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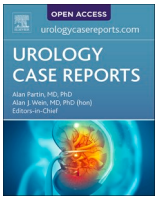

Oncology

Rare adrenal schwannoma treated with robotic-assisted adrenalectomy: A case report

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A B S T R A C T

Schwannomas are benign neoplasms that arise from peripheral nerve sheaths. Typically found in peripheral nerves of the head, neck, and extremities, these tumors seldom arise in the retroperitoneum. We report the case of a 50-year-old male with a 5 cm right adrenal mass removed via robotic-assisted laparoscopic approach due to concerns for adrenocortical carcinoma, which surgical pathology revealed to be a rare adrenal schwannoma. This is the reported case.

1. Introduction

A schwannoma is a benign peripheral nerve sheath tumor that develops from Schwann cells. They are found most commonly in the head, neck, and extremities.[1] However, while rare, 1–3% of schwannomas arise from the adrenal gland.[2] Adrenal schwannomas are linked to the Schwann cells of the phrenic nerve, vagus nerve, and sympathetic trunk, which innervate the adrenal glands.[3] They are most often found incidentally on imaging and can be misdiagnosed as adrenal cortical adenoma.[4] While adrenal schwannomas are typically asymptomatic, some individuals may present with abdominal or flank pain.[5] Like other schwannomas, most adrenal schwannomas are benign and have a good prognosis after surgical excision. There have been few published reports of adrenal schwannomas to date, consistent with its low incidence rate. We report a case of a 50-year-old male with an asymptomatic, incidentally discovered adrenal schwannoma.

2. Case presentation

A 50-year-old male with a medical history including hypertension, Rocky Mountain spotted fever, alpha-gal allergy, autoimmune hepatitis, hemochromatosis, non-alcoholic steatohepatitis, and cirrhosis was referred by his primary care physician to Gastroenterology at our institution. The referral was due to elevated liver function tests and hepatosteatosis observed on ultrasound.

Further evaluation by the Gastroenterology team included magnetic resonance imaging (MRI) of the abdomen with and without contrast, initially intended to exclude hepatocellular carcinoma. This MRI incidentally revealed a 4.9 × 5.0 × 4.0 cm oval mass arising from the medial limb of the right adrenal gland (Fig. 1).

The mass demonstrated heterogeneous progressive enhancement post-contrast. Given these imaging characteristics, differential diagnoses included adrenocortical carcinoma, pheochromocytoma, and potential metastasis. A hormonal analysis to evaluate for a functional adrenal mass was performed, and the patient's serum electrolytes, glucose, aldosterone, renin, and 24-h urine catecholamines and metanephrines were all within normal limits. Consequently, urology was consulted for further assessment and management.

At the time of evaluation, the patient was asymptomatic. His vital signs were notable for mildly elevated blood pressure at 141/90 mmHg. Both his review of systems and physical examination were otherwise unremarkable. The patient underwent a robotic-assisted right adrenalectomy. The patient's postoperative course was uneventful, and he was discharged on post-operative day two.

Histopathological examination of the excised mass revealed an encapsulated neoplasm. It was composed of cytologically bland spindle cells arranged in short fascicles and exhibited features characteristic of Antoni A and B patterns, including more densely cellular areas with nuclear palisading and paucicellular areas, respectively. A pathological diagnosis of adrenal schwannoma was made.

3. Discussion

Schwannomas are tumors that arise from Schwann cells enveloping peripheral nerves, predominantly found in the head, neck, and flexor surface of extremities.[6] It is rare for schwannomas to develop from the adrenal gland. Adrenal schwannomas account for 1–3% of all schwannomas and only 0.2%–0.5 % of adrenal tumors.[6] Thus far, only about 40

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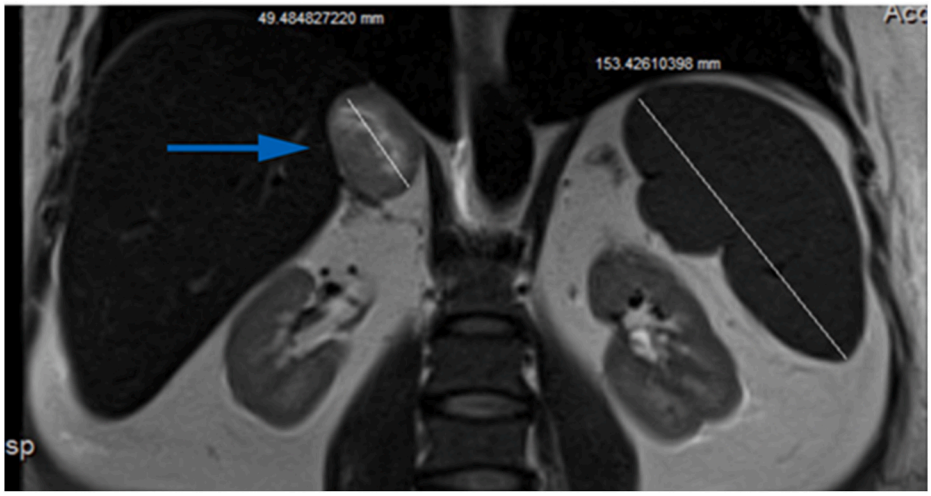

Fig. 1. Blue arrow shows a 4.9 cm oval mass arising from the right adrenal gland.

cases of adrenal schwannomas have been reported in the medical literature.[7] The present case represents an addition to this limited body of literature, contributing to our understanding of diagnosis and intervention for this uncommon type of tumor.

Adrenal schwannomas typically manifest in middle-aged individuals around the fifth decade of life and are frequently discovered incidentally during imaging studies unrelated to adrenal concerns.[8] While the patient presented is male, adrenal schwanommas occur about 1.2 times more frequently in women.[9] Most cases—including the one presented here—are asymptomatic, but about 30–40 % of patients, potentially associated with larger tumor sizes, can present with non-specific pain in the abdomen, flank, or back.[5,10] In a case series of 33 patients, all except one patient demonstrated normal laboratory findings, with the exception being elevated urinary catecholamines.[5] This is consistent with our patient, whose serum electrolytes, cortisol, and urine catecholamines and metanephrines were all within normal limits.

Morphologically, adrenal schwannomas are encapsulated, round, hypovascular tumors with a homogenous texture. However, up to 40 % of cases have reported a heterogeneous appearance, due to calcifications and cystic or hemorrhagic degeneration.[11] Sizes of adrenal schwannomas have ranged from 1.0 to 15 cm across reports, with an average size of 5.7–8.1 cm.[6] Considering the association between adrenal incidentalomas larger than 4 cm and an elevated risk of malignancy, the adrenal schwannoma observed in this case represents a tumor of typical size but raises concerns regarding its potential malignancy.[12] This underscores the importance of thorough investigations, most importantly resection and possible immunohistochemical staining, to establish an accurate diagnosis. Histopathologically, these tumors demonstrate hypercellular areas with spindle-shaped Schwann cells (Antoni A areas) and nuclear palisading (Verocay bodies), alternating with loosely textured hypocellular areas (Antoni B areas), as was the case here.[13]

Despite the challenging nature of preoperative diagnosis due to the lack of specific clinical or radiological features, a systematic review of primary adrenal schwannomas indicates a favorable post-surgical prognosis. Among 85 patients, there were no instances of recurrence or distant metastasis at an average follow-up period of 45 months.[10] Our decision to move forward with surgical removal aligns with this approach. Our patient remains well at two months post-adrenalectomy, supporting the overall favorable prognosis associated with surgical intervention in adrenal schwannoma cases.

4. Conclusion

Adrenal schwannomas are rare tumors most often found incidentally and hard to clinically differentiate from other potentially malignant adrenal masses. Difficult to diagnose with imaging or laboratory studies, histological and immunohistochemistry analyses are necessary to make a definitive diagnosis of adrenal schwannoma. Coupled with the fact that adrenal masses greater than 4 cm have an increased risk of malignancy, surgical excision with adrenalectomy should be considered standard management for a suspected tumor of this size. As illustrated by this case, it is prudent to consider adrenal schwannoma in the differential diagnosis of an incidentally found adrenal mass.

CRediT authorship contribution statement

Glendon Markollari: Writing – review & editing, Writing – original draft, Data curation. **Hunter J. Kraus:** Writing – review & editing, Validation. **Naveen Pokala:** Writing – review & editing, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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