old female presented with right flank pain with a vague lump; slowly increasing in size in the right lumbar region for two years. The pain was intermittent and dull aching in nature, spontaneous in onset, with no aggravating factors. The patient sometimes took oral analgesics for relief. A contrast-enhanced CT scan of the abdomen revealed a horseshoe kidney entity with lower poles fused at the L4 level, an exophytic cystic lesion arising from the lower pole of the right moiety with enhancing solid component (Type IV Bosniak) [Table/Fig-1]. A 14 mm right renal pelvic calculus was also noted.



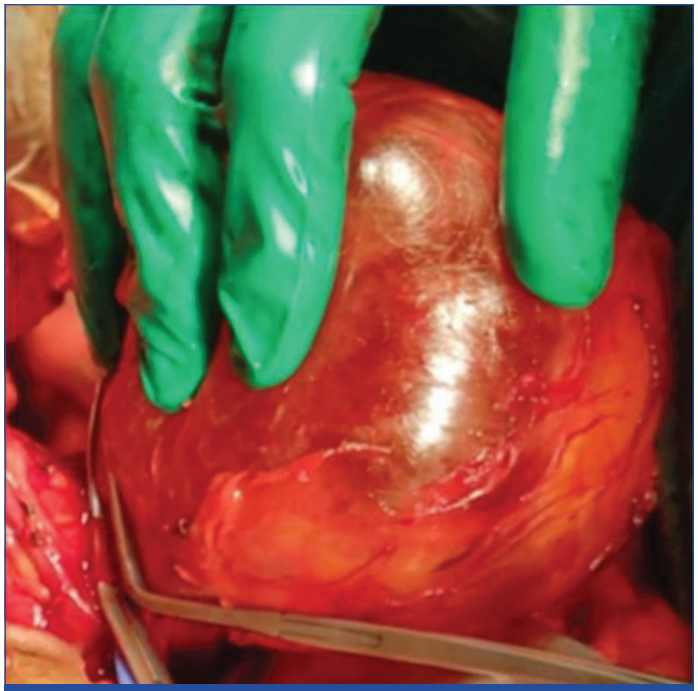
[Table/Fig-1]: CT scan showing cyst with solid component in right moiety in horseshoe kidney.

On the intraoperative retrograde pyelogram, incomplete duplication of the right ureter was noted [Table/Fig-2]. Given abnormal anatomy and a large volume of mass, the decision to perform an open surgery was made. A 12 cm solid cystic eccentrically located mass

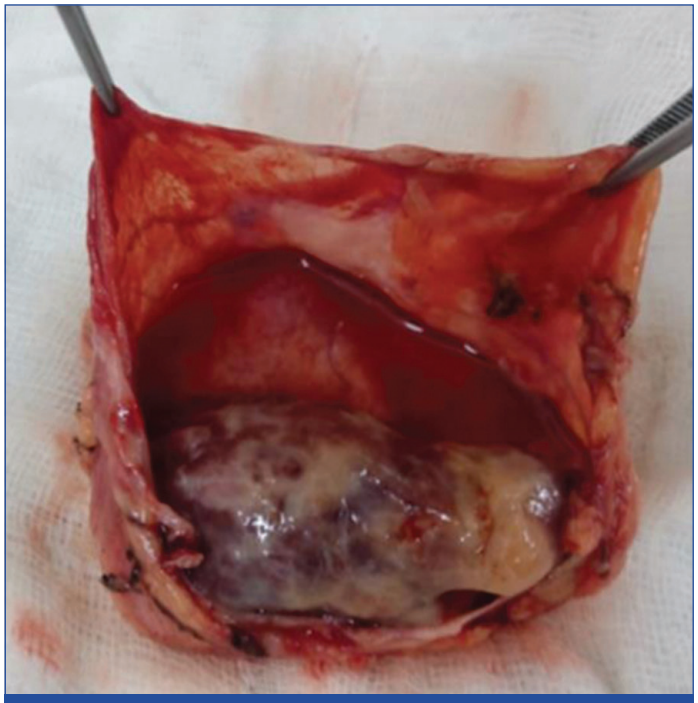
[Table/Fig-2]: Retrograde pyelogram of right-side showing incomplete ureteral duplication.

was noted arising from the lower pole of the right moiety [Table/Fig-3]. Wide local excision of the mass was performed, in view of abnormal anatomy and eccentric location of the mass, along with pelvic stone removal, followed by Double-J (DJ) stenting.

On bench dissection, a cystic mass with a solid component was noted and sent for histopathological evaluation [Table/Fig-4]. This finding was suggestive of the cuboidal epithelium of the cyst wall and the sections from the attached tissue mass showed interlacing fascicles of spindles and areas of hyalinisation. Immunohistochemical studies were positive for desmin and smooth muscle actin, while cyst wall epithelium was positive for Pax-8. These features were suggestive of renal cysts with leiomyoma. Postoperative course was uneventful, patient was discharged on POD-4. Patient did fine on follow up, Right DJ stent removal was done after six weeks. Patient was asymptomatic on follow-up visits at three and six months.



[Table/Fig-3]: Intraoperative image of the leiomyoma mass.



[Table/Fig-4]: Specimen image confirming CT findings of cystic mass with solid component.

DISCUSSION

The horseshoe kidney is one of the most common renal fusion developmental disorders of the genitourinary tract, with approximately 1-4 per 1000 births and males are more affected than females [1]. Almost 25% of horseshoe kidneys in their lifetime require operative treatment [2]. There are many congenital anomalies and syndromes associated with horseshoe kidney [3]. The risk of developing cancer is similar in cases with and without horseshoe kidneys [4]. The rate of developing Wilms' tumour, transitional epithelium cancer and renal cell carcinoma is higher in patients with horseshoe kidneys [5,6]. Presenting signs of horseshoe kidneys comprise flank pain, palpable mass or haematuria. A large number of renal cancers arise in the isthmus, suggesting teratogenic factors are responsible for the abnormal migration of nephrogenic cells to form the isthmus [7].

Renal leiomyomas are rare, benign tumours of the kidney originating from smooth muscle cells. These smooth muscle tumours are located mainly in the urinary bladder and uterine myometrium, but their presentation in the renal parenchyma is rare since smooth muscles are located only in the renal blood vessels, renal capsule, pelvis and calyces [8]. Small lesions are majorly asymptomatic and identified during autopsy, and the larger lesions might cause pain and swelling in the abdomen. Renal leiomyomas are majorly reported in adults from the second to sixth decade of life, with females being more affected than males [8]. Data from a study by Gupta S et al, proposes that these tumours have hormonally responsive smooth muscles as in cases of uterine leiomyomas, suggestive of its increased prevalence in females [9].

Microscopically, they are well circumscribed with long fusiform spindled cells and eosinophilic cytoplasm with elongated nuclei. Immunohistochemically, these tumours show positivity for actin, smooth muscle myosin, vimentin, desmin and laminin. They have a very low Ki-67 proliferation index [10].

Horseshoe kidneys have been a common developmental derangement, and associated tumours have been reported. The exact pathogenesis of tumour formation in them is unknown, but it could be a result of chronic obstruction, lithiasis or infection, which is more common in these patients [11]. In the above-presented case, the lesion was located at the lower end of the right kidney along with the presence of calculi at the right renal pelvis, favouring the possible reason for developing the pathology. Je BK et al., examined 366 children and young adults and 14 foetuses with horseshoe kidneys [12]. One-half of the cases had extrarenal disease, of which gastrointestinal and vertebral anomalies were most common.

Their malignant counterpart, renal leiomyosarcoma, is more common in adults. Both renal leiomyoma and leiomyosarcoma arise from smooth muscle fibres of the renal capsule, pelvis or a blood vessel, differentiating them is crucial for final diagnosis and prognosis. Renal leiomyomas can be differentiated from leiomyosarcoma by the absence of mitotic activity, pleomorphism, hyperchromatic and perilesional invasivity [13,14]. In the above case, imaging could not confirm the diagnosis since the lesion was enclosed in a cystic mass. Immunohistochemistry confirmed this smooth muscle cell tumour and detected the pathology.

A case of primary leiomyosarcoma in a horseshoe kidney in a female with Turner syndrome has been reported and published in Japan in 2014 [15]. Due to the risk of aggressive behaviour which may cause metastasis, open right heminephrectomy and division of the isthmus were performed. Patient was asymptomatic and disease free at six months follow-up.

In the documented few cases of renal leiomyoma, radical nephrectomy was done in all cases because of a lack of accurate pre-op diagnosis [16]. Leiomyomas are often detected incidentally, and radical nephrectomy happens to be the gold standard management. However, less radical approaches may be considered in cases of radiologically benign-appearing masses. The prognosis is excellent, and no extrarenal invasion has been reported [17,18].

The aim of presenting this case is to highlight the rare occurrence of leiomyoma in a female with a horseshoe kidney, which was difficult to diagnose on imaging, and after immunohistochemical studies, the confirmed diagnosis was made. Knowledge of such rare associations focuses on designing definite treatment protocols for minimising radical nephrectomy. To the best of our knowledge, no case of renal leiomyoma in a horseshoe kidney has been reported till now. Reporting such cases globally is necessary for establishing consistent treatment guidelines.

CONCLUSION(S)

With such a rare presentation of smooth muscle tumours in horseshoe kidneys, clinicians should be careful in accurate diagnosis before planning for radical nephrectomy. Histologically, leiomyomas might be similar to angiomyolipomas or leiomyosarcoma. Hence, immunohistochemical evaluation is crucial in confirming the

diagnosis. Being benign and non-aggressive a surgical excision or lesser radical approach can be performed.

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