fièvre persistante, de bicytopénie, de taux élevés de triglycéride, de ferritine et de LDH ; chez une patiente présentant des arthralgies dans ces antécédents, une lymphopénie, une protéinurie > 0,5g/24h, une péricardite et des ANN positifs. Une corticothérapie à la dose de 2mg/Kg/j a été démarrée, associée à des transfusions itératives. Une interruption thérapeutique de la grossesse a été réalisée. Mais, la patiente s'est rapidement aggravée, et a été transférée en réanimation où elle est décédée dans un tableau de défaillance multi-viscérale.

Conclusion

Le SAM est une maladie rare, en particulier chez la femme enceinte ; moins de dix cas ont été, jusque là, rapportés (1, 2). Dans le tableau 1 nous avons résumé quelques cas de SAM associé à la grossesse, décrits dans la littérature. La physiopathologie du SAM n'est pas encore complètement élucidée. Le terrain dysimmunitaire semble être responsable ; une hypersécrétion de cytokine serait à l'origine de ce syndrome. Certains auteurs ont émis l'hypothèse de sécrétion, par les lymphocytes Th1 activées, de l'interféron stimulant les macrophages. L'auto-amplification incontrôlée des macrophages va aboutir à la phagocytose des cellules hématopoïétiques et la production rapide de TNF alpha (Tumor Necrosis Factor alpha), d'où l'initiation de la cascade inflammatoire entraînant la fièvre, l'hyperferritinémie, et d'autres symptômes associés (4). Pour certains auteurs, la grossesse constituerait en elle-même un facteur de risque de survenue du SAM (5). Les modifications immunitaires et l'activation de cytokines induites par la grossesse pourraient créer un terrain propice au développement de ce syndrome chez des femmes génétiquement prédisposées (1, 6). Le pronostic du SAM est sombre et dépend de l'étiologie, la mortalité est entre 30 et 50% selon les séries (3). Ce syndrome expose au risque de mortalité maternelle par défaillance multi-viscérale (7), comme c'est le cas chez notre patiente. Concernant le fœtus, l'activation des cytokines au cours du SAM serait responsable de retard de croissance intra-utérin, de pré-éclampsie ou de HELLP syndrome (7).

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Neoplasia in horseshoe kidney: A diagnostic and therapeutic dilemma

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Renal cancer that originates in a HK is an unusual entity (1). Only 166 cases of renal malignant tumors in HKs have been reported on the international literature up to July 2006 (2). We searched the literature and found 30 other cases by June 2013. The occurrence of renal carcinoma in HKs is no higher than in the normal population (3). A HK presents a unique anatomic challenge secondary to its low fixed location, abnormal vasculature and presence of an isthmus. The blood supply to the HK is often atypical (3). It may arise directly from the aorta, the inferior mesenteric artery, the common or external iliac arteries, or the sacral arteries (4). Thus, the urologic surgeon must be familiar with the exact vascular distribution and the contribution of each vessel to the tumor supply to facilitate surgery, to create an exact strategy for the operative intervention and to prevent bleeding complications (3). An abdominal angiography/ arteriography with selective delineation of the renal vessels should be performed preoperatively in addition to a CT scan (3,5). Contrast-enhanced MR angiography improved the capability of multiplanar imaging and visualization of major segmental renal arteries, renal veins and even accessory vessels (6). Surgical procedures involved in the treatment of malignant tumours in the HK are complex and challenging, thus, surgical planning is fundamental for a successful treatment. A midline laparotomy is mostly indicated, to facilitate exposure and gain access to the often-variable vasculature, with division of the isthmus (7). If the tumor involves one kidney in a HK, the functional isthmus has to be resected along with the tumor. If the tumor arises from the isthmus, isthmusectomy with bilateral lower pole heminephrectomy is needed (5). There are few reports of laparoscopic ablative surgery in HKs, of laparoscopic and retroperitoneoscopic heminephrectomies (8,9) and even partial nephrectomy (10). From our own experience, three more patients with malignant tumours involving HKs are reviewed and their management discussed.

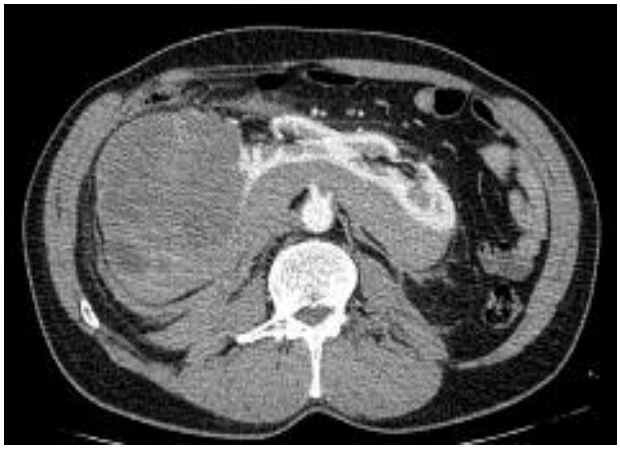
Case 1:

A 50-year-old man presented to our emergency for macroscopic hematuria after lumbar trauma. His blood pressure was 130/85mm Hg. The examination of the abdomen revealed a hard, bimanually palpable, non-tender mass, about 10 cm in size, in the left lumbar region.

CT scan revealed a HK of fused lower poles with a tumor on the right kidney. The tumor was 10,4 by 9,3 cm in size and limited to the kidney. A central scar was present in the neoplasm (Fig. 1). There was no evidence of extra-organ spread on the CT scan. Angiography concluded to double right and left renal arteries arising directly from the aorta. His routine haematological and biochemical investigations were within normal limits. Laparotomy was performed through a midline abdominal incision, the posterior peritoneum was incised from the ileocecal junction to the ligament of Treitz, and the HK was

exposed. In the right kidney, the tumor displaced the lower calices and involvement of the renal pelvis could not be excluded. There were no signs of perirenal infiltration, venous invasion or lymphatic spread. The isthmus of the HK was divided, and a right hemi-nephrectomy was performed, taking care to ligate the supplying vessels. Histology showed a relatively well-differentiated renal cell carcinoma in both of the tumors (pT3, Grade2). The patient had an uneventful post-operative course. He is presently well after one year follow-up, with a normal CT scan and renal function.

Figure 1 : Preoperative CT scan reveals a HK with a 9-cm enhancing mass in the right part.



Case 2

A 65-year-old male patient, with no significant past medical or surgical history, was referred for isolated left lumbar pain. He was afebrile and physical examination revealed a hard palpable, non-tender mass, about 8 cm in size, in the left lumbar region. Basic hematological and biochemical investigations were unremarkable. Urinalysis, urine cytology, and urine cultures were negative. CT scan revealed a HK with an 8 cm mass arising from the left kidney (Fig. 2). It revealed also a 3 cm homolateral supra renal tumor. A central scar was present in the renal neoplasm. The Angio-CT scan showed that the right kidney was fed by four-vessel supply that were originating from the aorta and has only one venous. No distant lesions were observed during staging. Through a subcostal incision, a left radical nephrectomy with isthmusectomy and left adrenalectomy were performed (Fig. 3). Parenchymal sutures were placed and the collecting system of the right kidney was closed. Bleeding was minimal and no intraoperative complications occurred. No macroscopically involved lymph node was observed intraoperatively. The renal vein was not infiltrated. On histologic examination, both tumors proved to be clear-cell-carcinomas, Fürhman grade 2 pT2aM1. A follow-up CT scan with contrast media 6 months after the operation showed a left supra renal local recurrence at the previous tumor site. He is actually under antiangiogenic therapy.

Figure 2 : Abdominal CT: HK with mass on the lower pole of the left kidney.

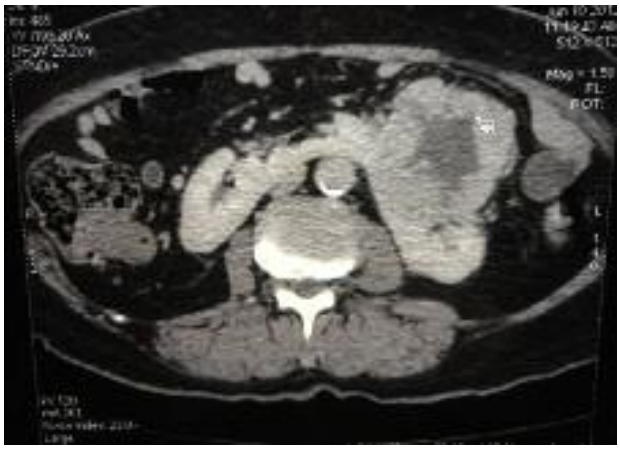
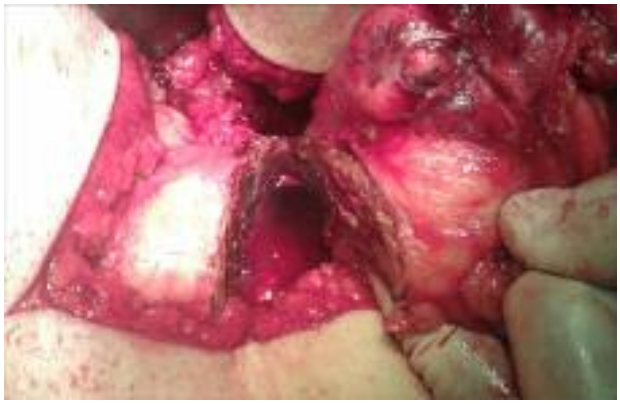


Figure 3 : Gross intraoperative photograph: isthmusectomy.

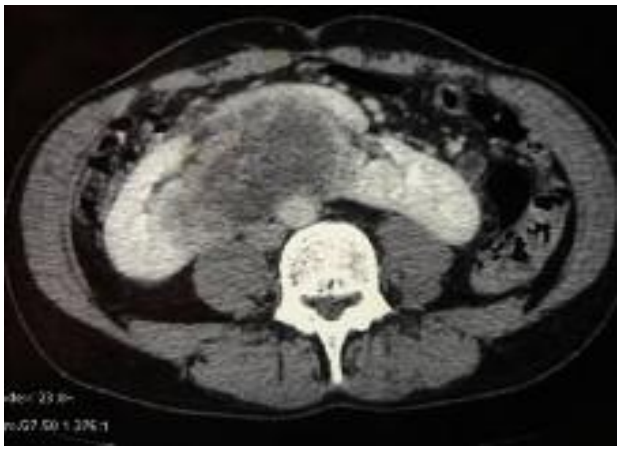


Case 3

Male, 47-year old patient, complaining of abdominal pain and constipation underwent investigation in enterology department. Physical examination revealed a hard, palpable, non-tender mass, about 10 cm in size, in the right lumbar region. His routine haematological and biochemical investigations were within normal limits. A CT scan was performed, showing a HK with a solid expansive lesion measuring 11 x 8,2 x7 cm in size, in the isthmus and the lower portion of the right kidney (Fig. 4). Multiple macroscopically involved lymph nodes were observed on CT scan. The Angio-CT scan concluded to a right and left main renal arteries arising directly from the aorta and an additional vessel to the right lower kidney portion originating from the left common iliac artery. There were two principal right renal veins and one principal left renal vein all drain to the vena cava, and an accessory left renal venous to the common iliac venous. The kidney was approached through a midline transperitoneal incision and the entire HK was exposed. We found a large tumor mass depending mainly on the isthmus and invading the aorta. A multiple and large para-aortic lymph node were also noted towards the hilum.

The radical excision was impossible and very hazardous, thus, we made only a biopsy of the tumor. The para aortic lymp nodes were also removed. The pathological examination of the surgical specimen revealed clear cell carcinoma with lymphatic metastases. Antiangiogenic therapy was planned.

Figure 4 : CT Scan showed a HK, with a large tumor in the isthmus and the lower pole of the right kidney.



Conclusion

Tumors arising within HK are uncommon and may be difficult to identify and to treat. HKs show a lot of variety with regard to supporting arteries and veins. Therefore, a selective study of the renal vessels by angiography should be performed preoperatively. Radical nephrectomy is recommended and patients need a strict follow up.

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Pilomatricome de localisation exceptionnelle avec une complication rare.

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Le Pilomatricome est une tumeur bénigne prenant naissance du follicule pileux, elle se présente comme une masse sous cutanée unique dure et mobile. On la trouve généralement au niveau des régions pilleuses du corps, surtout au niveau de la tête et le cou. Les extrémités constituent sa deuxième localisation. Le pilomatricome est fréquent chez l'enfant avec une incidence plus grande chez la fille. Depuis sa première description en 1880, il a y eu une compréhension progressive de sa présentation clinique et morphologique. Toute fois des difficultés persistent à établir le diagnostic aussi bien clinique que cytologique [1]. Nous traitons une observation de pilomatricome inhabituel par sa localisation au niveau du membre supérieur, sa taille importante (3 cm) et son apparition rapide chez une fille de 10 ans traitée chirurgicalement avec une récidive locale qui reste une complication rare.

Observation

Il s'agit d'une fille âgée de 10 ans qui a consulté pour une tuméfaction de la face antéro externe du bras droit évoluant depuis trois mois. L'examen clinique a trouvé une tuméfaction nodulaire de trois centimètres de diamètre, dure, indolore, adhérente à la peau mais mobile par rapport au plan profond. La peau en regard avait un aspect sain (Fig. 1).

Figure 1 : Tuméfaction nodulaire de trois centimètres de diamètre, dure, indolore. La peau en regard a un aspect sain.