

Renal hemangiopericytoma in 15 year old female-treated laparoscopically

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ABSTRACT

Hemangiopericytoma (HPC) is rare in the urogenital system, most frequently occurs in the pelvis, head and neck, and meninges, it is extremely rare in the kidney. US, CT, or MRI do not show any specific sign of renal HPC that might aid in the differential diagnosis with other renal tumors, that's why almost all cases are diagnosed after pathological results. The majority of cases are identified in patients with an unspecific clinical manifestations associated with hematuria or hypertension, hypoglycemia or additional paraneoplastic syndromes. Here we present a rare case of hemangiopericytoma in 15 year old female, with drug refractory hypertension.

Introduction

Hemangiopericytoma (HPC) is an uncommon perivascular tumor, first described by Stout and Murray in 1942.¹ Renal HPCs are extremely rare, with only 50 cases previously reported in the literature. HPC is classified as a soft-tissue vascular tumor arising from pericytes, cells first described by Zimmerman in 1923,² that are normally arranged around capillaries and postcapillary venules and modulate blood flow and permeability. Consequently, HPC may occur anywhere capillaries are found. We present a case of HPC in 15 year old female, who was treated by nephron sparing laparoscopic surgery.

Case report

A 15 year old female was admitted in the pediatric clinic due to dizziness and vomiting. During the exam accidentally was found high blood pressure 180-130 mmHg. Next 1 month patient had fluctuation in the BP following medical treatment. From the abdominal ultrasonography the small lesion 1,5 cm, was found on the left kidney, under the renal vessels. CT confirmed heterogenic small lesion under the renal hilum, with double pyelocaliceal system and two ureters [Fig. 1]. No tumor infiltration was found in the pyelocaliceal system, vessels and lymph nodes. Laboratory tests, including complete blood count, serum electrolytes, creatinine, and urea, were normal. The patient underwent surgery. We performed transperitoneal partial nephrectomy by laparoscopic approach [Fig. 2]. We use intraoperatively laparoscopic ultrasound to find the tumor and define its borders. A well shaped exophytic

tumor was excised completely. The surgical specimen was 1,5 cm in diameter with white color. Microscopic pathology examination confirmed the diagnosis - HPC [Fig. 3]. Next day after operation, the patient had normal BP without medications. She was discharged 3 days after surgery without complications. After the follow up of 3 months, the patient remains well with no evidence of increasing BP.

Discussion

HPC is classified as vascular tumor of soft tissues from the pericytes, modified cells of smooth muscle tissue that were described by Zimmerman in 1923.² This tumor occurs more often in younger patients with average age of diagnosis 40 years, than other types of renal cell carcinomas and there was no difference in the incidence of this tumor between man and women, also no age variation between sexes in the diagnosis.³ In the present study, the patient is 15 year old, in the cases described so far in the literature, this is the second youngest patient diagnosed with hemangiopericytoma. We found that unlike other cases where surgery was performed by conventional approach, this is the first case in which partial laparoscopic nephrectomy was performed. The most common symptoms described are hematuria, hypoglycemia and arterial hypertension, rarely can be found a painless abdominal tumor.⁴ Hematuria is most commonly seen in patients with large tumor formations infiltrating the renal pyelocaliceal system. Hypoglycemia is associated with rapid glucose metabolism by tumor, unlike hypertension is expected to be due to the production of renin by the tumor that does not appear after the removal of the tumor,⁵ which was observed in our

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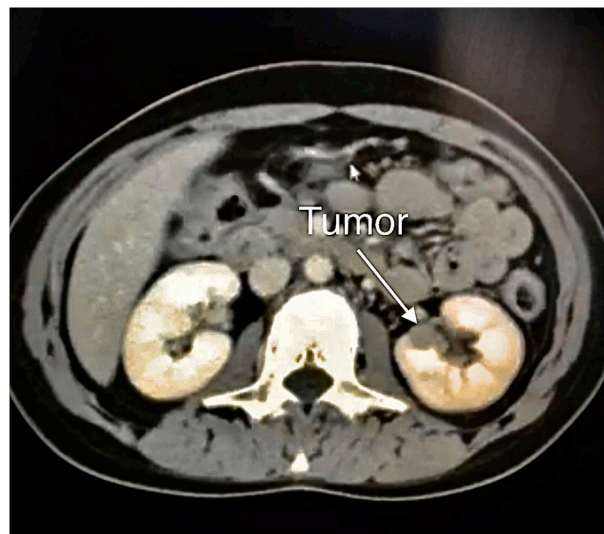


Fig. 1. CT- Arrow show well-defined, heterogeneous, enhancing small soft tissue lesion on the left kidney, approximately 1.5 cm × 1,5 cm in size.

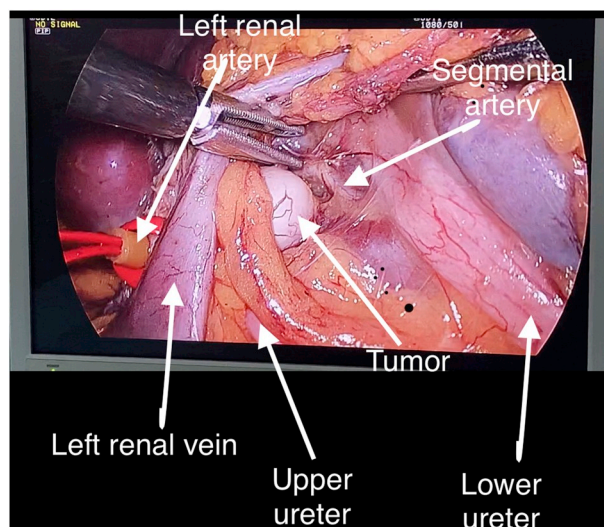


Fig. 2. The intraoperative laparoscopic view-arrows show different anatomical structures and tumor under the renal hilum.

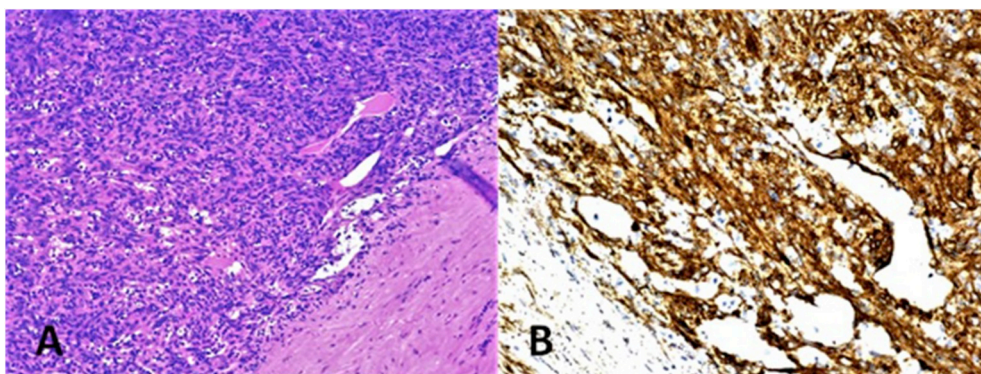


Fig. 3. HPC. (A) Histology shows round, well circumscribed tumor with thick fibrous capsule within and outside the renal parenchyma. It's composed of whorls and intersecting bundles of tightly packed spindle cells without atypia, mitotic figures or necrosis, with stromal hyalinization and focal formation of branched vessels. (B) Immunoprofile shows strong positivity for CD34 and single cells, that are positive for bcl2, CD99 and S-100 protein, Ki-67 < 10%. Tumor cells are negative for CK AE1/AE3. The renal parenchyma around the tumor is normal.

patient. US, CT or MRI do not show different signs that distinguish HPC, and that may be included in the differential diagnosis with other renal tumors. The main treatment for HPC is surgery with complete radical excision of the tumor. If possible partial nephrectomy should be performed, because it may provide improved renal function and

oncological outcomes compared with radical nephrectomy. Radiotherapy, chemotherapy or adjuvant or neoadjuvant immunotherapy seem to be inefficient to treat HPC.⁵ The diagnosis of hemangiopericytoma is usually the result of excluding the possibility of other vascular and mesenchymal tumors, according to the histological pattern and the

immunohistochemical results.

Conclusion

HPC is a rare tumor in urogenital system and patients may present with different clinical manifestation. The imaging features are not specific to make a preoperative diagnosis. Surgery is the most effective modality of treatment for the disease and with this case we showed that minimally invasive surgery including partial resection is possible, feasible with satisfactory outcomes for the patients. Accurate diagnosis of HPC is proven after surgical treatment with the immunohistochemical result.

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