Haemangiopericytoma of the Kidney

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Citation

D Sharma, A Ghosh, M Kumar, B Gupta, V Shukla. *Haemangiopericytoma of the Kidney*. The Internet Journal of Urology. 2007 Volume 5 Number 2.

Abstract

Haemangiopericytoma (HPC) is an uncommon mesenchymal neoplasm of kidney. It affects young males unlike other common renal tumors and presents with a painless renal mass occasionally associated with haematuria. A 41 year old male presented with a left side 15x15 cm abdominal mass in his left lumbar and hypochondrium region. Contrast enhanced computed tomography showed a heterogenous mass with calcification, fluid filled cystic areas and showing a mild enhancement peripherally. Patient underwent left radical nephrectomy. The tumor was well circumscribed with solid and cystic areas had caused a superior polar hydronephrosis. The histological, histochemical and immuhistochemical findings suggested the diagnosis of HPC. Patient recovered well and is asymptomatic one year after surgery.

INTRODUCTION

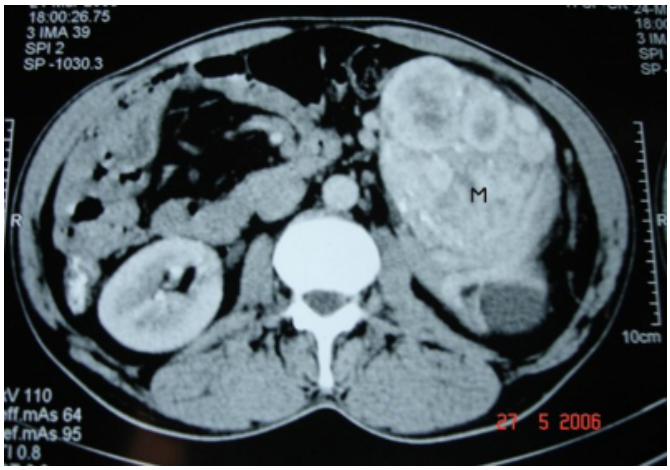
Hemangiopericytoma (HPC) is an unusual soft tissue vascular tumor featuring uncontrolled proliferation of pericytes, which are the cells spiraling around capillaries described by Zimmerman[$_1$] Though HPC may occur in any part of body; it predominantly occurs in the lower extremity and retroperitonium[$_2$] and very rarely involves urogenital system[$_1$]. We descried a case of renal HPC in a 41 year old man and present a brief review of literature.

CASE REPORT

A 50 year old male presented with a left side abdominal mass for 3 months. He had no history of haematuria or any other constitutional symptoms. On examination a large 15x15 cm bosselated mass occupying left lumbar and hypochondrium regions was found. Contrast enhanced computed tomography showed a sharply defined heterogeneous mass of size 13x9x20cms with cystic component, calcification and necrosis which had a mild enhancement at periphery, in the left renal area (Figure 1).

Figure 1

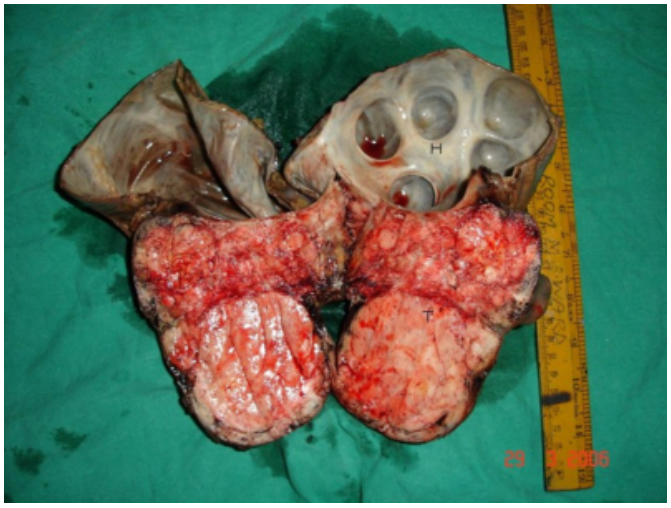
Figure 1: Contrast enhanced computed tomography showing a heterogenous mass (M) with calcification and necrosis with a mild peripheral enhancement



Suspecting a renal cell carcinoma patient was explored and left radical nephrectomy was perfommed. Grossly, tumour sized 18x12 centimeter was within the renal capsule, irregular surface bosselated, firm in consistency and had caused a superior polar hydronephrosis. Cut section showed well circumscribed, solid homogeneous, grayish white nodular mass in the renal parenchyma with cystic and hemorrhagic areas along with few satellite nodules adjacent to main the mass (Figure 2).

Figure 2

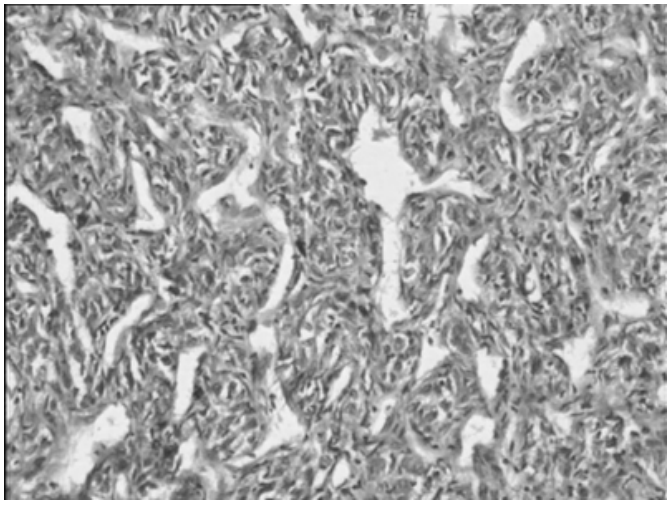
Figure 2: Gross specimen showing a well circumscribed mass (T) with cystic and hemorrhagic areas along with suprapolar hydronephrosis(H).



Histopathology of the lesion revealed a cellular, richly vascular, spindle cell neoplasm displaying moderate nuclear pleomorphism. The typical staghorn vascular spaces lined by endothelium were present, around which the spindle cells were lying peripheral to the basement membrane (Figure 3).

Figure 3

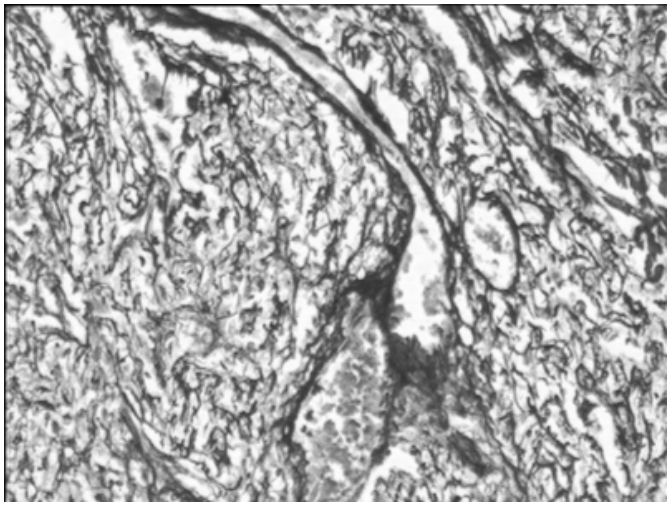
Figure 3: Characteristic vascular pattern, anastomosing vessels of varying caliber surrounded by moderately pleomorphic spindle cells (Hematoxylin and eosin x 400).



Mitotic activity of 0-2 per ten high-power fields was observed. A silver stain confirmed the extra vascular location of neoplastic cells and showed pericellular reticulin fibers (Figure 4).

Figure 4

Figure 4: Silver stain of the lesion demonstrating the extravascular location of the tumor cells and an individually pericellular reticulin (Reticulin stain x 400)



Neoplastic cells were immuno-reactive for vimentin and CD34; and non-reactive for cytokeratin, S-100, CD68 and desmin. Renal capsule, renal vessels and perinephric tissue was free of tumor infiltration. These findings suggested the diagnosis of HPC. Patient recovered well postoperatively and is asymptomatic one year after surgery.

DISCUSSION

In the wide spectrum of renal tumors primary sarcomas of kidney are rare, representing only 1-3% all malignant neoplasm of kidney [3]. The first case of primary renal HPC was documented by Black and Heinman in 1955 [4]. Since then only 38 cases has been reported in English literature.

Patients of renal HPC are slightly younger (mean 40.3, range 16-68 yrs) than the patients of other types of renal tumors [1]. They usually present with a painless abdominal mass which often acquires a huge size because of insidious growth. Haematuria is uncommon and is only seen in advance cases after infiltration of the pelvicalyceal system [1]. Infrequently symptoms like hypoglycemia and hypertension attributed to extensive metabolism of glucose and increased renin production by the tumor cells respectively has been observed which usually subsides after surgery [5,6].

No characteristic radiological sign has been described for HPC that can aid the preoperative diagnosis. However the pattern of large vessels surrounding the tumor during arterial phase enhances the possibility of pre-operative embolization [3].

Histopathology of HPC is characterized by a monotonous

cellular proliferation, stag horn vascular pattern lined by endothelial cells with solid aggregates of spindle shaped cells around them and minimal collagenization. Silver stain out lines the vascular basement membrane, which can be used to confirm that the proliferating cells are lying outside the basement of vascular endothelium. The neoplastic progenitor cells stains usually for vimentin and in a smaller percentage of cases with CD34 [2]. The diagnosis of HPC is one of exclusion, which includes differentiating it from solitary fibrous tumor, synovial cell sarcoma, deep benign fibrous histiocytoma, phosphaturic mesenchymal tumor and many others showing pericytomatous pattern focally. Immunohistochemical (IHC) analysis provides substantial help in excluding these tumors in conjunction with a thorough histological examination. Electron microscopy provides further evidence to differentiate between these tumors (HPC) and solitary fibrous tumors composed of fibroblast which in recent reports seems to be closely related to HPC [1]. In the present case differentiation from other renal and perirenal spindle cell lesions like sarcomatoid renal cell carcinoma, sarcomatoid urothelial carcinoma, leiomyosarcoma, malignant fibrous histiocytoma and solitary fibrous tumor was considered. Pericytic vascular pattern, minimal collagenization and lack of specific differentiation as suggested by IHC led us to categorize the lesion as HPC. Histological features associated with malignant behaviour are increased cellularity mitotic activity more than four per ten high-power fields, areas of necrosis and hemorrhage [1,7]. None of these features were found in the present case.

Surgery is the main stay of treatment and radical approach is necessary [1,3]. Pre-operative embolization reduces the tumor size as well as may offer a better surgical field [1,8]. Routine lymphnode dissection is not recommended as the tumor spreads primarily by the hematogenous route and only rarely through the lymphatics [9]. Radiotherapy is generally condemned [9,10]. Chemotherapy, as well has not been accepted as effective adjunct to therapy in major series of HPC in various location [9].

Clinical behavior of HPC is variable. Metastases appear in ≈15% of cases in various anatomic sites though others have reported up to 50% [1]. The factor that influences survival are related to size, age at diagnosis and histological patterns

CONCLUSION

HPC is a rare tumor occurring generally below 50 years. Computed tomography and magnetic resonance imaging has minimal role in diagnosis, however angiography can pre-operative embolize. IHC is helpful to exclude other tumors. Surgery is the main stay of treatment.

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