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Atypical presentation of an ancient retroperitoneal schwannoma mimicking a renal hydatid cyst: a case report and literature review

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Abstract

Schwannomas are benign neurogenic encapsulated peripheral nerve tumors of Schwann cells that rarely occur in the retroperitoneum. We present a case of a 22-year-old man who presented with right flank pain and was initially diagnosed as hydatid cyst based on the imaging and borderline results of anti-echinococcus (IgG). Albendazole was prescribed and taken for 4 months with no response and later underwent surgical excision of the mass. The diagnosis and morphology confirmed the nature of the schwannoma. Complete surgical excision remains the gold standard for the management of these tumors. The preoperative diagnosis is usually difficult; however, the definitive diagnosis is made upon histopathological examination.

Keywords Schwannoma, Retroperitoneal mass, Renal hydatid cyst, Surgical excision, Case report

1 Introduction

The American Journal of Neuroradiology defines "ancient schwannoma" a benign encapsulated tumor of long-standing duration [1], and the term has been used mainly in extracranial schwannomas such as the head and neck region or on the flexor surfaces of the extremities [2]. The term "ancient" refers to the histological degenerative features, which are acquired with increasing age in these tumors [1]. Benign neurogenic tumors (schwannomas, neurofibromas), fibromatosis, and renal angiomyolipomas are the commonly found in the retroperitoneum [2]. However, when associated with von Recklinghausen disease, they are malignant [3]. Schwannomas that occur in the retroperitoneum are uncommon, representing only 0.5–5% of all schwannomas [4]. They tend to occur more frequently in women when compared with men [5].We report a case of a 22-year-old male, initially diagnosed with a hydatid cyst. Upon follow-up after 4 months, it was later identified as an ancient schwannoma. The preoperative diagnosis is usually difficult; however, the definitive diagnosis is made upon histopathological examination.

2 Case presentation

We have reported this section as recommended in the CARE Checklist of information.

2.1 Patient information

A 22-year-old male patient, visited the Sindh Institute of Urology and Transplantation, Karachi, in January 2023. He complained of pain in his left flank that radiated to his back. The pain had been gradually increasing in intensity over the past year. The patient had no significant medical or surgical history and was a non-smoker and farmer by occupation. The abdomen was normal on examination, and both testes were palpable in the scrotum. An abdominal ultrasound was performed, which revealed a cystic mass located above the

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upper pole of the kidney, adjacent to the duodenum anteriorly, the inferior vena cava medially, and the renal hilum laterally. The patient's hemoglobin level was 12.2 gm/dL, and their total leucocyte count was 5600/uL. Their random blood sugar level was 166 mg/dL, and renal function was preserved. To rule out primary retroperitoneal germ cell tumors or metastasis from gonads, the patient's testicular cancer tumor markers were checked, including alpha-fetoprotein, human chorionic gonadotropin, and lactate dehydrogenase; the results were normal.

2.2 Image findings

A CT-CAP triphasic was planned and performed, which reported a well-defined round hypodense cystic area in the retroperitoneal region on the right side abutting the duodenum anteriorly, the inferior vena cava medially, and the renal hilum laterally. It measured 4.5×4.3×5.7 cm (AP×TV×CC). The features were suggestive of a hydatid cyst (Fig. 1). The liver, gallbladder, pancreas, and both adrenal glands were unremarkable. Both kidneys showed normal parenchymal enhancement with no calculi or hydronephrosis. No significant abdominal lymphadenopathy was noted. Additionally, there were no abnormal findings in the chest.

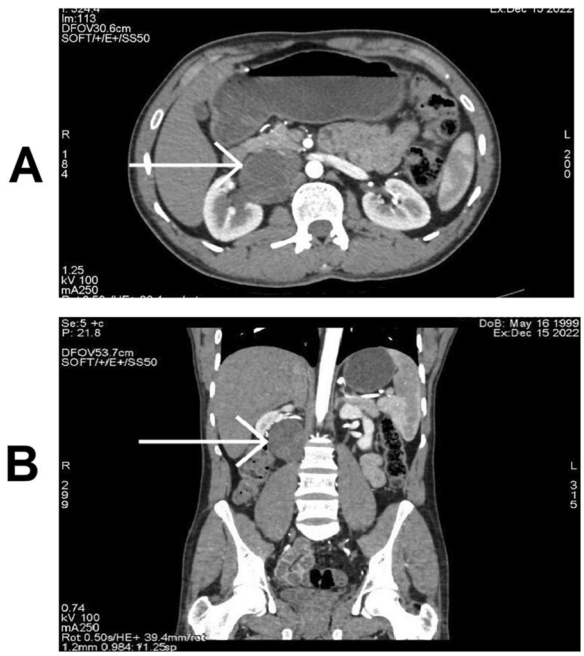

Fig. 1 CT scan axial (**a**) and coronal (**b**) views demonstrating hypodense cystic area in retroperitoneum on right side abutting duodenum, inferior vena cava, and renal hilum. *CT* computed tomography

2.3 Diagnosis and treatment

The provisional diagnosis of a hydatid cyst was made based on the imaging and borderline results of anti-echinococcus (IgG), and the patient was prescribed to take albendazole for 4 months. A repeat CT-CAP was performed after the aforementioned timeline, demonstrating an unchanged complex cystic structure in the retroperitoneum on the right side. An informed consent was taken, and an upfront surgical resection of the mass was planned and performed. A supra-twelfth right flank incision was made to allow entry into the retroperitoneum. The renal hilum, vessels, and ureter were identified, and a well-defined mass was noted abutting the inferior vena cava. The mass was mobilized from surrounding structures and feeding vessels were ligated. After excision, the specimen containing mass and feeding vessels was submitted for histopathological analysis. The right kidney was spared, and there was no iatrogenic injury noted to surrounding structures. Postoperatively, the patient was shifted to the intensive care unit for observation. On the third postoperative day, the patient was stepped down to the regular ward. He had a smooth recovery and was discharged from the hospital on the 5th postoperative day.

2.4 Histopathological findings

Informed consent was taken for histopathological examination. On gross examination, the mass was cystic-cum-solid tumor measuring 6×6 cm in size. On cutting, it was predominantly cystic, multiloculated and filled with hemorrhagic fluid. No gross necrosis was seen. On microscopic examination of solid areas, it revealed a neoplastic lesion exhibiting hypocellular and hypercellular areas. The hypercellular areas were composed of spindle cells with elongated tapered ended nuclei and eosinophilic cytoplasm. Verocay bodies were observed in addition to edematous changes in the hypocellular areas. Moreover, focal degenerative atypia was noted; however, no atypical mitosis, necrosis, or evidence of malignancy was seen. Immunohistochemistry (IHC) for Vimentin and S-100 was diffusely positive (Fig. 2). The morphological and IHC features were consistent with the diagnosis of ancient schwannoma.

2.5 Follow-up

The case was discussed in a tumor board meeting, and it was decided to keep the patient on surveillance. On a follow-up visit at 3 months, a CT-CAP was performed, and there was no recurrence noted. The patient was advised to follow-up every 6 months.

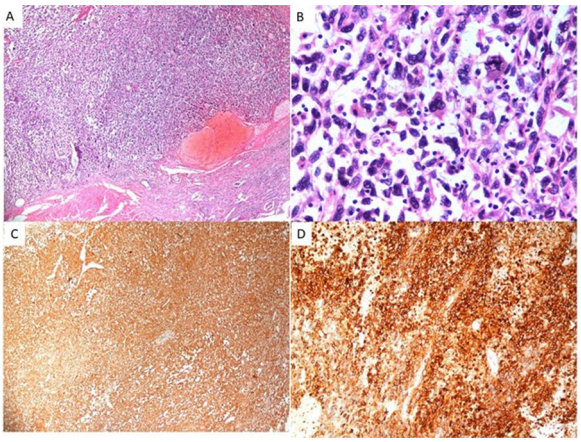

Fig. 2 **a** Low-power view showing a predominantly spindle cell tumor (H&E, ×40). **b** High-power view showing highly pleomorphic and bizarre tumor cells (H&E, ×400). **c** IHC for vimentin showing diffuse positivity in tumor cells (IHC for vimentin, ×40). **d** IHC for s-100 showing diffuse positivity in tumor cells. *IHC* immunohistochemistry, *H&E* hematoxylin and eosin stain

3 Discussion

Retroperitoneal masses are a diverse group of lesions that arise in the retroperitoneal spaces, presenting a diagnostic challenge for radiologists [6]. The most commonly found masses in the retroperitoneum are benign neurogenic tumors (schwannomas, neurofibromas), fibromatosis, and renal angiomyolipomas [2]. Schwannomas that occur in the retroperitoneum are rare, representing only 0.5–5% of all schwannomas [4] commonly occurring in the head and neck region or on the flexor surfaces of the extremities [7]. Ancient schwannoma constitute 0.8% of all soft tissue tumors [8]. Patients are mostly diagnosed between ages 40 and 60 years, and the male-to-female ratio is 2:3 [9]. Cystic changes are frequently observed in 50–60% of cases in retroperitoneal tumors of this type. Retroperitoneal schwannomas have nonspecific symptoms which vary depending on their size, location, and anatomical position, resulting in slow growth. This delay in diagnosis is due to the nature of the tumor [10].

There have been numerous case reports documenting the occurrence of schwannoma in rare locations such as the renal, hilar, or perirenal region [11–16], and most of these have presented as either complex cystic or solid masses mostly confusing as renal cell carcinoma in the preoperative diagnosis. Isolated renal hydatid cyst is rare, and it forms 2–4% of hydatid disease. Their infrequent occurrence is due to nonspecific signs and symptoms, lack of distinguishing radiological features, and its primary observance in middle-aged females. We thereby report herein a case report presenting a unique presentation of a schwannoma mimicking a renal hydatid cyst in a young male [17].

Diagnosing renal hydatid disease is challenging as there are no specific symptoms or signs that definitively confirm it. Only 50% of cases receive a reliable diagnosis, with 71% receiving a presumptive diagnosis based on a combination of clinical history, radiological findings, and serological and urinary studies [18]. Renal hydatid cysts can remain symptomless for extended periods or lead to complications like infection, abscesses, necrosis, hemorrhage, or obstruction [19]. Ameur and colleagues reported 34 cases, noting various clinical presentations, including pain, hematuria, the presence of a mass, hydatiduria, prolonged fever, and hypertension [20]. Notably, there are no specific serological or immunological tests that can definitively confirm the presence of a hydatid cyst [19].

Radiological studies have an important role in the diagnosis of renal hydatid disease. However, since the patient did not show any improvement after treatment for renal hydatid cyst, we did a histopathological examination; to our surprise, the morphological and IHC features were consistent with the diagnosis of ancient schwannoma. The tumor's histopathological features include the presence of a visible capsule surrounding it, and its histological composition consists of both Antoni A regions and Antoni B regions within a myxoid stroma [21]. A theory was proposed by Enzinger, stating that the degeneration of Antoni B areas resulted in cyst formation, which coalesced over time and formed a single cystic cavity [22]. The degenerative features of these tumors, including cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification, and degenerative nuclei with pleomorphism, lobulation, and hyperchromasia, are distinctive [1].

On CT scan, schwannoma appears as a clearly delineated and uniform mass, with rim enhancement of its fibrous capsule after contrast injection. Recently, mass needle biopsy has gravitated more attention, especially for young patients who may require different therapeutic approaches, from observation to surgical excision. Core needle biopsy, a simple and safe procedure, has an accuracy rate of 98% in distinguishing between benign and malignant soft tissue tumors [23]. In large retroperitoneal tumors, MRI offers superior visualization of the tumor's origin, vascular architecture, and involvement of nearby organs. According to some authors, angiography may also be recommended to assess the tumor's vascularity and/or for presurgical embolization [21].

Schwannomas are resistant to radiotherapy and chemotherapy; therefore, surgical intervention is the only treatment option available. However, the requirement for negative margins of soft tissue is a topic of debate, especially when sacrificing adjacent tissue or organs is necessary. Benign schwannomas have a positive prognosis,

but the most common complication is tumor recurrence, which occurs in 5–10% of cases and is likely due to incomplete removal [5]. To minimize the surgical risk and preserve adjacent vital tissues in benign types, it is recommended to perform subtotal resection. Benign schwannomas have a good prognosis, and the occurrence of recurrence is rare [23].

4 Conclusion

We presented an unusual case of retroperitoneal schwannoma imitating a renal hydatid cyst. A provisional diagnosis of a hydatid cyst was made for a patient with retroperitoneal cystic structures. After a course of albendazole and an unaltered CT scan, surgical resection was performed without complications, sparing the right kidney. Histopathological examination of the resected mass, which exhibited cystic and solid characteristics, revealed an ancient schwannoma with spindle cells, Verocay bodies, and positive immunohistochemistry for Vimentin and S-100. The patient had a smooth recovery and was discharged on the 5th postoperative day.

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Author contributions
All authors have read and approved the manuscript. HB and AA proposed the study concept and design. MN and ASH acquired the data. HB, AA, and MN drafted the manuscript. ASH critically revised the manuscript.

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Availability of data and materials
All data generated or analyzed during this study are included in this published article.

Declarations

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Consent for publication
Yes, consent of publication was obtained directly from patient of legal age.

Competing interests
The authors declare that they have no competing interests.

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