Renal carcinoma in horseshoe kidney with endoluminal duodenal invasion

Case 11288

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Section: Uroradiology & genital male imaging
Area of Interest: Abdomen Kidney Gastrointestinal tract
Procedure: Diagnostic procedure
Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
Authors: Rigiroli Francesca, Tonolini Massimo
Patient: 51 years, male

Clinical History:

Middle-aged man suffering from lower right abdomen and iliac pain radiating to ipsilateral testicle, haematuria, and melaena. Admits asthemia and 10-kg weight loss during the last months, occasional night fever and sweats. Physical examination detects palpable periumbilical mass. Laboratory tests reveal anaemia (8.3 g/dL haemoglobin), raised (50 mg/L) C-reactive Protein.

Imaging Findings:

Initially, plain radiographs (Fig. 1) showed distended stomach with air-fluid level, and a large roundish opacity in the mid-abdomen corresponding to palpatory finding instead of the usual bilateral renal opacities. Multidetector CT (Fig. 2) detected a large heterogeneously enhancing mass arising ventrally from the isthmus of a horseshoe kidney, which was previously unknown. The tumour apex was clearly seen invading the lumen of the third duodenal portion, as endoscopically confirmed. Some sizeable poorly marginated liver metastases were present, and renal cell carcinoma was confirmed by biopsy of the dominant liver lesion. Metastatic disease, adjacent organ invasion, and isthmus location contraindicated surgery, which would have required combined total nephrectomy plus pancreaticoduodenectomy. The patient survived approximately one year, during which the primary carcinoma and liver metastases showed good response following sunitinib chemotherapy (Fig. 3), with persistent duodenal invasion and appearance of right moiety hydronephrosis.

Discussion:

One of the commonest genitourinary anomalies, horseshoe kidney (HK) is encountered with a 0.25% prevalence in the general population, is twice more common in men than in women, and consists of two distinct kidneys on each side of the body connected at their lower poles by a parenchymatous or fibrous isthmus across the midline. The blood supply to HK may involve one renal artery on each side, duplicate or triplicate renal arteries. The isthmus may be reached by a renal artery branch, or may have an independent artery originating from abdominal aorta. Representing 2-3% of all adult neoplasms, renal cell carcinoma (RCC) mostly occurs in the sixth and seventh decades of life. Although in people with renal fusion anomalies the risk to develop RCC is the same as in the general adult population, the association of HK and renal tumours is uncommon, with fewer than 200 reported cases. RCC accounts for over 50% of cases, whereas other neoplasms including transitional cell or squamous carcinomas, Wilms tumour, lymphomas, carcinoid and sarcomas have also been reported [1, 2, 4]. HK is usually asymptomatic and mostly diagnosed incidentally during US, CT and MRI imaging studies. Symptoms,
when present, are due to urolithiasis, hydronephrosis, or urinary infection. In the general population, RCC is often diagnosed in advanced stages with venous or adjacent organ invasion, or distant metastases. Conversely, direct RCC extension to the duodenum has been reported only once [5]. Multidetector CT currently represents the gold standard for RCC staging, including multiplanar image reformations that show the tumour’s size, its relationship with the healthy renal parenchyma, collecting system and nearby organs, and CT-angiographic visualization of the renal vasculature. [6-8]. Locally advanced RCC requires en bloc surgery such as radical nephrectomy plus pancreaticoduodenectomy, combined with interferon or sunitinib adjuvant chemotherapy [5]. Heminephrectomy represents the surgical treatment of choice for RCC arising in HK, which usually allows preservation of sufficient renal function. Alternatively, angiographic embolization has been proposed to reduce vascular supply to the tumour, decrease bleeding and ease surgery [1, 3, 4].

In our opinion, in this patient the fusion anomaly probably favoured the otherwise exceptional duodenal invasion by a tumour arising from the isthmus. In patients with congenital kidney anomalies, a precise tumour staging with CT is even more important than in those with normal urinary anatomy, in order to choose and plan the optimal, specific surgical approach [6-8].

**Differential Diagnosis List:** Horseshoe kidney with renal cell carcinoma invading the duodenum, Lymphoma, Duodenal carcinoma, Pancreatic carcinoma, Renal abscess, Xanthogranulomatous pyelonephritis

**Final Diagnosis:** Horseshoe kidney with renal cell carcinoma invading the duodenum

**References:**


Chapin BF, Delacroix SE, Jr., Wood CG (2011) Renal cell carcinoma: what the surgeon and treating physician need to know. AJR Am J Roentgenol196:1255-1262

Description: Supine (a) and upright (b) radiographs exclude free intraperitoneal air and bowel obstruction.
A large, roundish faint opacity (arrows) occupies the centre of the abdomen, corresponding to palpatory finding. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Supine (a) and upright (b) radiographs exclude free intraperitoneal air and bowel obstruction.

Note distended stomach with air-fluid level. A large, roundish faint opacity (arrows) occupies the centre of the abdomen, corresponding to palpatory finding. 

Origin: Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Preliminary unenhanced scans (a..c) show some sizeable, ill-defined hypoattenuating liver lesions (+), a large ovoid-shaped inhomogeneous mass with speckled calcifications (arrow in b) abutting the horseshoe kidney. **Origin:** Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Preliminary unenhanced scans (a..c) show some sizeable, ill-defined hypoattenuating liver lesions (+), a large ovoid-shaped inhomogeneous mass with speckled calcifications (thin arrow in b) abutting the horseshoe kidney. **Origin:** Tonolini M, Department of Radiology, “Luigi Sacco,” University Hospital – Milan (Italy)
Description: Preliminary unenhanced scans (a..c) show some sizeable, ill-defined hypoattenuating liver lesions (+), and a solid tissue mass (arrow in c) in the lumen (*) of the third duodenal portion.

Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: After intravenous contrast, some sizeable poorly marginated hypovascular liver lesions (+) consistent with metastases are confirmed. Origin: Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
**Description:** The horseshoe kidney across the midline shows a large (11x7 cm), ovoid-shaped partly exophytic mass, with heterogeneous appearance corresponding to enhancing solid tissue at the periphery and extensive central necrosis. **Origin:** Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The horseshoe kidney across the midline shows a large (11x7 cm), ovoid-shaped partly exophytic mass, with heterogeneous appearance corresponding to enhancing solid tissue at the periphery and extensive central necrosis. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The inhomogeneous mass arises from the isthmus of the horseshoe kidney. The inferior vena cava and both renal veins are patent. Note poorly marginated liver metastases in the right lobe.

**Origin:** Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
**Description:** The duodenal lumen at the third portion (*) is occupied by an enhancing soft-tissue (arrows) corresponding to cranial invasion from the renal carcinoma arising from the horseshoe kidney isthmus. Note poorly marginated liver metastases (+). **Origin:** Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
**Description:** The duodenal lumen at the third portion (*) is occupied by an enhancing soft-tissue (arrows) corresponding to cranial invasion from the renal carcinoma arising from the horseshoe kidney isthmus. Note poorly marginated liver metastases (+). **Origin:** Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Unenhanced (a) and post-contrast (b) images show size reduction of the renal carcinoma arising from the horseshoe kidney isthmus, with persistent speckled calcifications (arrow in a). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Unenhanced (a) and post-contrast (b) images show size reduction of the renal carcinoma arising from the horseshoe kidney isthmus, with appearance of delayed perfusion in the right renal moiety. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: The liver metastases (+) are reduced in size, better demarcated and markedly hypoattenuating indicating positive response to treatment. Origin: Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
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Description: The liver metastases (+) are reduced in size, better demarcated and markedly hypoattenuating indicating positive response to treatment. The renal carcinoma arising from the horseshoe kidney isthmus decreased in size. Origin: Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Multiplanar contrast-enhanced images (f..h) show persistent neoplastic invasion (arrows) of the duodenal lumen (*) at its third portion. The liver metastases (+) are reduced in size, better demarcated and markedly hypoattenuating indicating positive response to treatment. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Multiplanar contrast-enhanced images (f..h) show persistent neoplastic invasion (arrows) of the duodenal lumen (*) at its third portion. Note the reduction of the anteroposterior diameter of the caudal portion of the tumour. Origin: Tonolini M, Department of Radiology, "Luigi Sacco" University Hospital – Milan (Italy)
Description: Multiplanar contrast-enhanced images (f..h) show persistent neoplastic invasion (arrows) of the duodenal lumen (*) at its third portion. Note appearance of hydrenephrosis (arrowhead) and delayed perfusion of right renal moiety. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Note appearance of hydronephrosis and delayed perfusion of right renal moiety. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)