

Nephroblastoma in Horseshoe Kidney A Rare Combined Entities (About 2 Cases)

Driss Hanine*, Zakaria Aboulam, Meryam Ramzi, Rachid Oulahyane, Mounir Kisra

Visceral Pediatric Surgery Department « A » Children Hospital Rabat - CHU Ibn Sina, Morocco

*Corresponding author

Driss Hanine

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Abstract: The horseshoe kidney is the merger generally lower pole of both kidneys by a bridge mesenchymal or fibrous infrequent, the severity of this congenital anomaly is the high incidence of associated renal pathologies. The authors report a case where you can combine horseshoe kidney and renal tumor malignancy, the aim is to draw attention to the diagnostic process in this case at the end to ensure optimal care. The first child aged of 6 years and half, consulted for abdominal pain symptomatic paroxysmal Rebels associated with abdomino-pelvic mass median. Ultra-sound and an assessment was made suspect CT digestive lymphoma or neuroblastoma. The indication for surgical biopsy was asked. Histological study had concluded nephroblastoma. A new assessment including Doppler ultrasound imaging and angio-scanner was performed. The diagnosis of nephroblastoma in a horseshoe kidney was retained with pulmonary metastasis. The second case, we had a child aged of 2 years and half, consulted for abdominal mass of chance discovery, whose imaging objectified nephroblastoma in a horseshoe kidney. Surgery for both children consisted of a complete and economical excision respecting the healthy renal parenchyma. Uncommon disease, his ignorance little affect pejorative diagnosis is based on imaging quality when in doubt scanned biopsy-guided posterior approach and fine needle is possible. In our first case, our surgical biopsy induced transformation stage; radiation is therefore necessarily associated with surgery and chemotherapy.

Keywords: malformation, horseshoe kidney, Kidney tumour, Nephroblastoma.

INTRODUCTION

Kidney tumors are one of the most common groups of solid tumors in children; since the beginning of the 1980s, the nosology of these tumors has progressed a lot, thanks to the work of the large series brought together by the National Wilms Tumor Study (NWTS) for North America and the international pediatric oncology society for Europe ; as well as the progress of molecular genetics that has upset the understanding of these tumors.

Nephroblastoma accounts for 6-8% of pediatric malignancies and 90% of solid kidney tumors in children. This tumor affects mainly the child between 1 and 5 years (average of 3 years and half), without clear predominance of sex.

The horseshoe-shaped kidney corresponds to an ectopic situation of the two kidneys united at their upper or lower poles by a fibrous or mesenchymal bridge. This anomaly of seat and shape is seen in 1/400 birth, in boys more than girls. It can constitute a non-pathological anatomical variant, as it can be associated in a large number of cases with a pathological state.

The risk of developing a tumor in a horseshoe kidney is higher than in a normal kidney. The presence of a tumor in a horseshoe kidney is an unusual situation in the sense that this kidney most often has anatomical relationships and mainly vascular abnormalities that will act on the management of this pathology. This explains the variability of therapeutic proposals and the absence of codified protocols. The management must be multidisciplinary and must respond to two contradictory imperatives, firstly to be curative and secondly to preserve the maximum of renal parenchyma to prevent progression to renal failure.

In our study, we report two cases of nephroblastoma on kidney horseshoe in a child of 2 years and a half and another 6 and a half years. The purpose of this study is to analyze the characteristics of such a localization, especially radiologically, and to discuss the choice of the surgical technique.

CASE REPORT

We report in our study 2 cases of renal nephroblastoma on kidney in children.

Case 1

Six and a half year-old male, with a history of surgery for appendicular peritonitis at the age of 3 years, admitted for painful abdominal mass discovered by the mother after episodes of food vomiting that had been evolving for a week in the context of apyrexia and conservation of the general state; and in whom the clinical examination finds a scar of medial laparotomy as well as a distended abdomen with a hard mass of regular contours palpable under the umbilic.

The child underwent an emergency ultrasound showing horseshoe kidneys with presence of a medial pre-vertebral lesion process of the sub-renal stage in close contact with the parenchymal bridge (Isthmus).

The patient then benefited from a CT scan (Figure 1) which objected to a retroperitoneal starting point process, with regular polylobe contours, not including major cystic and fleshy double component vessels without calcifications; measuring 14x08x12 cm with no ADP.

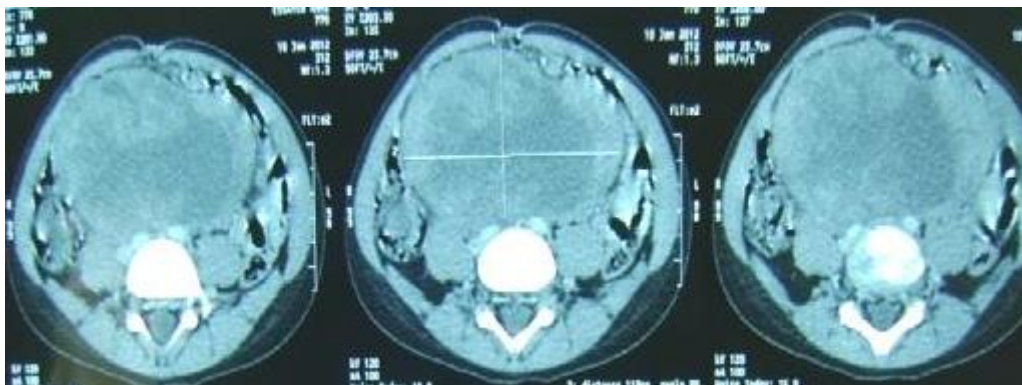


Fig-1: CT image of the 1st patient showing a heterogeneous tissue mass with necrotic rearrangements evaluated at a volume of 1346.8 cm³, well limited and with a retro-peritoneal starting point

The diagnosis at this stage is not precise between a neuroblastoma, lymphoma or other malignant kidney tumor which led to the realization of a biopsy having objectified metastatic mesenchymal nephroblastoma and leading to a stage III iatrogenic manner.

The extension assessment had shown to CT thoracic diffuse parenchymal lesions of secondary appearance (Figure 2). The child thus received 6 courses of neoadjuvant THC. The total regression of secondary pulmonary lesions was recorded as well as a regression of 30% of the volume of the renal tumoral process with the VCI and the renal veins being permeable.



Fig-2: CT image of the first case showing diffuse secondary parenchymal lung lesions

The child was operated on after recovery of the old median incision. The exploration having found a mass appended to the isthmus of 5 cm of well limited size respecting the two kidneys united in horseshoe by their lower poles.

Complete resection of the mass with a safety margin of 1.5 cm by passing in a macroscopically healthy tissue while respecting part of the isthmus as

well as a resection of the parietal path of the surgical biopsy (as described in the literature).

The pathological study concluded with mesenchymal nephroblastoma, classified as stage IV according to SIOP 2001 [1,2]. The patient then benefited from postoperative chemotherapy and adjuvant radiotherapy with good progress.

Case 2

Male child of 2.5 years old, without any particular pathological antecedent admitted for abdominal mass of the right hypochondrium evolving since 1 month with progressive increase of volume and in whom the clinical examination finds a distended abdomen with hard mass on palpation from the right hypochondrium to the right iliac fossa, as well as bilateral testicular ectopia.

The child underwent an abdominal ultrasound demonstrating a superior right polar lesion process, tissue echo, structure, oval, well-defined, echogenic, homogeneous, with absence of calcifications within it,

measuring 96x73x174mm and exerting a mass effect on calyx cavities that are dilated.

A CTM (Figure 3) showing horseshoe kidneys was completed with a large process at the expense of right hypodense hypodense tissue, with clear margins containing areas of necrosis and enhancement after PDC injection, measuring 100x102x126mm. This process invades the antero-internal lip of the contralateral hemi-kidney downwards and inwards and then massively the right kidney backwards which is enlarged with dilation of the calyx cavities that are crushed. It invades the prevertebral renal parenchymal bridge with persistence of a thin parenchymal tongue. The contralateral left renal pedicle is visible and permeable with no individualization of the right renal pedicle.

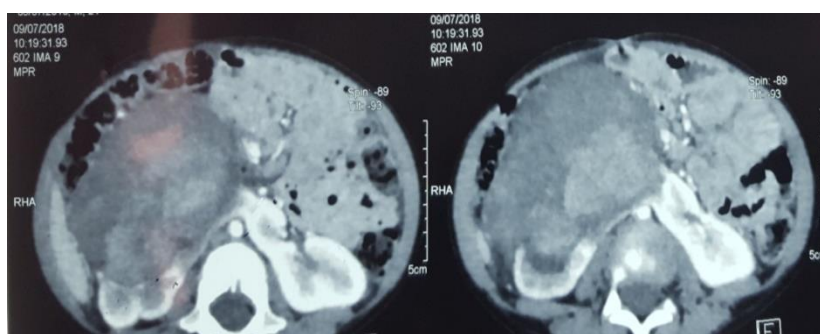


Fig-3: Scanning image of the 2nd patient objectifying horseshoe kidneys with a bulky process at the expense of the right hemi-kidney

The extension assessment being without particularities. After receiving 4 cures of adapted CTH, the patient was operated with the exploration several vascular connections between the mass and the VCI as well as the AMI that was ligated and sectioned, spotting of the left kidney which is without abnormalities as

well. that his pedicle which was put on lake, spotting of the right renal pedicle which is invaded and his ligation section, complete resection of the mass after his dissection compared to the different planes as well as the resection of the lower bridge linking the 2 kidneys with a margin healthy (Figure 4).

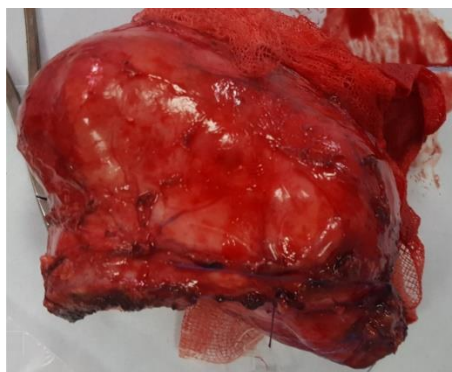


Fig-4: Tumor mass completely resected to its fresh state

The anatomo-pathological study having concluded in a stage I nephroblastoma. With a follow-up of 6 months the child did not present a recurrence with good clinical evolution.

DISCUSSION

Nephroblastoma or Wilms tumor is one of the most common solid tumors: 6 to 12% depending on the

country and the epidemiological methods. It is a tumor of infancy, which affects the child under 5 years as the case of our two patients. It is exceptional in adults.

Thanks to therapeutic trials since the 1960s, the cure rate increased from 20% in 1920 to more than 90% in 2000. [3,4]. The discovery of a tumor process on a kidney in horseshoe (RFC) allows the authors to

take stock both on the peculiarities of this association and on the difficulties of diagnosis and treatment.

The incidence of certain types of cancer seems to be increased by this anomaly (nephroblastoma and carcinoids). The diagnosis of RFC is sometimes difficult, imaging, especially arteriography, being essential, both to confirm the morphological anomaly, to situate the tumor process and to define a surgical strategy by visualizing the vascular pedicles which are eminently variable.

Surgery of excision, adapted to the tumoral localization, represents the essential therapeutic time [3]. The aim is to evaluate the diagnostic and therapeutic difficulties that persist and compare our two cases reported with data from the literature to correct our attitude. The incidence of RFC is estimated at 0.25% of the general population with a frequency at 1/1800 or 1/1400 [5,6].

There is a clear male predominance with a sex ratio of 2 as the case of our two cases. All urological conditions can be seen but the incidence of some of them seems higher than on normal kidney. The first cancer sighting on an RFC was reported by Hildebrand in 1895, and since then the reported cases are relatively rare. All histological types have been described but with varying incidences. Thus nephroblastomas (Wilms tumors) represent 14.3%.

The incidence of Wilms tumors developed on RFCs appears to be above average. As a result, the discovery of a kidney kidney in a child seems likely to require special monitoring related to this risk factor. One-third of patients with a horse-shoe kidney are asymptomatic. The kidney horseshoe is however a source of complications: there is also a syndrome of the junction pyelo-ureteral (JPU) in 20% to 30% of cases.

The latter is in fact often linked to a high implantation of the ureter on the pelvis and / or compression by an abnormal vessel, and / or at the crossing of the ureter by the renal isthmus. However, care should be taken not to take an apparently dilated bassinet as an obstruction because of its internal orientation. Thus, urinary infections can occur in 30% of patients and stones in 20% to 80% of them. 5% to 10% of the horseshoe kidneys can be revealed by the detection of an abdominal mass [3, 7-10]. However, all of these data could not provide a guiding element allowing the distinction between renal nephroblastoma on the horseshoe kidney nephroblastoma on anatomically normal kidney [11, 12].

Clinically

The mass being asymptomatic as master symptoms 46% as for the 2nd case and abdominal pain in 2nd place at 23% as for the 1st case. The diagnosis period for our patients being 1 month for the 1st and a

week for the 2nd case. The average time in literature is also 1 month

Imagery occupies a prominent place in the positive diagnosis of nephroblastoma on a horseshoe kidney. It also allows the assessment of lesions, the pre-therapeutic assessment, the follow-up as well as the screening of children at risk and their surveillance [12-14]. It is therefore essential to obtain quality imaging in order to establish this diagnosis with certainty. However, one must know the limits of imagery and never interpret it in isolation. It is always necessary a radio-clinical and histological confrontation.

Abdominal ultrasound alone usually provides the diagnosis of the renal tumor, it most often allows to visualize an abnormality of the position of the kidney and to highlight a possible parenchymal localization. It does not always allow to analyze the isthmus portion. This is indeed possible only 27 times out of 34 for Strauss, the non-definition of the lower pole of the kidney making suspect an RFC [15]. It shows the tumor, specifies its retro-peritoneal seat intra-renal, shows its limits and relationships with neighboring organs, it specifies its echogenicity, solid, cystic or more often mixed. It provides information on the existence of haemorrhage, necrosis and / or calcification intra tumor, it specifies the size of the tumor in three dimensions, and therefore the volume and precise the state of the liver.

Doppler ultrasound can tell us about the vascularization of the kidney and can show the renal artery and inferior vena cava. Abdominal CT is sometimes essential before beginning the treatment to ensure a higher degree of confidence in the diagnosis in the litigious cases, it specifies the situation of the kidneys, the structure of the isthmus and the dimensions of the tumoral process, makes it possible to measure the tumor in its 3 dimensions, seeks a thrombosis of the IVC and allows the study of the hepatic parenchyma.

If the clinical picture is painful, it is always necessary to think of an intra-tumoral haemorrhage or a tumoral rupture, the 3 stages of rupture can be seen:

- Cracking: Peri-tumoral image in crescent, spontaneously hyperdense
- Per peritoneal rupture: spontaneously hyperdense perirenal area
- Intra-peritoneal rupture: peri-hepatic fluid effusion, inter-hepato-renal effusion and in the dead-end of the Douglas.

These tumors are always chemo sensitive, it is possible to perform preoperative chemotherapy before opting for surgery as the case in our 2 patients.

The resection must be complete, however it will be necessary to visualize the healthy kidney and its

pedicle before resecting the tumor and ligate the pedicle of the diseased kidney.

In the light of these data, it is noted that patient management has complied with the recommendations of the SIOP 2001 protocol [16, 17] with good progress, a review made of abdominal ultrasonography and thoraco-abdominopelvic CT are recommended to the end of the treatment.

However, the gesture not to be done is to surgically biopsy the mass, so to the slightest doubt it is necessary to perform a percutaneous needle biopsy ultrasound or scan-guided not to change the therapeutic attitude and induce iatrogenic way a stage transformation to another advanced.

CONCLUSION

The horseshoe kidney is the most common malformation of renal symphysis. The occurrence of cancer on this type of malformation raises particular diagnostic and therapeutic problems. Imaging and in particular arteriography represent an essential step in the surgical strategy by visualizing the various morphological and especially vascular anomalies. The surgery, adapted to the local conditions of the RFC is, as in all kidney cancers, the essential therapeutic time. The therapeutic progress made by chemotherapy and the multidisciplinary management of Wilms' tumors has significantly improved its prognosis. Research is currently focused on oncogenesis and the identification of new prognostic parameters to alleviate treatment and further reduce therapeutic aftereffects.

REFERENCES

1. Beckwith Bj. Md, Hon Trocpath. Revised SIOP working classification of renal tumors of childhood. *Med pediatri oncol.* 2002 ;38 :77-78.
2. Vujanic, Md, Gordan M. Bengt S. Revised international society of pediatric oncology (SIOP) Working classification of renal tumors of childhood. *Med pediatri oncol.* 2002; 38 :79-82.

3. Faucompret S, Farthouat P, Deligne E, Louis C, Breda Y. Cancer sur rein en fer a cheval. A propos d'un cas de decouverte fortuite. In *Annales d'urologie* 2002 Jan 1 (Vol. 36, No. 2, pp. 81-86). Elsevier Masson.
4. Breslow Ne, Olshan A, Beckwith JB, Green DM. Epidemiology of Wilms tumor. *Med Pediatr Oncol* 1993;21:172-81.
5. Campbell M. Anomalies of the kidney. In: *Urology*. Philadelphia: WB Saunders. 1970.
6. Segura JW, Kelalis PP, Burke EC. Horseshoe kidney in children. *JUrol.* 1972;108:333-6.
7. Nagar RC, Sanwal BL. Squamous cell carcinoma in a horseshoe kidney. *J Indian Med Assoc.* 1989; 87 : 14-6.
8. Hayashi T, Fukuda H, Hagiwara T, SAKAI K. Renal celle carcinoma in a horseshoe kidney. *Hinyokika Kiyu.* 1991; 37:613-5.
9. Trulock TS, Ricketts RR, Verras A, Kim TH, Hawkins HK, Woodard JR. Wilms tumor arising in horseshoe kidney. *Urology.* 1985; 25 : 306-9.
10. Smith-Behn J, Meme R. Malignancy in horseshoe kidney. *South Med J.* 1988; 81: 1451-2.
11. Glenn JF. Analysis of 51 patients with horseshoe kidney. *N Engl J Med.* 1959;26: 684-7.
12. Sharma SK, Bapna BC. Surgery of the horseshoe kidney—an experience of 24 patients. *Aust N Z J Surg* 1986;56:175-7.
13. Lemerle J, Tournade MF. Néphroblastome (tumeur de wilms) *Rev prat (Paris).* 1993, 43, 17
14. Lemerle J, Valayer J, Tournade M. Tumeurs du rein. Flammarion médecine sciences. 1989.
15. Strauss S, Dushnitsk YT, Peer A, Manor H, Libson E, Lebensart PD. Sonographic features of horseshoe kidney: review of 34 patients. *J Ultrasound Med.* 2000; 19 : 27-31.
16. Lemerle J. Tournade MF. Néphroblastome (tumeur de wilms) *Rev prat (Paris).* 1993;43:17.
17. lemerle J, Valayer J, Tournade M. Tumeurs du rein. Flammarion médecine sciences. 1989.