Renal Preservation In A Giant Retroperitoneal Schwannoma Mimicking An Adrenal Tumor

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Citation

S Kumar, S Singh, N Acharya, Radotra, A Kishore, A Mandal. *Renal Preservation In A Giant Retroperitoneal Schwannoma Mimicking An Adrenal Tumor*. The Internet Journal of Urology. 2005 Volume 4 Number 1.

Abstract

We present a case of a 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 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 a 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

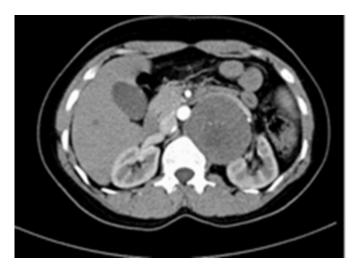
A 25-year old female presented with left flank pain of 3-year duration. Clinically a lump was palpable in the left lumbar and subcostal region. Her hematological and biochemical parameters were normal. Ultrasonography (USG) showed a well-defined mass in the left retroperitoneal region pushing the left 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).

Figure 1

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



Figure 2



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 dignostic. 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.

Figure 3

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

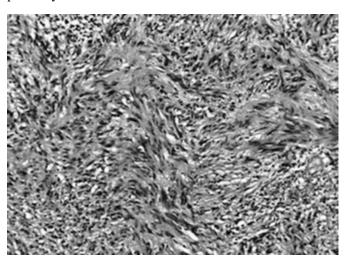
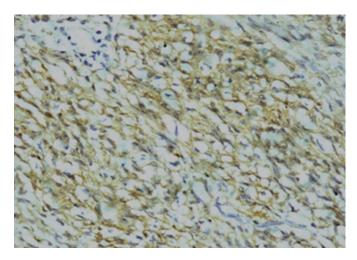


Figure 4



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 protein can distinguish Schwannomas from other spindle cell tumors. The treatment of both benign and malignant Schwannomas is primarily surgical excision because chemotherapy and radiotherapy are rather ineffective. These tumors are usually treated with En-bloc resection with the surrounding organ for complete extirpation. Janaka et al previously reported a similar case of giant retroperitoneal benign Schwannoma mimicking adrenal tumor. In that case, En-bloc resection including the kidney and regional lymph nodes was performed [2]. With good anatomical mapping of tumor relationships with renal hilar vessels with dedicated computer generated 3-D reformatted images and meticulous dissection intraoperatively, one can preserve kidney as in our case

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