Renal Preservation In A Giant Retroperitoneal Perinephric Schwannoma Mimicking An Adrenal Tumor And Posing Threat To Kidney

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Citation

Abstract
We present a case of 25 year old female presenting with left flank pain. The ultrasound and CECT imaging showed an adrenal mass, which after excision turned out to be a retroperitoneal Schwannoma. The mass was removed with the preservation of the kidney. On follow-up of 10 months, the patient is doing well.

INTRODUCTION
Schwannomas are rare tumors arising from Schwann cells of the peripheral nerve sheath. The retroperitoneal localization of Schwannoma is extremely rare. It has been shown that only 0.7% of Schwannomas arise in retroperitoneum[1]. This data, together with the absence of a typical clinical picture and the lack of peculiar instrument signs, make a preoperative diagnosis of this lesion really hard to ascertain. Nephrectomy is usually contemplated during excision of large tumors in relation to renal hilum[2]. This is an unique case of giant retroperitoneal Schwannoma mimicking an adrenal tumor, encroaching upon renal hilum and posing threat to kidney. The tumor was successfully excised preserving the kidney.

CASE PRESENTATION AND MANAGEMENT
A 25-year old female presented with left flank pain of 3 year duration. A clinically lump was palpable in the left lumbar and subcostal region. Her hematological and biochemical parameters were normal. Ultrasonography (USG) showed a well-defined mass in the right retroperitoneal region pushing the left kidney anteriorly. CECT abdomen and pelvis revealed a 16 x 6 x 5 cm heterogeneous mass lesion in relation to upper pole and hilum of the left kidney stretching aorta and renal vessels (Figure 1a&b). The serum cortisol and catecholamines were normal. Urine vanillyl-mandelic acid was 4 mg/day and urine cortisol level was normal. CT angiography was done to see the relation of renal vessels to the tumor. FNAB was not diagnostic. The tumor was approached with a chevron incision with midline extension up to xiphisternum. There was a large lobulated, well encapsulated mass stretching mesocolon and renal vessels anteriorly. The mass was in relation to upper pole and hilum of left kidney and extending behind diaphragmatic crus. After reflecting left colon, spleen and pancreas medially, left kidney and renal vessels were dissected meticulously away from tumor and the tumor was completely excised. Histologic examination showed spindle cell tumor pallisading in relatively more cellular areas. Immunostaining with S-100 protein shows strong positivity, suggestive of benign retroperitoneal Schwannoma (Figure 2a&b). At 10-month follow-up, the patient is doing well.
DISCUSSION
Retroperitoneal Schwannomas are extremely rare. Because of varied clinical presentation and no specific imaging characteristics, it is very difficult to predict these tumors preoperatively [3]. These tumors can mimic a multitude of different conditions like pancreatic tail tumours, hepatic tumor in caudate lobe, lymph node metastases etc [4]. Preoperative radiologic imaging may give some clue if cystic changes are seen because other retroperitoneal tumours rarely form cysts. Cystic changes are noted in 63% of benign and 73% malignant Schwannomas resulting from alterations in vascular wall [5]. However, a correct diagnosis is usually obtained only after surgical enucleation and immunohistochemical studies with S-100. The tumors expression of high concentration of the stain for S-100.

Figure 1
Legend 1 (a&b): CT abdomen showing the Schwannoma in relation to upper pole and hilum of the kidney.

Figure 2

Figure 3
Legend 2 (a&b): Microphotograph showing spindle cell tumour pallisading in relatively more cellular areas. Immunostaining with S-100 protein shows strong positivity

Figure 4
蛋白可区分施万瘤与其他梭形细胞肿瘤。良性施万瘤的治疗主要是手术切除，因为化疗和放疗都相当无效。这些肿瘤通常通过整块切除，连同周围器官进行完整的切除。Janaka et al.先前报道了一个类似的巨大腹膜后良性施万瘤，其表现形式类似于肾上腺肿瘤。在那个案例中，整块切除，包括肾脏和周围淋巴结，被实施。可以通过良好解剖学的映射和详细的计算机生成的3-D图像与术后彻底的术中解剖学术，一人可以保存肾脏，正如我们的案例所显示。

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References
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