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Case Report

An incidental large adrenal schwannoma: A case report ☆

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ABSTRACT

Adrenal schwannoma is a rare tumor of Schwann cell origin that represents less than 0.2% of all adrenal tumors. These typically benign tumors are most often found in the head, neck, and limbs. However, schwannomas can also rarely occur rarely in the adrenal gland within the retroperitoneal cavity. In the adrenal gland, these tumors arise from the medulla and are difficult to diagnose, often misdiagnosed as other benign or malignant entities. In this article, we report the case of a 43-year-old female with a large left adrenal mass revealed by biopsy to be a schwannoma. We focus on the use of radiological imaging modalities and immunohistochemical analysis to optimize diagnosis and treatment intervention of this rare tumor.

Introduction

Schwannomas are benign tumors that originate from Schwann cells in the myelin sheath of peripheral nerves. These typically benign tumors are most often found in the head, neck, and limbs, and, rarely, in the retroperitoneal space [1]. In the adrenal gland, schwannomas make up less than one percent of adrenal tumors [2], with, as of 2022, approximately 60 cases having been reported in the literature. Patients usually present with nonspecific mild symptoms such as persistent abdominal discomfort or nausea and vomiting secondary to mass effect on adjacent organs, leading to delayed presentation and extensive tumor growth. This results in a more difficult surgical excision of the mass and extensive blood vessel involvement. Furthermore, the nonspecific clinical nature along with the scarcity of literature describing characteristic imaging features render this tumor difficult to distinguish from other benign or malignant entities such as adrenal adenomas, myelolipomas, adrenocortical carci-

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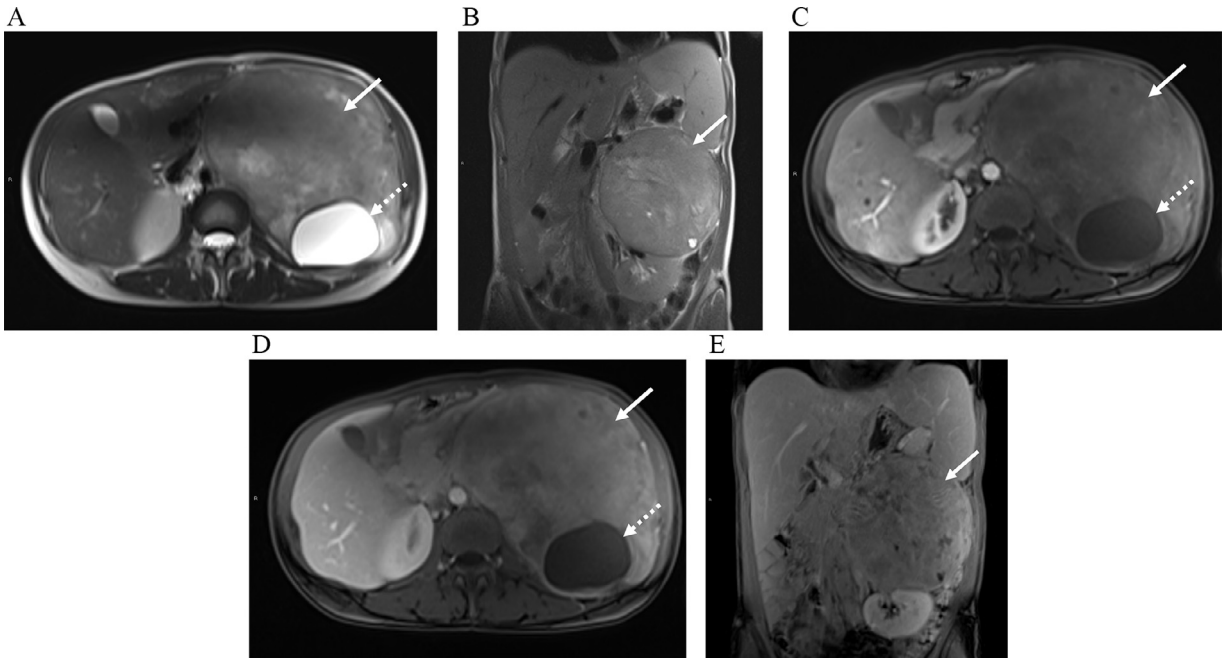

Fig. 1 – Axial (A) and coronal (B) T2-weighted MR images show a large well-circumscribed left suprarenal mass (arrows) with heterogeneous T2 signal intensity and internal cystic component (dotted arrow). Axial IV contrast-enhanced T1-weighted fat suppressed MR images in the arterial (C) and venous phase (D) and coronal IV contrast-enhanced T1-weighted fat suppressed MR image with a 5-minute delay show progressive heterogeneous enhancement in the large suprarenal mass (arrows). No internal enhancement within the cystic component (dotted arrows).

nomas, neuroblastomas, and other non-functioning adrenal tumors (that do not produce catecholamines or steroids). Here, we report the case of a 43-year-old female who was diagnosed with an adrenal schwannoma and underwent complete retroperitoneal resection of the tumor.

Case presentation

A 43-year-old Asian female with an insignificant past medical history presented to the primary care clinic following vague abdominal discomfort persisting for about 10 months and an unintentional weight loss of 10-15 lbs. Her family history was significant for bone cancer in her paternal grandfather. Physical examination revealed a nontender mass in the left lumbar region and subsequent lab work included CEA, CA19-9, aldosterone, renin, serum electrolytes, cortisol, and urinary metanephrines, all of which were unremarkable. The patient was scheduled to receive a magnetic resonance imaging (MRI) scan of the abdomen and follow up as an outpatient.

The MRI with and without contrast (Fig. 1) revealed a large, solid-cystic (almost solid) mass with heterogenous post-contrast enhancement arising from the left suprarenal region measuring approximately 15.6 × 10.9 × 13.5 cm with a posterior cystic component measuring 6.1 cm. Other findings of note included an enlarged spleen measuring 14.8 × 12.8 cm without any focal mass, as well as associated mass effect and inferior displacement of the left kidney. Differential diagnoses included a malignant adrenal lesion, favoring primary adrenocortical carcinoma. Two weeks later, an ultrasound-guided fine needle aspiration was performed, and pathology revealed lesional cells positive for S100 and SOX-10 proteins while negative for SMA, DOG-1, and myogenin, consistent with schwannoma.

Two and a half months later, a computed tomography (CT) scan of the patient's abdomen and pelvis with and without contrast (Fig. 2) was conducted for re-evaluation. A solid, heterogeneously enhancing left retroperitoneal mass measuring up to 17 cm was seen with cystic components suggestive of internal hemorrhage and/or necrosis, and consistent with biopsy-proven schwannoma of the left adrenal gland. The lesion did not appear to invade any adjacent structures but exerted mass effect on the splenic and left renal vasculature, as well as the left kidney, which was displaced inferiorly. Internally, there appeared to be a few areas of necrosis and/or hemorrhage with fluid level in a loculation or collection in the posterior aspect of the lesion measuring up to 6.1 cm.

Due to the size of the mass and its effect on surrounding structures, the mass was resected by neurosurgery via retroperitoneal approach. Surgically, the mass was a bulky, nodular, tan-yellow, myxoid and gelatinous interior with central necrosis and no evidence of mitosis. The patient has made an uneventful recovery from surgery, and is strong in the left lower extremity with no deficit. Follow-up evaluation of the tumor over time with serial imaging was recommended on a 4-to-6-month basis.

Discussion

We present a case of adrenal schwannoma, a rare benign tumor arising from the myelin sheath of peripheral, cranial,

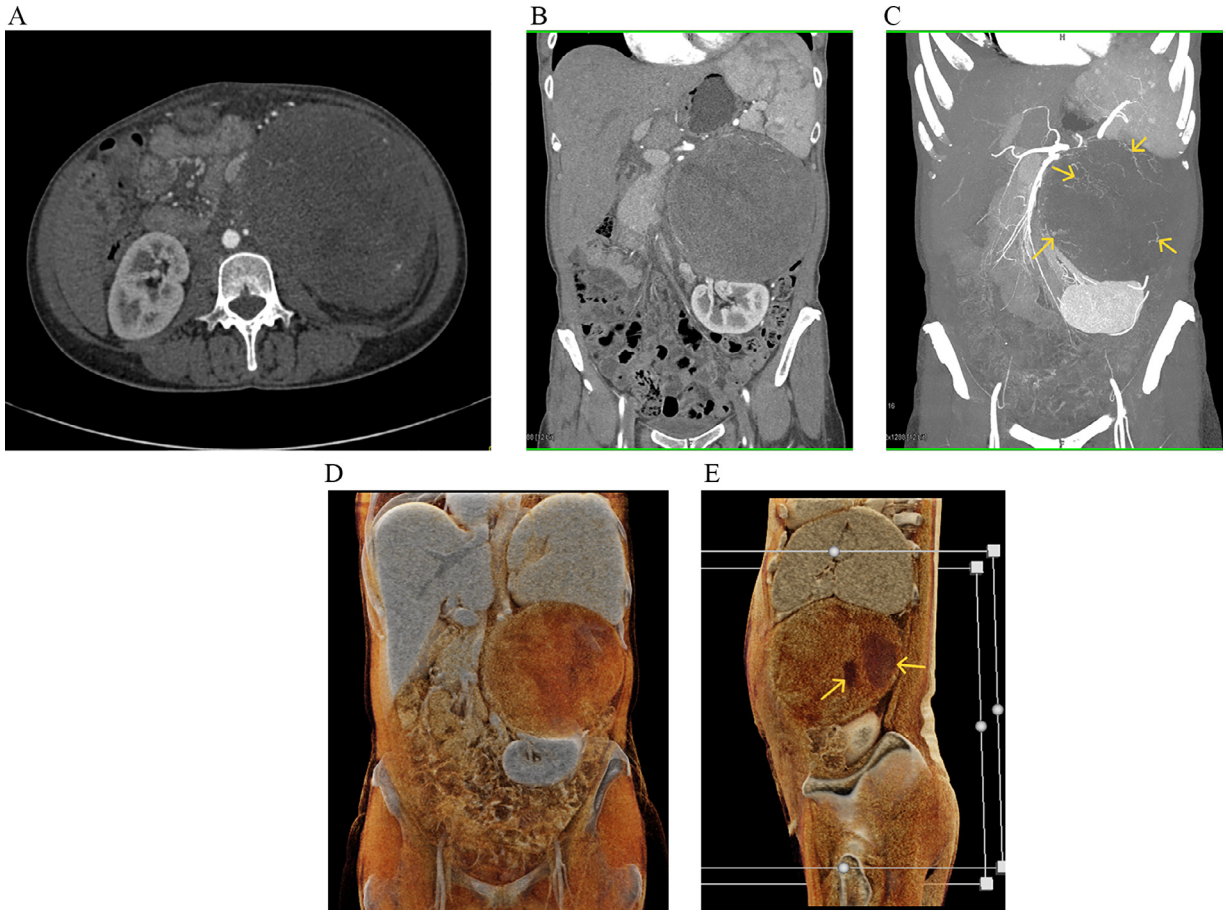

Fig. 2 – CT scan demonstrates 17 cm solid mass with areas of necrosis on axial and coronal views (A,B). Maximum intensity projection imaging (C) shows areas of neovascularity (arrows) and compression of the left kidney. Finally, the cinematic rendered images (D,E) show the tumor necrosis (arrows) with displacement of the left kidney downward. Sharp margins of the tumor are nicely defined.

motor, or sympathetic nerves, accounting for 0.7%-5% of all schwannomas [3]. Although not definitive, adrenal schwannomas are thought to arise from the Schwann cell nerve fibers innervating the adrenal medulla, specifically the vagal and phrenic nerves along with the sympathetic trunk. This tumor tends to affect patients between the ages of 20 and 50 years [4] and is more common in females with a female to male ratio of 1.8:1, consistent with our case's demographic [5]. Most adrenal schwannomas are benign and slow growing but there have been cases of malignancy (<1%); however, there is no biological evidence explaining a correlation between tumor size and malignancy potential [6]. As in this case, benign lesions do not present invasive characteristics or metastases and simply displace surrounding organs.

Most adrenal schwannomas are found incidentally during physical examinations and are referred to as "incidentalomas." Incidentalomas are defined by the European Society of Endocrinology and European Network for the Study of Adrenal Tumors (ESE/ENSAT) as an adrenal mass detected on imaging not performed for suspected adrenal disease [7]. Depending on the cell origin of these adrenal masses, they can be hormone-secreting or nonfunctioning. In our case, adrenal schwannomas are benign lesions; therefore, endocrine function should be within normal ranges. Due to the space in the retroperitoneal cavity, adrenal schwannomas typically grow to large sizes before causing symptoms, most commonly mild abdominal or flank pain. Adrenal schwannomas are challenging to diagnose preoperatively; therefore, pathological analysis is required for diagnostic confirmation. Initial metabolic workup for adrenal schwannomas to rule out a functional mass includes analysis of serum electrolytes, renin, aldosterone, catecholamine, and cortisol. Immunohistochemically, because of their neuroectodermal origin, almost all reported adrenal schwannomas present with strong cytoplasmic and nuclear staining for S100 protein and SOX10 expression [8]. However, some schwannomas may be difficult to distinguish from neurofibromas due to the similar positive S100 protein staining and histological patterns [9]. The presence of a calretinin positive stain reportedly aids in the differentiation between the 2 and allows for a more definitive schwannoma diagnosis [10].

Radiological imaging plays an essential role in treatment planning and management through examining the size, location, and involvement of the tumor with other structures [11]. CT is a valuable imaging modality in the diagnosis and preoperative planning of adrenal schwannomas. The classic ra-

diologic features of primary adrenal schwannoma on CT are a well-circumscribed oval or round unilateral mass with heterogenous soft-tissue density, progressive contrast enhancement, and cystic/necrotic changes [12]. In a review conducted by Xu et al., the sizes of adrenal schwannoma masses range from 0.6 to 14 cm, with an average diameter of 5.5 cm [13]. Additionally, larger masses (>8-10 cm) can present with cystic formation, bleeding, necrosis, and calcification due to secondary degeneration signaling the final stage of long-term tumor growth [12]. Adrenal schwannomas are hypovascular, and the progressive enhancement pattern contributes from the equal involvement of the tissue, septa, and walls of the lesion [11]. In the present case, the tumor was heterogeneously enhancing suggestive of internal hemorrhage and/or necrosis.

On MRI, schwannomas show heterogenous high signal intensity on T2 weighted images with cystic areas, smooth profile, and low signal intensity on T1 weighted images [11]. The tumor in our case displayed heterogenous enhancement with a posterior cystic component and internal enhancement consistent with previous findings of adrenal schwannoma. However, in our case the differential diagnosis after the initial MRI was a malignant adrenal lesion such as adrenocortical carcinoma, possibly due to the large size, heterogenous enhancement, and internal hemorrhage. Adrenocortical carcinoma may be differentiated from schwannoma through its common irregular margins, a lack of capsule, local invasion, and distant metastases [12]. However, both adrenocortical carcinoma and adrenal schwannomas share overlapping features of central tumoral necrosis/hemorrhage, heterogenous enhancement, size greater than 6 cm at presentation, and more common on the left than right adrenal gland, which can lead to diagnostic confusion [14].

Treatment for adrenal masses larger than 6 cm is commonly managed by excision, either laparoscopically or open [15]. The retroperitoneal approach, such as in our case, offers clear access to the adrenal gland and renal hilum, potentially reducing surgery duration and facilitating faster postoperative recovery. In Western medical practice, guidelines from the National Institutes of Health/American Society of Endocrine Surgery recommend that adrenal incidentalomas—those tumors discovered incidentally without presenting symptoms—measuring under 4 cm in diameter, lacking signs of malignancy, and not secreting hormones, should be under surveillance rather than immediate removal [16]. Surgery is advised for tumors that are rapidly increasing in size, have an irregular and uneven shape, and show signs of necrosis or infiltration into surrounding structures. For tumors larger than 6 cm in diameter, proactive surgical intervention is suggested due to the increased risk of complications such as cystic degeneration and a softer texture [16]. Although adrenal tumors typically do not invade adjacent organs, their large size may displace nearby structures. These tumors typically have a favorable prognosis following complete resection; however, 5-10% recur even after complete resection therefore close follow up is required [17]. In the present case the patient did not have severe clinical symptoms; however, given the increase in size of the mass and effect on surrounding organs the mass was excised. Given the rarity of the disease and lack of cases with a preoperative diagnosis, specific guidelines for management of these tumors have yet to be established.

Conclusion

Adrenal schwannomas are a rare manifestation with a lack of characteristic radiological features. These tumors are commonly confused with other adrenal tumors, as they lack specific clinical and imaging features, which leads to difficult clinical management and surgical planning. On imaging, high suspicion of these tumors should be noted if the mass appears as well circumscribed, unilateral, and slow growing with cystic degeneration along with mild or asymptomatic clinical symptoms.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for participation in research, which includes the permission to use data collected in future research projects such as the presented case details and images used in this manuscript.

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