HORSESHOE KIDNEY: A CASE REPORT AND REVIEW OF LITERATURE.

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ABSTRACT

The horseshoe kidney (HK) is a very rare renal malformation. It designates a transposition by fusion of the parenchymas of the two poles of the kidney. Its discovery is incidental to an autopsy. However, it is most often asymptomatic. Thus, we report a case of horseshoe kidney accidental discovery in a 58-year-old man. We take this opportunity to review the literature on embryological, clinical and therapeutic aspects of this malformation, which is one of the renal ectopies diagnosed in sub-Saharan Africa.

Key words: Kidney, Horseshoe, malformation.

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INTRODUCTION

The horseshoe kidney (HK) is a renal malformation secondary to parenchymal fusion of two homologous poles of both kidneys during their ascent in utero. It is a renal malformation frequently found in autopsies but currently, more often, by medical imaging (ultrasound, computed tomography and magnetic resonance imaging) [13,14,51]. Its incidence is 1 case per 400 to 600 births [3,16,24,33]. Clinically, HK is often incidental; in other words, it is most often revealed by a contusion secondary to a trauma of the lumbar spine whether anterior or posterior [18]. HK is also the subject of many diseases such as lithiasis and urinary tract infections [30, 59], tumors [48] and especially cysts [26]. We report a case of HK accidental discovery and then conduct a literature review describing its anatomo-radiological aspects.

CASE REPORT

This was a 58-year-old male subject who was consulting for abdominal pain and dorso-lumbar spine. This symptomatology had been evolving for a year; it occurred spontaneously, intermittently and without any notion of triggering factor or irradiation. This pain improved without or under analgesic treatment. We do not know any particular family or personal pathological history. The physical examination revealed a subject in good general condition whose urination was normal. The lumbar pits were without special features as well as the upper and middle ureteral points. There was no lumbar contact or renal bloating. The hernial areas were free. There was no vesical globe. The abdomen was soft. The rectal exam showed a flat prostate. CBC, creatinine and total prostate specific antigen PSAT were normal.

Uro-CT showed a fusion of the two kidneys (horseshoe kidney with lower concavity). The parenchymal isthmus was located in front of the large vessels and at the height of
L1. Both kidneys secreted in the same time. The renal cavities were not dilated. There was no pyelo-ureteral junction syndrome or renal lithiasis or other associated abnormalities. Lifestyle advice was given in part because of the complications of trauma. Symptomatic treatment was also instituted. The patient, residing in Mali, was subsequently lost sight of.
DISCUSSION AND LITERATURE REVIEW


In Senegal, the incidence of this renal abnormality is not known. In contrast, its annual incidence in the general population varies by author: 1/353 cases for Harper [38], 1/400 cases for Glenn [29] and O’Brien [56], Bell [6] and Cox [17], 1/425 cases for Campbell [16], 1/468 cases for Nation [50], 1/500 cases for El Badoui [23], Doukkali [21], Benchekroun [5], 1/600 Moore [49] and Saunders [71] in 1977 and 1/800 in adults and 1/400 in childhood [52, 66]. However, this frequency would be increased due to the clinical latency of this renal abnormality [77]. For Gutierrez [37], there are one or two cases of horseshoe kidney observed on 200 UV performed. Ilmura [40] estimates that the incidence of HK is higher in men than in women. For Iwanaga [42], this incidence is twice as high for men as for women. Some authors in all HK series of studies have observed that this abnormality is two to three times more common in boys than in girls [2, 5, 8]. El Aasraoui [26] notes cases, three boys and one girl in her report. This same observation is reported by Fekkak [29] with a sex ratio of 2 as well as Atala [2] and Ilmura [40]. Campbell [16] observes in his series that the horseshoe kidney is twice as numerous in children as in adults during autopsies. He found 61 of 19,046 autopsy cases in children and 61 of 32,834 in adults. These results are confirmed by the work of Nelson [52] and Stroosma [70] which note an incidence of 1/600 to 1/800 in adults and 1/400 in children respectively. For Sturrock [65], the incidence of this malformation as well as any other malformation should be significant when considering stillbirths and children who died in the perinatal period. In our observation, the subject is male.

The HK can remain asymptomatic and be discovered fortuitously either during a systematic assessment or during a renal trauma. Elsewhere, it may be revealed by pain at the extension of the trunk (5 to 10% of cases) [26]. The pain caused by the hyper-extension of the dorso-lumbar spine is often suggestive of HK [26]. This pain reflects the compression of the coeliac plexus by the isthmus of the HK. This is the case in our observation although these abdominal and spinal pains are not well systematized. Digestive disorders can sometimes direct investigations to this abnormality [26] and abdominal masses.

In fact, it is the complications that reveal the most HK and require surgery [29]. Conditions such as pyelo-ureteral junction syndrome, obstructive urinary lithiasis (especially renal), upper urinary tract infections (acute and chronic pyelonephritis, pyonephrosis, renal abscess, perinephric phlegmon) [30, 57, 59] and tumors of the superior excretory pathway may be circumstances of discovery of a HK due to noisy symptomatology. We note no complication in our observation which is supported by data from biology, bacteriology and medical imaging (Uro-CT).

However, it is necessary to note the discoveries per-operative and during the autopsies. Prenatal diagnosis is possible during the first trimester of pregnancy using high frequency transvaginal ultrasound [9].

The HK can be accidentally discovered either during a systematic check-up or during a radiological or ultrasound scan, computed tomography for another condition [7].

observation describes this modality. Ultrasound of the urinary tract allows to note the verticalization of the kidneys. However this modality is not realized in our context. Ultrasound can be used to diagnose urogenital disorders and abnormalities associated with HK. Intravenous urography (IVU): the lower poles of the kidneys are close together, the caliceric tree is oblique at the bottom and inside.

Uro-computed tomography (Uro-CT) is used to diagnose the HK and assess the functional value of the parenchymal bridge [48] and to diagnose other conditions or related malformations [60]. The Uro-CT is efficient. It provides useful, detailed and sufficient information about the anatomy of the RFC and its vascular variations and the conditions of associated conditions as well as the anatomical relationships of the HK with other nearby organs [55]. It is essential in case of HK disease: diagnosis and monitoring.

Resonance imaging (MRI), like CT, is an excellent imaging exam for diagnosing HK. It provides detailed information about the anatomy of the HK, its vascularization and related conditions, and anatomical reports.

Renal scintigraphy with DMSA (dimercaptosuccinic acid) or DTPA (diethylene triamine penta-acetic acid) marked with technetium-99 with forced diuresis stimulation (or Lasilix test) is used as a means of functional exploration to evaluate the permeability of the urinary excretory tract and to assess its relative functional value. This contributes to the diagnostic orientation and improvement of the therapeutic strategy of obstructive forms of HK [10]. However, many reported cases of HK [45] are incidentally diagnosed on bone scintigraphy with technetium-99.

In general, kidney abnormalities are classified in six categories by Campbell [in 68] in 1986: abnormalities of number, volume and structure, situation, form and fusion, rotation, vascularization. The fourth category of Campbell was studied in 1930 by the Russian Ognew [58] who established a simplified morphological classification. Its results have been summarized in five types by Matsumoto [47]: Type A(a): fusion of the upper poles, Type A(b): fusion of the lower poles, Type B(a): fusion by fibrous tissue, Type B(b): complete fusion of the kidneys, Type B(c): median fusion (by the medial edges). The HK case reported in our observation is type A(a). The HK has a diversified shape. This shape variation could explain how the two kidneys fuse together. Thus the reversed U, L, or U and L shapes are often described.

Sturrock [65] had found during the dissection of two cases of horseshoe kidneys in an ectopic position with an inverted L shape: the left kidney forming the vertical and the right kidney the horizontal element for both kidneys. The left kidney was in a normal anatomical condition for the first and the left kidney was in a lower anatomical condition for the second. Our subject has an inverted U-shaped HK.

Many authors establish different types of RFC vascularization including that of Graves [34] in six types. Thus, the variations from the origin of the renal artery to the lower segment of the kidney according to Graves [34] arise as follows: Type A: Hilum; Type B: From the main stem to its junction with the aorta; Type C: From the aorta to the main stem; Type D: From the aorta, but also giving rise to the testicular or ovarian artery; Type E: The artery of the lower segment comes from the aorta at a certain distance from the main stem; Type F: The lower segment of the artery and its posterior branch develop separately from the aorta. In our subject, the type of vascularization is not specified.

The HK integrates the general framework of a polymalformation syndrome. As a result, many abnormalities are associated with it. Thus, we can have: Urogenital

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abnormalities[14]: pyelo-ureteral junction syndrome 15 to 80% of cases, retro-cave ureter 0.1 to 2.8% of cases, ureteral duplicity, testicular ectopy 6.9% of cases, cryptorchidia 4%, hypospadias 2.8 to 5.7% of cases, unicorn uterus 5.4% of cases.

Cardiovascular abnormalities: the risk of congenital heart disease is multiplied by 60 in case of HK according to some authors [13,19]. Ichikawa [41] demonstrates that major venous abnormalities including a double inferior vena cava, a retroaortic left renal vein and a circumaortic left renal vein are frequently associated with HK [41]. However, no exploration for anomalies associated with the HK is carried out in our context. Sturrock [65] observes during a dissection of both kidneys in horseshoe, in addition to vascular abnormalities, a case of associated abdominal aortic aneurysm although observations of aortic aneurysm in cases of HK are rare [17]. For Olsson [54], HK is often associated with cardiovascular abnormalities.

In our observation, we note no etiological factors although some explorations are not carried out. However, the literature reports that this anomaly occurs between the fourth and sixth weeks of development (some authors) [35, 75] and others until the ninth week [25, 64]. It is due to many causes such as: position factor and fusion abnormality, abnormalities of metanephric cell migration, intra-uterine factors (environment and teratogenic exposure) and the association of this abnormality with genetic factors and chromosomal abnormalities [74].

No clear genetic cause of HK is described in humans however a number of regulatory steps in kidney development are not completely elucidated and may therefore offer a future insight into etiology [62]. In the animal model, notochord is involved as a factor determining the position of the metanephrogenic tissue. HK is dominant in male subjects [33]. Family cases of HK are reported in a father and son [62] and monozygous twins [61]. It was noted in a family that three siblings have a HK and their mother has a rotation anomaly [22]. This finding is circumstantial evidence that genetics can play an etiological role. However, there are also case reports of monozygous twins where only one child is affected [46]. It is worth noting the existence of other associations of genetic abnormalities to the HK: trisomy 13 and trisomy 18 which respectively cause PATAU syndrome (combination of renal polykystosis, HK and pyelo-ureteral junction syndrome, cryptorchidism and hypospadias etc.) [12,30] and in Down syndrome, the incidence of HK would be less than 1% [67]. In Turner syndrome, HK occurs in 14% to 20% in patients [63] and 15% in women [20]. Grainger [32] also reports in its series, the association of congenital anomalies and especially of Down syndrome 18 and Turner’s syndrome at the HK.

Abnormalities of caryotype: tetrasomia 8p: 46XX/47XX+i is reported in the literature [43]: it performs a clinical picture combining skeletal abnormalities, rectal atresia, inverted situs, RVU, facial abnormalities and HK.

During migration, from the sacred region, the two metanephrogenic blastemes can come into contact with each other through one of their poles (usually the lower poles) [69]. The HK has a higher concavity when the fusion is between the two lower poles of the two kidneys and lower concavity if not. Our case describes this second possibility. The ascent of the horseshoe kidney is slowed down by the lower mesenteric artery (fork) [27]. However, in our case, no exploration is done for this cause. During this ascent of the kidney, flexion or caudal rotation should be sufficient to cause fusion. Fusion abnormalities that can occur symmetrically
or asymmetrically suggest other causes of the horseshoe kidney. The asymmetrical or symmetrical character of fusion would be the result of differential displacement of the two renal masses [73]. The isthmus has varying anatomical relationships. Its middle or lateral position relative to the spine determines whether the HK is symmetrical or asymmetrical [64]. Asymmetric systems are more often found on the left (70%) than on the right (30%) [33]. In 80% of cases, the isthmus contains the functional parenchyma and can be used as a matrix for surgery.

The HK has the same clinical application area as the normal kidney despite its often lower lumbar position. It is thus that the HK has the same therapeutic modalities (classical and endoscopic surgery) as the normal kidney with some peculiarities related to the anatomy of this kidney (vascularization and renal excretion pathways and associated abnormalities). Thus these peculiarities (especially the high insertion of ureters) represent a challenge in the management of patients with symptomatic stones by percutaneous nephrolithotomy (NLPC) and extracorporeal lithotripsy (LEC) [71]. The most common treatment modalities are NLPC and LEC [1], however in many cases NLPC and LEC are contraindicated or technically difficult to achieve [36]. The lack of difficulty in fragmenting the stones is observed by Choualb [11] in the management of urinary lithiasis in patients with HK. The HK can be transplanted as a block or split in two. The separation or non-separation of the horseshoe kidney is a function of vascular anatomical variation and the donor and recipient collection system [70]. However, the results are different one year after the transplant [37]: block transplant 95.7% survival rate compared to 86.5% survival if separation [70].

CONCLUSION

The horseshoe kidney is an anatomical entity of renal malformations. It is a common fusion abnormality. It has a complex anatomy as to its location, shape, type of classification and vascularization. It is often discovered by chance. In our observation, the RFC is discovered during an assessment for abdominal pain and dorso-lumbar spine. This HK has a lower concavity: this form represents 10% of cases. Additional explorations allowing the identification of other related malformations have not been carried out in our subject. This kidney is classified type A(a). However the type of vascularization has not been specified. No etiological factors have also been determined in our context. The HK is prone to infections and trauma. Thus the prevention of these complications is based on lifestyle advice. The search for other associated malformations must be imperative given that this renal malformation fits into the framework of a polymalformation syndrome.

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CONFLICTS OF INTEREST

The authors affirmed that there are not any conflicts of interest.
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