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CASE REPORT

An interesting case of a mucinous cystadenoma of a horseshoe kidney in a middle-aged woman

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Abstract

A 48-year-old female presented with complaints of right sided vague abdominal pain associated with abdominal lump.. Computed tomography revealed horseshoe kidney with Bosniak IIF cystic lesion arising from the interpolar region of right segment of the horseshoe kidney (HSK). In view of the persistent pain, patient underwent cyst excision. The final histopathology diagnosis was mucinous cystadenoma. On follow-up at 6 months post surgery, there was no recurrence. The histopathology is very unique and rare with only a few cases recorded in literature. The inner lining of the cyst wall composed of columnar epithelium secreting mucin and at some places by transitional epithelium. This indicates that the origin of the mucinous tumour was likely to be from a sequestration of the renal pelvic urothelium in the renal parenchyma.

Keywords: Horseshoe kidney, Mucinous cystadenoma, renal cyst, Boniak

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Introduction

The occurrence of a primary mucinous cystadenoma in a horseshoe kidney is a very rare case with only 3 cases reported in literature [1-4]. We would like to describe our case as the fourth one to add to the literature of this rare clinical scenarios.

Case Report

A middle aged female of age 48 years presented to the urology outpatient department with complaints of vague right sided intermittent abdominal pain over a period of 6 months. An ultrasound of the abdomen revealed horseshoe kidney with a well-defined hyperechoic lesion with some hypoechoic areas within the lesion arising from the interpolar region of the right segment. Patient was then

further evaluated by Contrast enhanced computed Tomography of the abdomen and pelvis which revealed a horseshoe kidney with the lower poles of both the kidneys fused anterior to the abdominal aorta at the level of L3 lumbar vertebra with the isthmus measuring 11 mm in diameter. There was evidence of well-defined hypodense non-enhancing cystic lesion of size 7.3 cm *6.5 cm *7 cm in dimensions in the retroperitoneal region arising likely from the interpolar region of the right division of the horseshoe kidney compressing and displacing right renal hilum. The cystic lesion shows few thick calcified non enhancing septae and multiple peripheral wall calcifications suggestive of Bosniak type IIF lesion (Figures 1 to 3).



Figure 1. CECT scan of abdomen and pelvis showing Bosniak II F lesion arising from the interpolar region of the right segment of the horseshoe kidney (Arterial phase-axial cuts)



Figure 2. CECT scan horseshoe kidney with the cyst and the isthmus at the level of L3 vertebra.



Figure 3. Coronal section cuts of CECT scan showing the origin of the cyst from the Right segment of the horseshoe kidney.

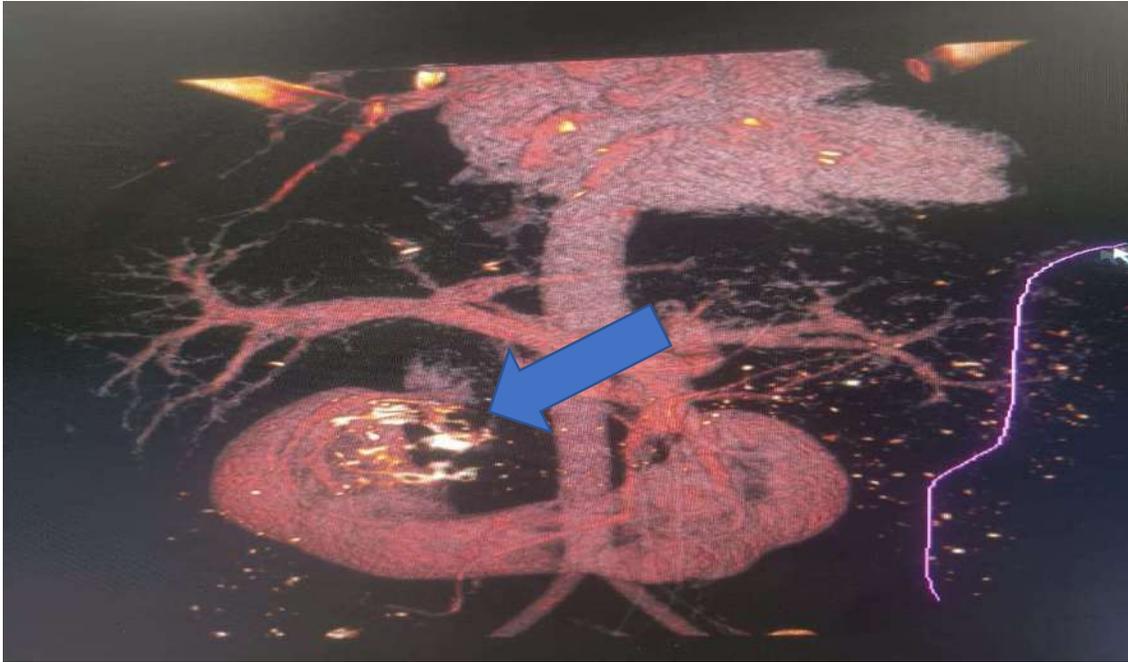


Figure 4. Reconstructed CT image of the horseshoe kidney with the origin of the cyst from the interpolar region of the right segment (arrow showing the cyst)

In view of the persistent pain, patient was planned for cyst excision. The cyst excision was done by an upper midline abdominal incision. After careful mobilisation of the right colon and kocherisation, the Right sided segment of the HSK along with the cyst was exposed. After meticulous dissection, the cyst

was carefully dissected out from the renal parenchyma away from the renal hilum and excised in toto. On cutting open the cyst; it was found to contain mucinous material. The procedure was uneventful. Patient was discharged 5 days after the procedure (Figures 4-9).



Figure 5. Preoperative photograph of the patient's abdomen marked with the location of the abdominal lump using a marker.



Figure 6. Shows the relation of the cyst to the liver, duodenum and the colon.

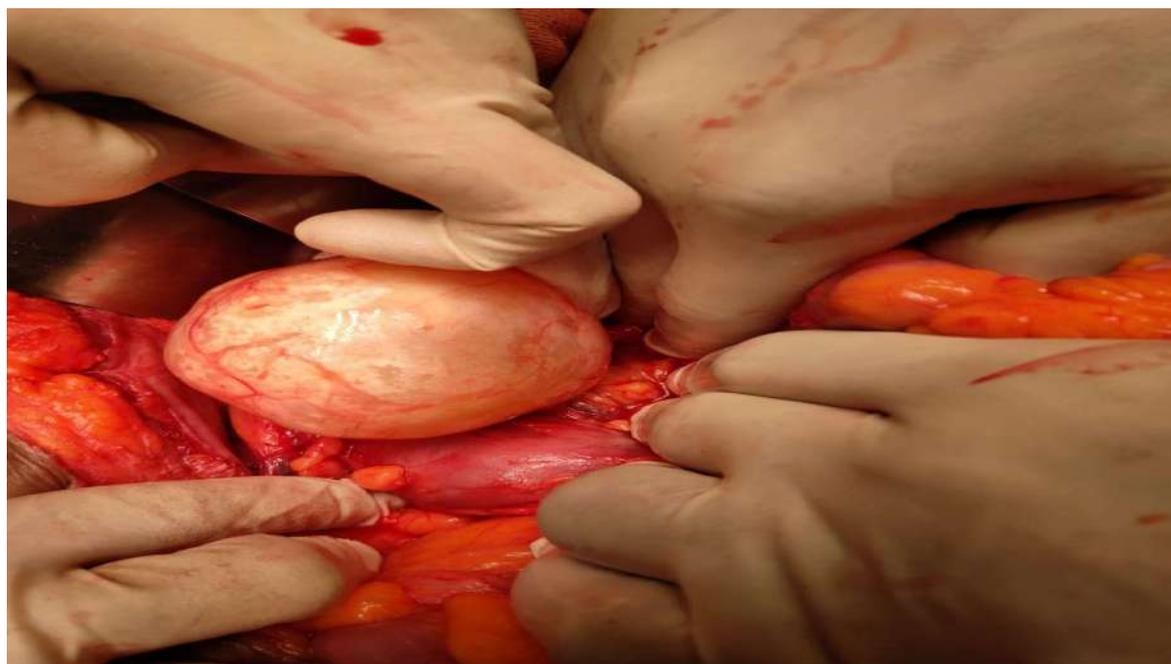


Figure 7. Shows the relation of the cyst to the right segment of the Horseshoe kidney.

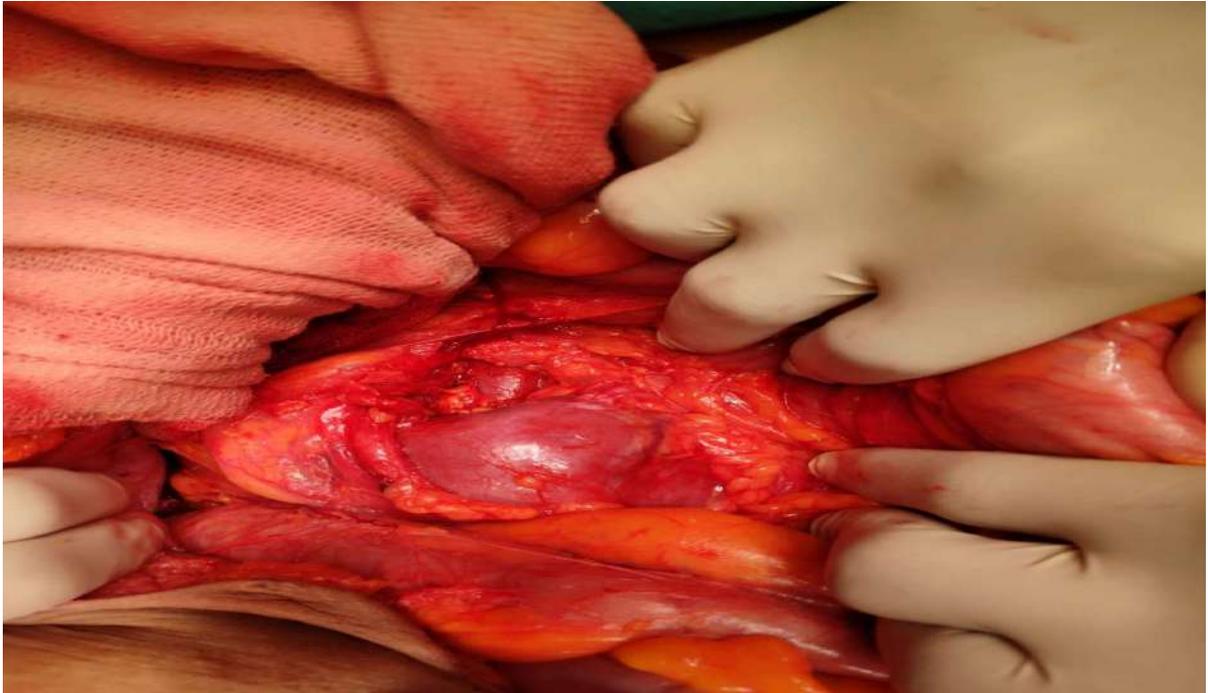


Figure 8. Shows the right segment of the horseshoe kidney along with the isthmus after complete removal of the cyst.

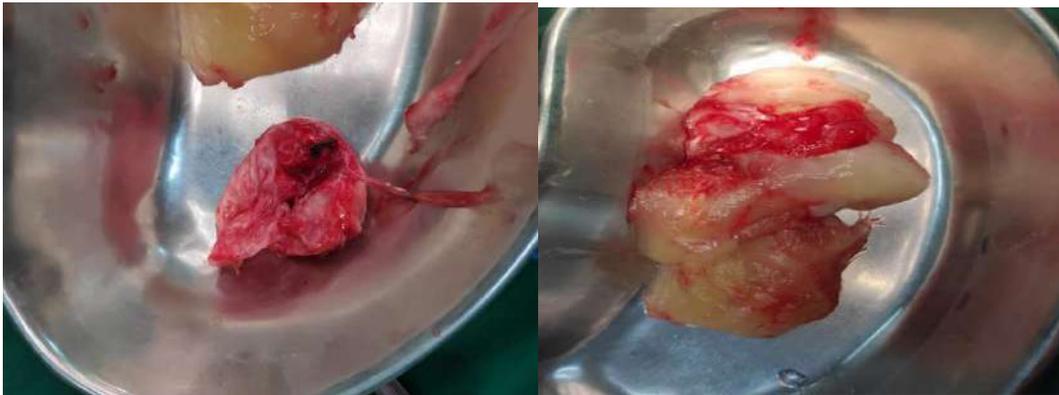


Figure 9. Shows cyst cut open with mucinous contents.

The final histopathology report revealed a fibromuscular cyst wall lined by a single layer of mucin secreting columnar lining epithelium admixed with transitional epithelium at some places. The fibromuscular wall was seen densely infiltrated with chronic inflammatory cells composed of lymphocytes and plasma cells. These findings were suggestive of final diagnosis of mucinous cystadenoma of horseshoe kidney. The patient has been on regular follow up post-surgery.

There is no recurrence over a period of 6 months.

Discussion

We describe an extremely unique case of mucinous cystadenoma in a horseshoe kidney. To our knowledge, there have been only 14 reported cases of mucinous cystadenoma of the kidney, and only 3 of such cases have been described in a horseshoe kidney. These tumors cannot be distinguished from a renal cyst based on clinical examination

findings, laboratory data, and imaging studies. None of the previous reports correctly diagnosed the disease before surgery; the final diagnosis was made only after the tumour was excised and final histopathology report was obtained. The density of simple renal cysts ranges from -10 to +20 HU on CT [3]. With our patient, the density of the cystadenoma ranged from 10 to 20 HU but with few septae and few calcifications and the radiologic features mimicked a renal parenchymal cyst- Bosniak IIF [3]. Also because of some floating membranes seen on ultrasound by the radiologist, even a differential diagnosis of hydatid cyst was on the cards. Mucinous cystadenoma of the kidney may therefore be missed by routine imaging studies. In the previous reports, the cystadenoma originated from the urinary collecting system in 8 cases and from the renal parenchyma in the remaining 4, based on histological findings [1-10]. Some theories by authors have postulated that the development of mucinous cystadenoma with an intestinal phenotype in the kidney involves urolithiasis, infections, or both, and these events may induce intestinal metaplasia [7]. In contrast, Liwnicz and colleagues suggested that the mucin secretion from the cystic tumour may be the cause of the stones [11]. On the other hand, other authors have speculated that from a sequestered segment of the renal pelvic urothelium in the renal parenchyma, especially in the case of an anomalous kidney, such as a HSK have been associated with such mucinous tumors [3,4]. In cysts arising from the renal

parenchyma, which include the 2 cases reported in horseshoe kidneys, the inner surface was not covered by urothelium. In our case, the histopathologic findings demonstrated an inner surface of the cyst covered by a mucin positive columnar epithelium admixed with transitional epithelium, suggesting that the origin of the mucinous tumour was likely from a sequestered segment of the renal pelvic epithelium.

Conclusion

We report an extremely unique interesting case of mucinous cystadenoma in a HSK. Thus, we conclude that horseshoe kidneys present with unique challenges to the urologist in his clinical practice.

Author Contributions

Ojas Vijayanand Potdar (Corresponding author): Design, patient history taking and writing the manuscript of the case report. Kaustubh Vaidya: Writing the manuscript of the case report. Mohammed Ayub Karamnabi Siddiqui- Design of the case report. Vivek Shaw: Patient history taking. Darshan Rathi- Images of the radiological investigations. Amrita Vikram Patkar: Writing the manuscript of the case report.

Conflicts of interest

The authors declares that they do not have conflict of interest.

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