CASE REPORT

Retroperitoneal ancient schwannoma involving the renal hilum

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Schwannomas, a soft-tissue tumor of a Schwann cell, involving the kidney are rare, with few cases available in the literature. Herein, we report a case of a rare variant, an ancient schwannoma of the renal hilum.

Key Words: neurilemmoma, kidney neoplasms, cystic kidney diseases

Introduction

A schwannoma, or neurilemoma, is a tumor that originates from Schwann cells. Schwann cells are derived from the neural crest and function to form myelin in the peripheral nervous system. While common in the head, as an acoustic neuroma, schwannomas involving the kidney are extremely rare. Retroperitoneal schwannomas involving the kidney are commonly misdiagnosed as renal cell carcinoma, resulting in radical nephrectomy. Similarly, the variant, an ancient schwannoma, is even less frequently seen. We report a case of a retroperitoneal ancient schwannoma of the renal hilum, which to our knowledge has been reported only once previously.

Case report

A 35-year-old female with a past history of nephrolithiasis was referred to our hospital for management of a left renal cystic mass. She had initially presented with complaints of abdominal pain and nausea. On examination, she had minimal tenderness to palpation in the left upper quadrant.

Abdominal ultrasound was performed and a left upper quadrant cystic mass was noted. CT imaging revealed an encapsulated and hypodense, cystic lesion with fine internal septations, Figure 1 and 2. On further evaluation, an MRI of the abdomen revealed a 6.2 cm x 6.7 cm x 6.5 cm encapsulated, round, cystic neoplasm. Thick, irregular enhancing internal septations were also present. The mass appeared to be located on the upper pole of the left kidney between the pancreatic tail and left adrenal gland. Although an adrenal cyst or pancreatic cyst could not be ruled out, these findings were suggestive of a cystic renal cell carcinoma.

After discussing with the patient her options of continued observation versus surgery, she elected to proceed with laparoscopic left radical nephrectomy. At the time of surgery, a greater than 6 cm cystic structure was noted on the superior aspect of the left kidney. The mass was densely adherent to the colonic mesentery and a large mesenteric artery was adherent to the cyst wall. The left adrenal gland, splenic vein, and body and tail of the pancreas were also adherant to the cyst. The cystic mass was excised along with the intact kidney without violation of the cyst wall. Frozen section of the mass was reported initially as an adrenal cyst. The patient tolerated the procedure well and there were no complications in the postoperative period.

Pathologic examination revealed a 7cm schwannoma with degenerative changes (ancient schwannoma) that had arisen from the left renal hilum. This tumor was described as a multicystic focally gelatinous mass located in the soft tissue of the renal hilum with no connection to the renal parenchyma. The left kidney had a benign cortical cyst while there was no abnormality in the left adrenal gland.

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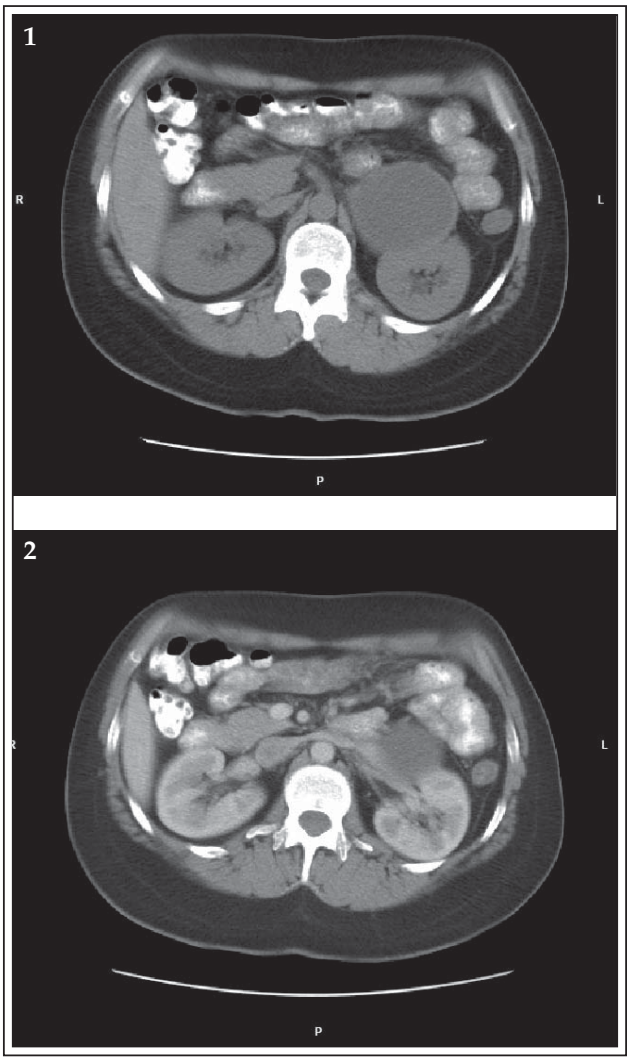

Figure 1 and 2. Non contrast and delayed contrast abdominal CT scans revealing cystic lesion related to the upper pole of the left kidney.

Discussion

Schwannomas, one of the more common soft tissue tumors, originate from Schwann cells of the nerve sheath. Typically encapsulated, they are usually solitary and slow growing tumors, rarely becoming malignant.[1] Though most commonly found in the head and extremities, they can be found less commonly in virtually any part of the body. The histological make up of the common schwannoma is described as an alternating pattern of Antoni type A and B. Antoni type A is composed of highly ordered compact spindle shaped cells, often forming palisading rows surrounding a more homogenous material which lacks nuclei. This unique pattern, known as Verocay bodies, is distinctive of schwannomas. Alternatively, Antoni type B has decreased cellularity and is arranged in a less orderly fashion.[1]

Schwannomas of the kidney are a rare occurrence with the literature reporting only 16 previous cases in the English language. The mean patient age of those reported and our patient was 53 years, with a female to male predominance, 11 and 6, respectfully.[2] The renal hilum was involved in only 2 of the 16 previously reported cases.[2] Schwannomas involving the kidney are difficult to diagnose, with non specific radiologic findings.[3] They are typically only diagnosed by histological examination following surgical excision.

Ancient schwannomas, also known as degenerative neurilemomas, is one of the rarest schwannoma variants. They were first described in 1951 by Ackerman and Taylor.[4] Characteristic degenerative changes are present, thought to be due to their slow, long term tumor development.[4,5] These changes include calcification, hemorrhage, hyalinization, and cystic changes, with at least one report of ossification found in the literature. Antoni type B predominates, providing a cytological atypia, leading to possible misdiagnosis of malignancy. With careful examination, mitotic figures are rarely seen.[1]

Our case of a retroperitoneal ancient schwannoma of the renal hilum is extraordinary with only one other case previously reported by Bezzi et al in 1996.[6] This previous case differed from ours in that the patient presented with obstructive symptoms and the mass was nonencapsulated. It is possible that cases previously reported to be malignant schwannomas involving the kidney were actually of the ancient variant type. Considering this and the lack of radiologic diagnostic criteria at this point in time, making a correct preoperative diagnosis of ancient schwannoma of the renal hilum is very difficult. □

References

1. Rosai J. Rosai and Ackerman's surgical pathology, 9th ed. Edinburgh, New York: Mosby; 2004:2264-2265.
2. Hung SF, Chung SD, Lai MK, Chueh SC, Yu HJ. Renal schwannoma: case report and literature review. *Urology* 2008;72(3)716.e3-6.
3. Isobe K, Shimizu T, Akahane T et al. Imaging of ancient schwannoma. *Am J Roentgenol* 2004;183:331-336.
4. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax: A clinicopathological evaluation of forty-eight cases. *Cancer* 1951;4(4):669-691.
5. Dahl I. Ancient neurilemmoma (schwannoma). *Acta Pathol Microbiol Scand* 1977;85:812-818.
6. Bezzi M, Orsi F, Rossi P. Nonencapsulated ancient schwannoma of the renal sinus. *Am J Roentgenol* 1996;166(6):1498.