CASE REPORT

A 32-year old Female Diagnosed with Primary Adrenal Hemangiopericytoma

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Hemangiopericytomas are rare tumors originating from pericytes in the wall of capillaries. It is a type of soft tissue sarcoma which commonly involves the lower extremities, pelvic retroperitoneum, and head & neck. A 32-year old female presented with a 5 year history of slowly-growing right flank mass. Physical examination findings showed a bulging right flank mass extending anteriorly, approximately 15cm x 15cm, firm, movable, nontender mass on bimanual examination. CT-scan with IV contrast revealed a large suprarenal mass, right. She underwent adrenalectomy with en-bloc nephrectomy, right with uneventful post-operative course. Histopathology of the specimen was read as hemangiopericytoma. Further testing by Fluorescence In-Situ Hybridization confirmed the diagnosis. Metastatic work-up was negative. Hence, this is the first reported case of primary adrenal hemangiopericytoma. Surgical removal is the mainstay of treatment of hemangiopericytomas. Radiotherapy and chemotherapy have no role in the management of the disease.

Key words: hemangiopericytoma, adrenal tumor, urologic malignancy, pericytes

Introduction

Initially described in 1942 by Stout and Murray, hemangiopericytomas (HPC) are rare tumors derived from capillary pericytes. Common locations include the lower extremities, pelvis and the head and neck.[1] In world literature as of 2009, 42 cases of hemangiopericytomas were reported but they involve only the kidney.[2] To date, this is the first reported case of a primary hemangiopericytoma of the adrenal gland.

Presented is a case of a 32 year-old female with a slowly-growing right flank mass for 5 years who was diagnosed and treated at SPMC.

The Case

Patient is a 32 year-old female, married, a housewife, non-smoker and non-alcoholic beverage drinker with no history of hypertension, diabetes or previous surgery. Her symptoms started 5 years prior to consult when she noticed a bulge on her right flank upon doing household chores. She denied any history of dizziness, headaches or hypertensive episodes. The mass was slowly-growing for several years but no consult was done and no medications taken. She tolerated the condition until 3 months prior to consult, the patient experienced loss of appetite and weight loss which prompted consult at the Outpatient

Department. She was seen awake, conscious, coherent with BP of 110/70, HR of 70, RR of 16 and Temperature of 36.7°C. Physical examination findings showed bulging right flank mass extending anteriorly, approximately 15cm x 15cm, firm, movable, nontender mass on bimanual examination (Figures 1 & 2).

CT-scan of the whole abdomen with IV contrast (Figure 3) showed a huge mass on the right suprarenal space measuring 18cm x 16cm x 16cm with compression of the kidney downward and inferior vena cava and aorta medially.

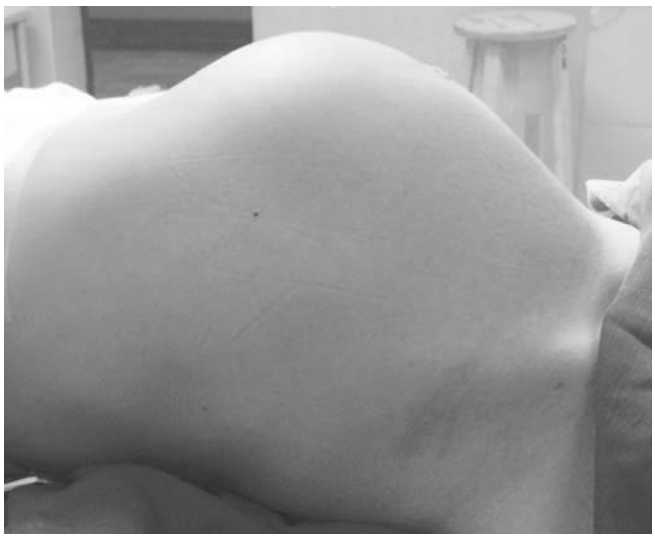


Figure 1. Bulging mass at right flank (side view).

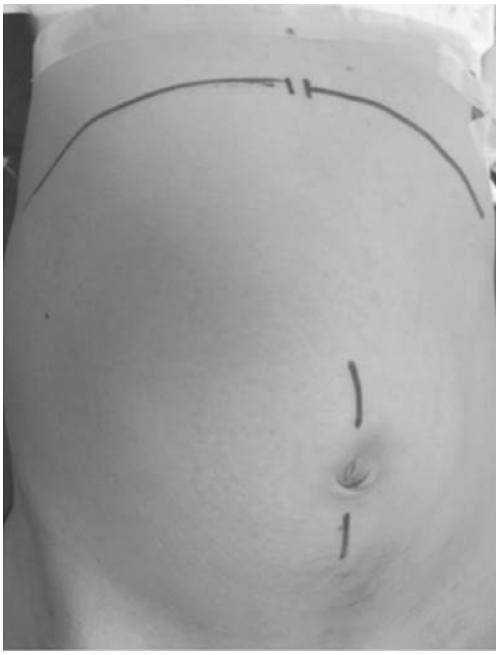
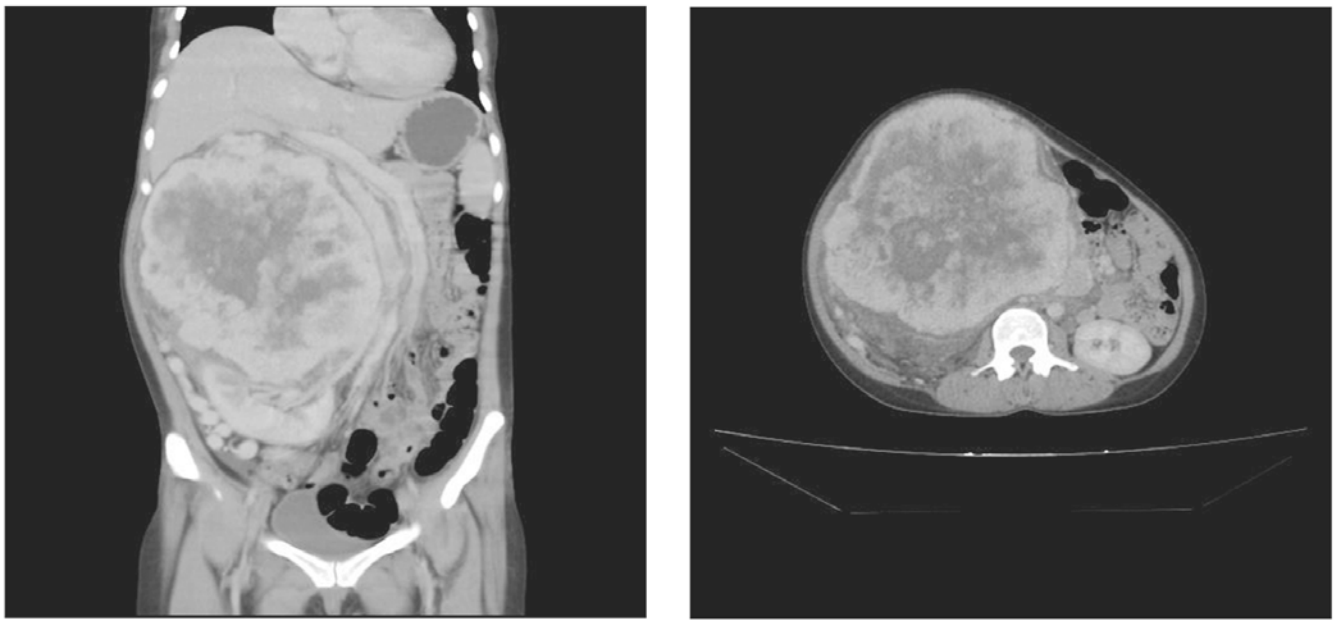


Figure 2. Bulging mass at right flank (front view).



A B

Figure 3. Abdominal CT-scan A. coronal view B. axial view.

Due to the location of the disease entity, differential diagnoses of flank masses included a renal, adrenal and retroperitoneal origin. Renal cause was ruled out due to the absence of hematuria and flank pain. Next step was to differentiate if it was a functioning or non-functioning adrenal mass. The three most common functioning adrenal diseases are pheochromocytoma, Cushing's syndrome and Conn's syndrome. Pheochromocytoma was ruled out because of the absence of headaches and tachychardia. Cushing's syndrome was ruled out due to the absence of Cushingoid features such as central obesity and buffalo hump. Conn's syndrome was also ruled out because there was no history of persistent hypertension or muscle weakness. The investigators were then left with a non-functioning adrenal mass versus a retroperitoneal mass, both of which require surgical excision.

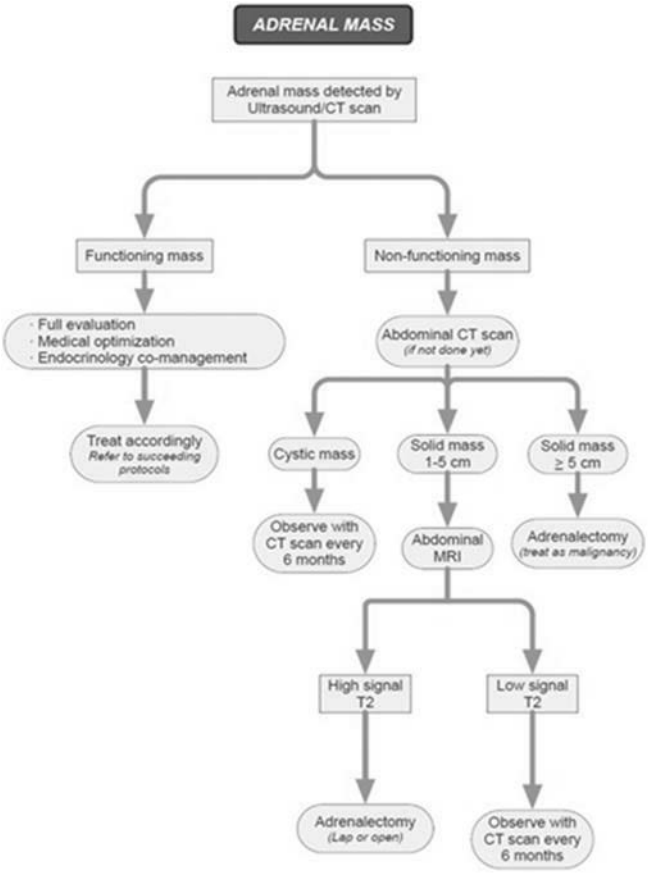


Figure 4. Management algorithm for adrenal mass (NKTI Clinical Practice Guidelines 2011-2012)

With a consideration of a non-functioning mass greater than 5 cms, the patient was scheduled for adrenalectomy with en-bloc nephrectomy, right via trans-abdominal approach. (Figure 4).

Intra-operatively, the mass was adherent to the IVC and venorraphy was done. (Figures 5 & 6). A total of 6 units of blood was transfused peri-operatively. Post-operatively, the patient progressed without complications. She was resumed on diet and extubated on the 2nd post-operative day and was discharged on the 7th post-operative day.

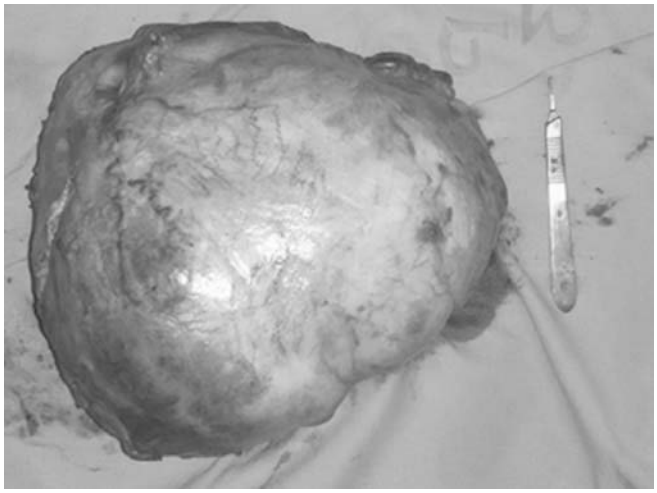


Figure 5. Adrenal mass.

Figure 6. Adrenal mass with right kidney.

On gross pathologic examination, the specimen measured 17cm x 22cm x 9cm, well-encapsulated mass with attached kidney. Cut section shows a tan, fleshy to solid surface with areas of necrosis. Hematoxy-eosin staining revealed sheets of atypical pericytes with numerous thin-walled ramifying vessels. There is very scanty intervening connective tissue. (Figure 7).

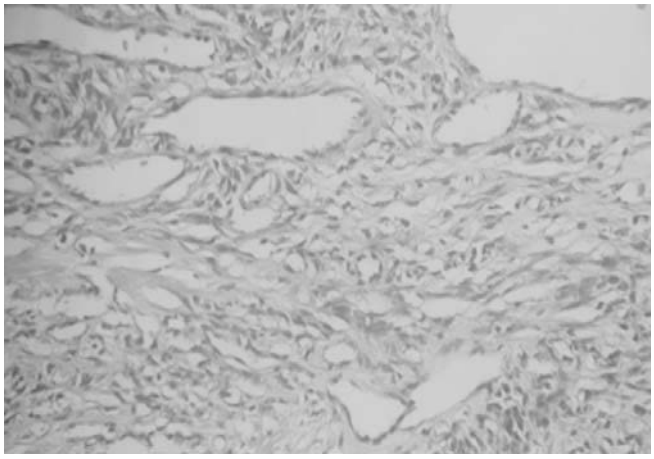


Figure 7. Atypical pericytes with thin-walled ramifying vessels.

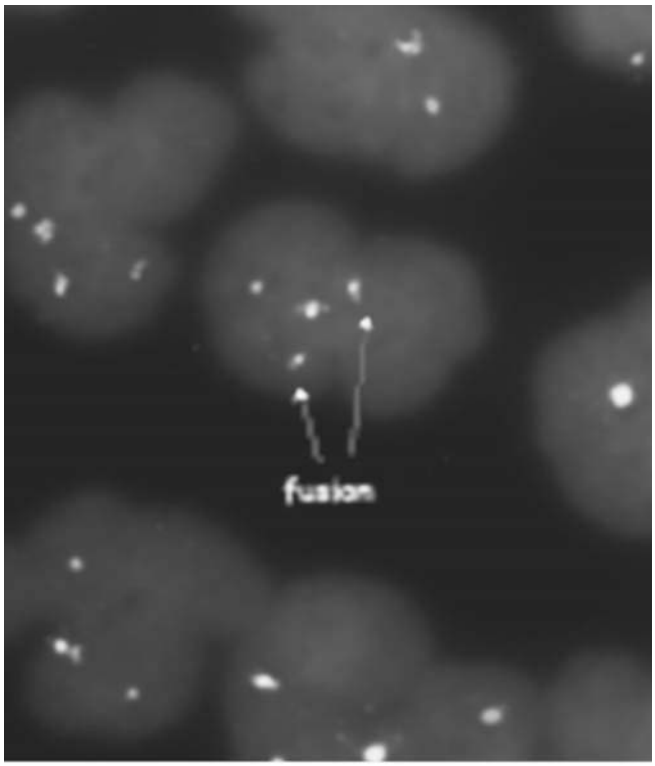


Figure 8. Fusion of DNA probes (pink: chromosome 12, blue-green: chromosome 19) signifies the translocation t(12;19).

Fluorescence in situ hybridization (FISH), the gold standard for diagnosis, was done in another institution and revealed 20 out of 50 informative cells (40%, cut off: 1.5%) had a translocation of chromosome 12 and chromosome 19, which is confirmatory for HPC (Figure 8)

Follow-up check-up of the patient was done at 3 months post-surgery and revealed a well-coaptated incision scar. The abdomen was soft and non-tender with no palpable masses. At 6 months post-op, a metastatic work-up was done which included CT-scans of the head, chest and abdomen which were all normal.

Discussion

Hemangiopericytomas are rare vascular tumors and can be found anywhere vascular capillaries are found. These tumors are commonly located in the musculature of the lower extremities, retroperitoneum, pelvis, head, neck and lungs.[3,4] In 2013, Filho et. al. reported a case of hemangiopericytoma of the kidney and was treated with nephrectomy.[5] Moreover, Pistolesi, et al. presented a case of meningeal hemangiopericytoma metastatic to the adrenal gland.[6] This is the first published case report regarding primary hemangiopericytoma of the adrenal gland.

The age of initial diagnosis of HPC is 470.3 years-old (range 16-86). There is a slight difference in male and female patients, with the predominance of females (17 vs 16).[6] Two distinct clinical entities have been described: the adult type and the infantile type, which has a favorable prognosis. In the adult type, as in the case, the reported 10-year-survival rate was 70%.[7] The eventual cause of death would be distant metastasis to the lungs or bone.[8]

Symptoms of HPC are unspecific. In 66% of cases, the presentation is a painless abdominal tumor,[9] Other symptoms are hematuria (if with infiltration to the kidney), hypoglycemia and

arterial hypertension. Hypoglycemia is attributed to the extensive metabolism of glucose within the tumor,[10] while hypertension is a result of renin production.[11]

Despite technical advances in the field of imaging, no characteristic signs of HPC have been described on ultrasonography, CT or MRI that might aid in the differential diagnosis. These studies usually depict a large mass, which may grow insidiously to a diameter of 25 cm, but with no pathognomonic features.[9]

Macroscopically, most tumors are well circumscribed lesions or a thin capsule, associated to complex network of vessels and, with small satellite nodules around the main mass. Only the minority is adhered to neighboring tissues.[5] Tumor size reported in the literature range between 2 and 25cm.[13] In the present case, the gross size of the tumor was 17cm x 22cm x 9cm, but in contrast, was very adherent to the inferior vena cava and posterior retroperitoneum.

The combination of histological and immunochemical patterns may provide a diagnosis of HPC and exclusion of differential diagnoses. (Table 1). HPC's would be positive for vimentin and CD34 while positive or negative on Bcl2 and SMA.

However, immunohistochemistry is of limited help in the identification of neoplastic pericytes and hence in the direct recognition of HPC.[7] A study of 163 sarcomas by Jhanwar, et al.[14] showed that hemangiopericytomas have a translocation of position 13 of chromosome 12 to position 13 of chromosome 19, [t(12;19) (q13;q13)]. Thus, cytogenetic analysis using FISH is confirmatory. The specimen showed 40% (cutoff: 1.5%) of informative cell had a t(12;19) abnormality, which confirmed the diagnosis of HPC.

Surgical removal is still the mainstay of the treatment of HPC, and should be as radical as possible in order to avoid incomplete resection and recurrence, which can be as high as 37.5%.[13] Ideally, a repeat CT-scan must be done every 6 months after surgery for 2 years then annually thereafter to document any recurrence. For large tumors, preoperative embolization can be done and has been reported in 2 cases.[15,16] There is no role of radiotherapy and chemotherapy in the treatment of HPC. In seven patients radiotherapy followed surgery, but only one was alive at 11 years, while the other six survived for a mean of 32 months.[17,18] Actinomycin D, nitrogen mustard, cyclophosphamide, vincristine, adriamycin, chlorambucil, fluoromethalone and methotrexate

Table 1. Immunohistochemistry result of hemangiopericytomas compared with other neoplasms (Source: Dabbs Diagnostic Immunohistochemistry, 4th edition 2013, p. 238).

	RCC w/ sarcomatoid features	Leiomyosarcoma	MPNST	PNET	Synovial Sarcoma	HPC
Vimentin	+	+	+	+	+	+
CD99	-	+/-	+/-	+	+	-
Bcl2	-	+/-	+/-	+/-	+	+/-
EMA	+	-	+/-	-	+	-
CK	+	-	+/-	-	+/-	-
E-Cadherin	+	-	-	-	+/-	-
CD34	-	+/-	-	+	-	+
S-100	-	+/-	+	-	-	-
Desmin	-	+	+	-	+/-	-
SMA	-	+	-	-	-	+/-
RCCA	+	-	-	-	-	-
Caldesmon	-	+	-	-	-	-
Inhibin	+/-	-	-	-	-	-

Abbreviation:
EMA - Epithelial Membrane Antigen
SMA - Smooth Muscle Antigen
PNET - Primitive Neuroectodermal Tumor
RCCA- Renal Cell Carcinoma Antigen
MPNST - Malignant Peripheral Nerve Sheath Tumor

were used but produced no favorable results in terms of sustained objective response of metastatic tumors.[19]

Conclusion

Hemangiopericytomas are rare tumors involving the capillaries and can occur anywhere in the body. Typical presentation would be painless growth, as in the patient.

Since HPC's present with a vague clinical picture and no pathognomonic features even with advances in image modalities, the diagnosis can only be confirmed after surgery. In this case, the specimen was sent to another institution for FISH for confirmatory testing. Furthermore, diagnosis of HPC's is only made thru a combination of histopathology, immunohistochemistry and cytogenetic analysis.

Surgery is the treatment of choice for HPC's. Adjuvant radiotherapy and chemotherapy have been used but showed no survival benefit.

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