CASE REPORT

Renal cell carcinoma with isolated metastasis to sigmoid mesentery: a rare resectable combination

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SUMMARY

Renal cell carcinoma accounts for 2%–3% of all malignancies in adults. It spreads via direct extension, lymphatic route as well as haematogenous route. Lymph nodes, lungs, bone, liver and brain are the usual sites for its metastatic spread. In the presence of limited metastatic disease with potentially resectable metastases, surgery offers the best chances of cure. In the present case, we describe a case of renal cell carcinoma with a solitary metastasis to the sigmoid mesentery in a patient with Von Hippel-Lindau syndrome. There was no retroperitoneal lymphadenopathy or tumour thrombus in the renal vein. The patient was managed with laparoscopic radical nephrectomy and excision of the sigmoid mesentery mass. At 6 months of follow-up, there is no evidence of recurrent disease.

BACKGROUND

Of the patients with renal cell carcinoma who present with a localised disease, up to 40% develop metastasis during follow-up.¹ Although the spread occurs unpredictably, the usual sites are downstream areas such as the lymph nodes and lungs. There are multiple case reports of unusual metastasis to different organ system with most common being gastrointestinal system.^{2,3} We report a case of unusual isolated synchronous metastasis to sigmoid mesentery in a clear cell renal cell carcinoma (RCC) associated with Von Hippel-Lindau syndrome.

CASE PRESENTATION

A 30-year-old female patient presented with history of gradual onset weakness in all four limbs for 6 months. She had no sensory symptoms and there

Figure 2 Contrast enhanced CT abdomen in axial plane at the level of kidneys showing heterogenous mixed density lesion in the upper pole of the right kidney in posteromedial cortex (arrow). A simple pancreatic cyst is also seen (asterisk).

was no history of fever. There were no other complaints. Her mother had died at a young age due to RCC. On examination, she was conscious, oriented and her higher mental functions were intact. Neurological examination localised the lesion to cervical spine. Abdominal examination was unremarkable.

INVESTIGATIONS

MRI of spine with brain screening revealed a cystic lesion with avid enhancing mural solid component involving the cervical spine at C2–C6 level (figure 1). A provisional diagnosis of haemangioblastoma in cervical spine was made. Since the patient also had history of renal cancer in mother, she was worked up for VHL syndrome and found to be positive for the same.

Ultrasonography of abdomen showed multiple well-defined anechoic lesions without solid component, septae or any echogenic debris within pancreatic parenchyma suggestive of multiple simple pancreatic cysts. Rest of the pancreas was normal. The main pancreatic duct was also normal. There was also a well-defined hypoechoic lesion with few hyperechoic septae and a peripheral isoechoic solid component involving the upper pole of right kidney suggestive of a complex cyst. On contrast enhanced CT abdomen, a lobulated heterogenous mixed density lesion measuring 3×2.1 cm was seen in the upper pole of right kidney with multiple enhancing septae and peripheral irregular enhancing solid component suggestive of Bosniak type IV cyst (figure 2). Two small simple cysts were

Figure 1 MRI of cervical spine in sagittal plane. (A) T2-weighted image (WI) showing hyperintense signal in the spinal cord. (B) Postcontrast T1-WI showing cystic lesion with avid enhancing mural solid component (asterisk).

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Figure 3 Contrast enhanced CT showing a solid enhancing lesion in the sigmoid mesentery in right hemipelvis (arrow).

also noted in midpole of right kidney. There was no retroperitoneal lymph node enlargement. Multiple pancreatic cysts were confirmed on CT and there was another simple cyst in segment VIII of liver. The CT also showed a well-defined solid enhancing lesion measuring 2.1×1.8 cm in the sigmoid mesentery which was located just above the urinary bladder (figure 3).

DIFFERENTIAL DIAGNOSIS

1. Synchronous metastasis from RCC: rare if occurs as an isolated finding; shows enhancement on CT similar to the renal mass.
2. Paraganglioma: may occur as one of the associated findings in patients with VHL syndrome; highly enhancing on CT.

TREATMENT

Free fractionated plasma metanephrines and the 24 hours urinary fractionated metanephrine levels were normal. The patient underwent right laparoscopic radical nephrectomy for the endophytic renal mass abutting the renal hilum along with resection of the pelvic mass (figure 4). Histopathological evaluation of the renal mass revealed tumour composed of small nests of tumour cells displaying mildly pleomorphic round to oval nuclei, vesicular chromatin, conspicuous nucleoli and variable amount of clear cytoplasm (figure 5). Sections from the mesenteric mass showed a tumour, predominantly arranged in papillary pattern lined by fibrovascular core. The papillae were lined by tumour cells with minimal nuclear atypia and moderate amount of clear cytoplasm (figure 6). On immunohistochemistry, the primary renal tumour cells as well as the metastatic tumour cells were positive for CD10

Figure 4 Right nephrectomy specimen showing mass lesion in the upper pole (arrow).

(figure 7). Based on the above findings, the renal lesion was diagnosed as clear cell RCC and the lesion in sigmoid mesentery was diagnosed as metastasis from clear cell RCC.

Three months later, the patient underwent C1–C2 laminectomy with excision of haemangioblastoma.

OUTCOME AND FOLLOW-UP

The patient initially had worsening of weakness after excision of haemangioblastoma which gradually improved and the patient was discharged on the eighth postoperative day. At 6 months of follow-up, the patient is ambulatory and asymptomatic.

DISCUSSION

RCC is mainly the malignancy of the elderly people and accounts for nearly 3% of all malignancies and 85% of renal malignancies. It usually presents in sixth to seventh decade.⁴ However, RCC is more frequent in younger patients in India and also occurs at early age.^{5,6} About 50% of patients with VHL syndrome develop RCC and it is characterised by early onset (third to fifth decade), multifocality and bilaterality.⁷ The familial RCC arises due to mutations in VHL gene located on short arm of chromosome 3 (3p).

The metastasis in RCC can be synchronous or metachronous and the most common site of metastasis include lung, bone, liver, adrenal gland, contralateral kidney, retroperitoneum and brain in the decreasing order of frequency.^{8,9} The rare or unusual sites include pancreas, large intestine, stomach, duodenum, small

Figure 5 Histopathological evaluation of the renal mass at (A) low magnification and (B) high magnification, showing tumour composed of small nests of tumour cells displaying pleomorphic nuclei, vesicular chromatin and variable amount of clear cytoplasm.

Figure 6 Histopathological evaluation of the mesenteric mass at (A) low magnification and (B) high magnification, showing tumour cells with minimal nuclear atypia and moderate amount of clear cytoplasm, arranged in papillary pattern lined by fibrovascular core.

Figure 7 Immunohistochemistry showing CD10 positivity in (A) primary renal tumour cells as well as (B) metastatic tumour cells.

intestine, biliary tract, peritoneum, spleen, omentum, oral cavity, paranasal sinuses, larynx, hypopharynx, thyroid, eye and orbit, skin, skeletal muscle, uterus, ovary, vulvo-vagina, breast, urinary bladder, testicles, skin, different group of skeletal muscles and cauda equina.^{3 10–22}

Here, we present a case of isolated unusual metastasis to sigmoid mesentery from clear cell RCC in a patient with VHL syndrome. Even though the patient did not have any symptoms related to RCC or the metastasis, extensive imaging workup helped to diagnose both the primary renal malignancy and the metastatic deposit. Since surgery is the mainstay of treatment for primary as well as metastasis, imaging plays an important role in diagnosing these lesions. The role of immunohistochemistry is important in establishing relation between the mass and the metastatic lesion, particularly in case of poorly differentiated tumours and CD10 is an important marker particularly for clear cell RCC.²³

To our knowledge, this is the first case report of isolated synchronous metastasis to the sigmoid mesentery in a case of RCC associated with VHL syndrome.

Learning points

- Renal cell carcinoma occurs in about half of the patients with Von Hippel-Lindau syndrome and is usually incidental.
- Sigmoid mesentery is a rare site for isolated metastasis from renal cell carcinoma.
- Complete surgical excision of the renal mass as well as the metastatic lesion is the best form of treatment for metastatic renal cell carcinoma.

Contributors PY and UPS performed the surgery. HL and UPS conceived and edited the manuscript. PY and SA prepared the manuscript and provided the images. All the authors approved the final manuscript.

Competing interests None declared.

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