Case Report

Cystic retroperitoneal renal hilar ancient schwannoma: Report of a rare case with atypical presentation masquerading as simple cyst

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ABSTRACT

Schwannoma is a benign soft tissue tumor of neural origin arising from the Schwann cells of the neural sheath. It has rarely been reported in renal or perirenal region. The preoperative diagnosis has mostly been confused with renal cell carcinoma in this location in most previous reports. We report a case that presented with a large "simple cystic" mass at the renal hilum. The preoperative differential diagnosis included hilar renal cortical cyst, renal sinus cyst, ureteropelvic junction obstruction (UPJO), or even a hydatid cyst. The final diagnosis was clinched only on histopathological examination.

Key words: Schwannoma, ancient, renal hilar, simple cyst

CASE REPORT

A 44-year-old lady presented to us with complaints of dull aching continuous pain in the right flank region for 1 year. She had intermittent acute exacerbations of pain associated with nausea/vomiting requiring hospitalization for pain relief. There was no associated fever, hematuria, lithuria, or lower urinary tract symptoms. On examination a large renal mass could be palpated bimanually in the right lumbar region. Ultrasonography revealed a large cystic hypoechoic area in the region of renal hilum suggestive of either UPJO with large extrarenal pelvis and gross hydronephrosis, or a parapelvic cyst causing extrinsic obstruction at ureteropelvic junction. All routine laboratory blood and urine investigations were normal. An intravenous urogram and CECT abdomen were done to further characterize the mass [Figure 1]. It confirmed an 11 × 12 × 10 cm simple cystic mass at renal hilum with proximal gross hydronephrosis and pushing the kidney superolaterally. There was no solid or enhancing component seen inside the mass. The right kidney showed delayed excretion of contrast with poor function and relatively thin parenchyma. However, no further additional anatomic information could be gained regarding the true pathology. ELISA for assay of antibodies to echinococcus was within normal limits.

An informed consent was taken for right retrograde pyelography and exploration. Retrograde pyelography confirmed gross hydronephrosis with a malrotated kidney; however, pelvis was not dilated to the size equivalent to that of the cyst on CECT, excluding the diagnosis of UPJO. A rib-cutting extraperitoneal flank approach was used through the 12th rib bed for excision/deroofing of the parapelvic cyst [Figure 2]. The cyst was densely adherent to the psoas fibers and sheath posteriorly, which were separated by a sharp dissection. The cyst was aspirated first to collect sample for cytology and then electively decompressed in a controlled fashion using suction canula. Care was taken to avoid any spillage of fluid by packing the surgical field with betadine soaked sponges. The fluid was straw colored and clear. There was no lamination of the cyst wall, unlike a hydatid cyst. The cyst opening was closed and further dissection done to separate it from ureter, pelvis, and lower pole of kidney. No attachment of the cyst was seen with the renal cortex. The cyst was excised in toto and sent for histopathological review which surprisingly revealed a cystic schwannoma typically positive for S-100 protein on immunohistochemical

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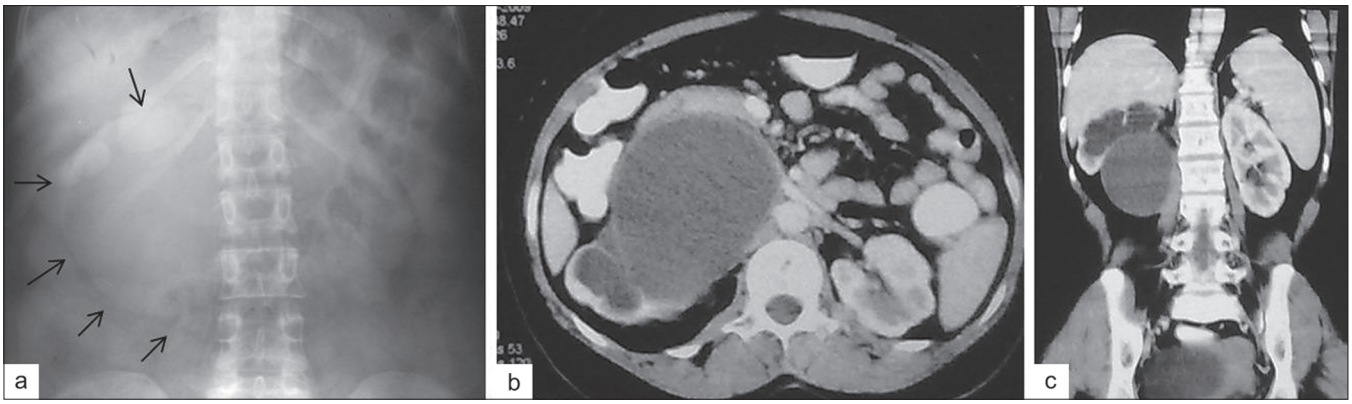

Figure 1: (a) Intravenous urogram. (b and c) Contrast enhanced CT scan showing a large cystic parapelvic right renal lesion with hydronephrosis. There was no apparent solid component inside the mass

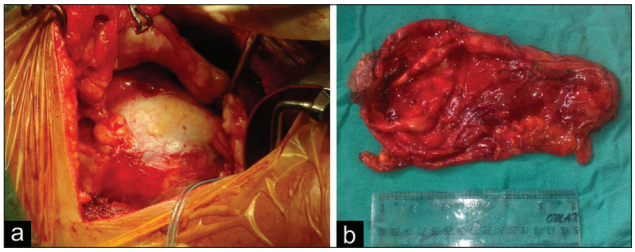

Figure 2: (a) Operative photograph showing the tense cystic mass at the right renal hilum. The ureter was separated and looped in the feeding tube. (b) Completely excised specimen

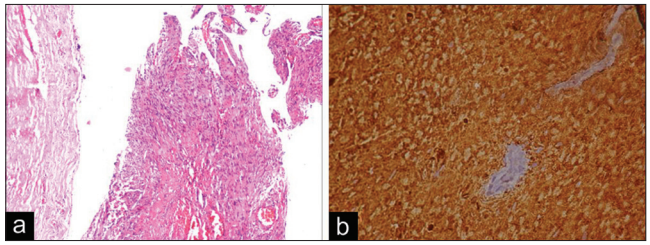

Figure 3: (a) HandE stained ×40 photomicrograph of the specimen showing a predominantly cystic tumor bounded by fibroconnective tissue capsule. The tumor was composed of Antoni A areas of compact spindle cells with focal nuclear palisading and few Antoni B areas of spindle to ovoid cells in a loosely textured matrix. Stroma showed many dilated thick-walled vessels and hemorrhage. (b) ×200 photomicrograph showing typical S-100 immunoreactivity of the schwannoma.

staining [Figure 3]. The cyst wall was 2-5 mm thick with no solid nodules.

At 3 months follow-up, the patient is asymptomatic. A renal dynamic scan was done which confirmed nonobstructive flow pattern.

DISCUSSION

Schwannomas are peripheral nerve sheath tumors which are usually benign. Retroperitoneum is a relatively uncommon site (0.7-3%) for schwannoma to occur.[1,2] They may be associated with von Recklinghausen's disease in 5-18% cases. Given the large loose areolar space available in the retroperitoneum, tumors in this location often present late with vague complaints related to compression of surrounding structures and often develop degenerative changes inside them. Such tumors which acquire histological degenerative changes over long standing duration are often referred to as "ancient" schwannoma. Cystic change is a fairly common (50-60%) association with this type of tumor in the retroperitoneum.[3] Rare presenting features like hematuria and secondary hypertension have been reported.[4]

Although there are several existing case reports of a schwannoma occurring at the otherwise quite rare location of renal, hilar, or perirenal region,[5-9] most of these have presented as either complex cystic or solid masses mostly confusing as renal cell carcinoma in the preoperative diagnosis. To the best of our knowledge, the presentation of a schwannoma mimicking a simple cyst has not been reported earlier. To this extent, the index case had a unique and atypical presentation, the histopathological diagnosis being a complete surprise.

Most cases are confirmed only on histopathology, since there are no specific features of schwannoma on imaging studies. The degenerative changes in ancient schwannomas may produce inhomogenous enhancement and complex cystic appearance in an otherwise well circumscribed mass on CT scan. MRI with gadolinium enhancement may provide a clue to the site of neuronal origin but provides no marked benefits over CECT.[10] A preoperative fine-needle aspiration cytology or biopsy is unreliable. Complete surgical resection with negative margins is the treatment of choice. Local recurrences are uncommon and malignant transformation is extremely rare. Laparoscopic resection of retroperitoneal schwannoma has also been described.[11]

Overall, the case represented a rare diagnosis, that too with an atypical presentation posing a diagnostic dilemma. No such previous presentation of a schwannoma has previously been reported in literature.

REFERENCES

1. Gubbay AD, Moschilla G, Gray BN, Thompson I. Retroperitoneal schwannoma: A case series and review. Aust NZ J Surg 1995;65:197-200.
2. Tortorelli AP, Papa V, Rosa F, Pacelli F, Doglietto GB. Image of the month – retroperitoneal schwannoma. Arch Surg 2006;141:1259-61.
3. Takatera H, Takiuchi H, Namiki M, Takaha M, Ohnishi S, Sonoda T. Retroperitoneal schwannoma. Urology 1986;28:529-31.
4. Singh V, Kapoor R. Atypical presentations of benign retroperitoneal schwannoma: Report of three cases with review of literature. Int Urol Nephrol 2005;37:547-9.
5. Alvarado-Cabrero I, Folpe AL, Srigley JR, Gaudin P, Philip AT, Reuter VE, Amin MB. Intrarenal schwannoma: a report of four cases including three cellular variants. Mod Pathol 2000;13:851-6.
6. Edwards B, Goodrich S, Sundaram CP. Retroperitoneal ancient schwannoma involving the renal hilum. Can J Urol 2009;16:4948-9.
7. Li Q, Gao C, Juzi JT, Hao X. Analysis of 82 cases of retroperitoneal schwannoma. ANZ J Surg 2007;77:237-40.
8. Ohta I, Lin PH, Rau CL, Wang KC. Evaluation of perinephric, retroperitoneal schwannomas: Case report and review of the literature. South Med J 2007;100:80-2.
9. Hung SF, Chung SD, Lai MK, Chueh SC, Yu HJ. Renal Schwannoma: Case report and literature review. Urology 2008;72:716-6.
10. Hughes MJ, Thomas JM, Fisher C, Moskovic EC. Imaging features of retroperitoneal and pelvic schwannomas. Clin Radiol 2005;60:886-93.
11. Yoshino T, Yoneda K. Laparoscopic resection of a retroperitoneal ancient schwannoma: A case report and review of the literature. Anticancer Res 2008;28:2889-91.

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