Neoplasia in horseshoe kidney - A case report

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Abstract

Introduction. Horseshoe kidney is a rare pathology that could be found singular or in association with other malformations or sometimes with malignant disease. The risk to develop stone disease is higher in a horseshoe kidney then in a normal one. Renal carcinoid tumors, Rokitansky syndrome, ano rectal malformations and neural tube defects are just other frequent associated disease. As its name suggests horseshoe kidney is composed of two renal units fused in 95% of cases at the inferior pole and the rest at the upper pole. The isthmus may be vascularized, or not. Topographically the renal vasculature is highly respected for both renal units, the real problem of the isthmus is that it has an aberrant vessels arrangement, where without a viable approach imaging mean, the risks of bleeding is very high.
Material and method. We are presenting a 52 year old patient, which has came to our clinic accusing diffuse pain at the left lumbar level without any complaints or symptoms that could require a routine check. We will focus on the peculiarities of surgical approach to this kind of disease and we will also make a discussion on the most frequent renal malformation.
Conclusions. Horseshoe kidney is one of the most common congenital anomalies which represent a therapeutic challenge in terms of both minimally invasive and classic approach. The peculiarities of vascularity make it difficult to approach such a type of kidney for both extracorporeal lithotripsy as well as for open surgery.

Key words: horseshoe kidney,surgical treatment

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Introduction

The horseshoe kidney is the most common renal fusion malformation, which consists in the union of the lower poles of the kidney by a parenchymal or fibrous isthmus. The incidence of horseshoe kidney is about 0.25% in overall population, representing 1:400 people, with a man:woman ratio of 2:1 [1,2]. Embryology, horseshoe kidney is formed due to an excessive angulation of the longitudinal axis of metanephrogene blastemas, with the fusion of the two lower poles during the 4th to 8th weeks of uterine life, during the process of ascension towards the upper abdomen [1-3]. Recently it was postulated a new theory that polar fusion is the result of a teratogenic process, but so far it has not been supported by clear evidence [2]. In most cases the ascension of the horseshoe kidney stops at the lower lumbar or pelvic level, because of the block created by the emergence of the inferior mesenteric artery, which prevents the completion of this process. It is also noticeable an anterior positioning of the renal hila, with the calyces oriented toward the posterior plane. The two ureters follow a path above the inferior poles and the isthmus of the two kidneys, which predisposes to complications such as hydronephrosis, infections and stone formation [1-4]. Due to the anterior location of the kidney to the aorta and inferior vena cava, renal vasculature of horseshoe kidney is not uniform as in a normal kidney, but is formed of a radial network of blood vessels of different size. The evolution of horseshoe kidney is mostly asymptomatic, its diagnosis may be pure coincidental or when the complications emerges. [1-3]

Case presentation

Patient E. L. 52 years, resident of an urban area, married, smoker and moderate alcohol consumer, is hospitalized in our clinic accusing diffuse pain at the left lumbar level.

The disease develops for about six months, the patient being at the first presentation in a medical service. Patient's history notes for hypertension and an upper gastrointestinal bleeding episode in 33 years. On admission, the patient's blood count revealed a hemoglobin of 20.6 mg/dl with normal other laboratory values.

The CT examination of the abdomen and pelvis with contrast revealed a tumor in the upper pole of the left side of the horseshoe kidney and a richly vascularized isthmus in front of the aorta.

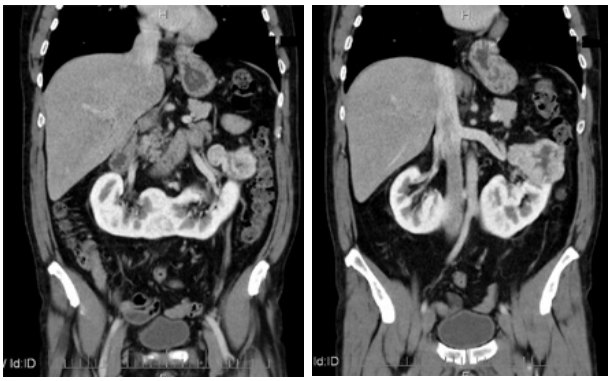


Fig. 1, 2: CT aspect, it can be observed the intense intake of contrast at the isthmic level alongside the tumor of the upper pole

Under general anesthesia the surgery is performed and is practiced left hemi-nephrectomy by peritoneal approach and also the excision of a portion of the isthmus which, as noted in the tomographic examination, was well vascularized. Postoperative evolution was favorable, so that the patient was discharged at 7 days after surgery with near normal renal function.

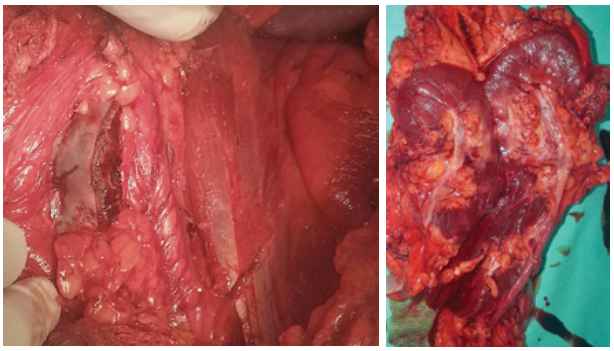


Fig. 3,4 Intraoperative aspect: it can be seen clearly the residual isthmic region and the great vessels posteriorly. The left kidney specimen removed with ureter and isthmic region at the inferior side of the photo.

Hystology examination revealed renal cell carcinoma of the upper pole of the left unity, with Fuhrman 2 nuclear grade.

Discussions

Although it was first described in 1521 by Jacopo Berengar da Carpi, horseshoe kidney remains until date an enigma for modern urology. In addition to the associated congenital anomalies, both systemic and genitourinary, it was observed that there is a higher incidence of horseshoe kidney in chromosomal syndromes [1, 2, 4-6]. The tumor pathology which occurs in horseshoe kidney is rare, currently being quoted about 200 cases of this type in literature [7, 8]. Howev-

er tumor incidence is 3-4 times higher in patients with kidney malformations than in normal ones, and it can occur both primary and secondary to complications of urinary stasis [9].

A feature of our case is the poorly symptomatic evolution of both the renal malformation and tumor formation, which was diagnosed by CT imaging. This investigation is very useful alongside MRI and angiography, in order to visualize the blood supply of the horseshoe kidney, which may originate from both the aorta and other large caliper vessels in contact with the kidney (mesenteric arteries and iliac) [3]. In our case there was a vascular anomaly rarely described in literature [5], that the main vascular sources were emerging from the aorta, but they were irrigating very well the isthmus region, where they were branching to supply the two renal hemi-units. In this case, given the risks, the operators proposed an open surgical approach thro a midline incision. For a better control of bleeding at the isthmus level the aberrant vascular sources were intercepted initially.

Among the known tumor types, the most common in this kind of malformation is the renal cell carcinoma, with an incidence of 47%. Other rare tumors are more common in patients with horseshoe kidney than in other patients [5, 10]. In this case, the tumor type and degree of differentiation had a role in the evaluation of postoperative survival and follow-up of subsequent therapeutic management. Although it is very rarely documented in literature, the local recurrence has a rate of about 1.8% and also the counter side recurrence, has a rate of 1.2% [10].

An alternative to open surgery is the laparoscopic approach, but in tumors occurred on horseshoe kidney, the indication for a laparoscopic approach is limited by the kidney's degree of rotation and vascular origin sources [8, 11]. Another therapeutic method that can be considered is the selective embolization, which it's not routinely recommended for all patients, but only in cases where open surgical approach is too risky or in patients who have a contraindication to surgery [10].

In conclusion any disease occurred on a horseshoe kidney must be carefully investigated before surgery, in order to asses properly the renal topography and anatomical peculiarities. Publications in literature can give us some necessary information for the investigation of this kind of pathology, but we must not forget that in every case of malformation, there are patient specific peculiarities. Before planning a surgery on a horseshoe kidney, it should be kept in mind that it may occur intraoperative complications which may require the resection of both renal unities. Therefore, the patient should be well informed before the operation on all the possible complications that may arise during the surgery and post operatory.

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