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Case Report
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Primary Adrenal Schwannoma: A Case Report and Review of Literature

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Abstract

Background: Primary adrenal schwannoma is a rare benign tumor that is thought to arise from peripheral nerve sheaths in the adrenal gland. It shares similar characteristics with schwannomas occurring in other anatomic regions, namely the head and neck. Due to its suspicious appearance on most imaging modalities, the tumor is assumed to be malignant until definitively diagnosed after surgical excision.

Case Report: A 60-year-old male was being investigated for an inguinoscrotal mass and was found to have an incidental adrenal mass on computed tomography. There was nonspecific local lymphadenopathy as well as nonspecific pulmonary nodules, raising suspicion for metastatic adrenal carcinoma.

Laboratory workup for hormone-secreting tumor was negative. Magnetic resonance imaging of the adrenals was organized, and confirmed a 54 mm × 55 mm left adrenal mass in contact with the renal cortex, with heterogenous enhancement, lobulated contours, and two enhancing nodules suspicious for metastatic lymphadenopathy.

A presumptive diagnosis of adrenal carcinoma was made by the treating urologist. Laparoscopic radical adrenalectomy was performed under general anesthesia, and required open conversion due to an adhered splenic vein. Pathological analysis after surgical excision revealed a schwannoma, a rare but benign adrenal tumor.

Conclusion: Retroperitoneal schwannomas account for 1% to 3% of all schwannomas and 1% of all retroperitoneal masses, and retroperitoneal schwannomas arising directly from the adrenal gland are even less common. Despite its rarity, schwannoma should be considered in the differential for adrenal incidentalomas due to the lack of precise preoperative diagnostic features.

Keywords: Schwannoma; Adrenal; Tumor; Incidentaloma; Adrenalectomy; Urology; Retroperitoneal

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Abbreviations

CT: Computed Tomography; FNA: Fine Needle Aspiration; MRI: Magnetic Resonance Imaging

Introduction

Primary adrenal schwannoma is a rare benign tumor that is thought to arise from peripheral nerve sheaths in the adrenal gland. It shares similar characteristics with schwannomas occurring in other anatomic regions [1], namely the head and neck [2,3]. Due to its suspicious appearance on most imaging modalities, the tumor is assumed to be malignant until definitively diagnosed after surgical excision [4,5]. We present a case of primary adrenal schwannoma treated at our center, along with a literature review regarding the investigation, diagnosis, and management of adrenal schwannomas.

Case Presentation

Presentation and imaging

A 60-year-old male presented with ultrasound findings of a progressively enlarging left inguinoscrotal mass and 10 lbs of unexplained weight loss, which was regained. Computed Tomography (CT) of the abdomen and pelvis with contrast was performed due to the suspicious history of his weight loss. The inguinoscrotal mass was consistent with a hydrocele; however, CT scan also revealed a 58 mm × 50 mm × 59 mm mass in the left adrenal gland. The adrenal mass had internal calcifications, with probable adjacent lymph nodes and a mildly enlarged left

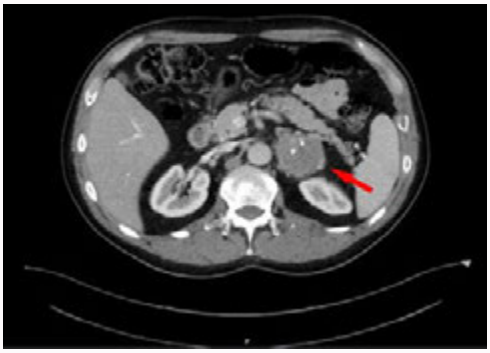

Figure 1: Axial CT with contrast shows the left adrenal incidentaloma measuring 58 mm × 50 mm × 59 mm (red arrow).

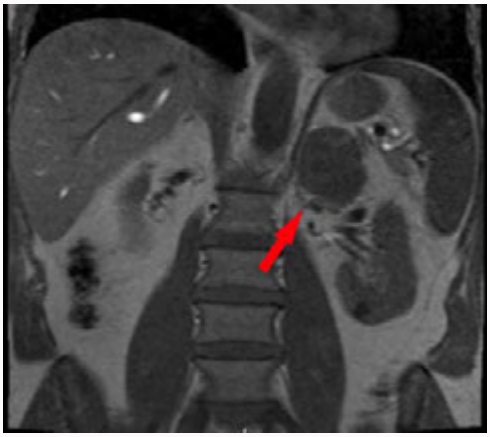

Figure 2: Coronal T1-weighted MRI shows a heterogeneously enhancing left adrenal mass (red arrow).

retrocrural lymph node. Two nonspecific pulmonary nodules were also noted on the CT. The findings were considered suspicious for an adrenal malignancy. Past medical history was significant for Zenker's diverticulum, and surgical and family history were non-contributory (Figure 1, 2).

Investigations

Complete blood count, serum electrolytes, plasma renin, plasma aldosterone, 24-h urine metanephrines and catecholamines, and creatinine clearance were all in their respective normal ranges, indicating a non-functioning tumor. Follow-up chest CT was conducted to investigate the pulmonary nodules, which were located in the lingula and measured ≤ 2 mm. Infectious, inflammatory, and metastatic etiologies were considered.

Magnetic Resonance Imaging (MRI) of the adrenals was organized, and confirmed a 54 mm × 55 mm left adrenal mass in contact with the renal cortex, with heterogenous enhancement, lobulated contours, and two enhancing nodules suspicious for metastatic lymphadenopathy. A presumptive diagnosis of adrenal carcinoma was made, and laparoscopic radical adrenalectomy was planned. Hydrocelectomy was deferred. The patient was counselled regarding the indications, procedure, risks, benefits, and recovery process, and provided informed consent for the operation.

Surgical approach

Laparoscopic adrenalectomy with open conversion was performed under general anesthesia. The patient was placed in the right lateral decubitus position, and was given Cefazolin preoperatively. Intraoperatively, a large mass was visible on the anterior aspect of the left adrenal gland. The adrenal gland was dissected from the spleen and the upper pole of the left kidney using a harmonic scalpel. The mass was adhered to a splenic vein posteriorly, and open conversion was required for complete dissection in order to reduce the risk of bleeding. A subcostal incision was made and the mass was dissected from the vein. The adrenal gland with the mass was completely removed and sent for pathology. The total estimated blood loss was 500 mL which was secondary to some bleeding from accessory adrenal vein. The patient tolerated the procedure well and there were no postoperative complications.

Pathology

The excised specimens consisted of the 55 mm × 45 mm × 41 mm suspected tumor, weighing 62 g, as well as a fragment of adipose containing normal adrenal tissue. The tumor specimen was a firm nodular mass with a whitish-yellow exterior, and a whitish-yellow/gray cut surface. Calcific and hemorrhagic areas were present along with edematous areas. Firm areas representing probable lymph nodes were present within the adipose tissue fragment.

On light microscopy, the nodular mass moderately cellular with spindle cell morphology and no atypia. Nuclei were elongated and the cytoplasm was mildly eosinophilic. Cellular areas were interspersed with edematous and hyalinized areas. Calcific foci and thick-walled blood vessels were present along with occasional lymphocytic infiltrates, and some "ancient" change was noted.

Immunohistochemistry showed positive S-100 and vimentin staining, patchy positive staining for Desmin and Myosin, weak staining for keratin, and negative staining for CD117, Melan-A, HMB-45, chromogranin, synaptophysin, and CD56. A final diagnosis of schwannoma was made, and the dissected lymph nodes had no evidence of malignancy.

Follow-Up

The postoperative period was uneventful and the hydrocele was surgically removed 3-years after the adrenalectomy. The patient was followed up for the past 5 years, he was recently diagnosed with prostate cancer, and will likely undergo radical prostatectomy.

Discussion

Retroperitoneal schwannomas account for 1% to 3% of all schwannomas and 1% of all retroperitoneal masses, and retroperitoneal schwannomas arising directly from the adrenal gland are even less common [3,6]. Similar to most cases of adrenal schwannoma, our patient was undergoing investigation for unexplained weight loss that was regained when the incidentaloma was discovered.

In most cases of schwannoma, laboratory investigations are normal. Typical investigations for adrenal incidentalomas include serum electrolytes, cortisol, adrenocorticotropic hormone, aldosterone, and renin, as well as 24-h urine collection for catecholamines, metanephrines, vanillylmandelic acid, 17-ketosteroids, and 17-hydroxycorticoids [1,6-12]. Complete blood count [6,8,9,11,13] and low dose dexamethasone suppression testing was completed in many cases as well [6-12]. To our knowledge, there have only been three reported cases of catecholamine-secreting retroperitoneal schwannomas in the English literature, of which two were adrenal [14,15] and one was extra-adrenal [16].

CT was the initial mode of discovery for the tumor in our patient,

and MRI was used to further characterize the lesion. We did not conduct Fine Needle Aspiration (FNA) to attempt preoperative diagnosis. FNA is contraindicated unless metastasis is suspected [10], due to the risk of seeding tumor cells or causing severe complications in the case of a pheochromocytoma or echinococcal cyst [1,17]. FNA is a suboptimal diagnostic tool, as demonstrated in a case where two consecutive biopsies failed to identify an adrenal schwannoma [18].

Schwannomas are difficult to diagnose on imaging due to nonspecific features. Similar to our case, calcification and heterogenous enhancement have been observed in previous schwannomas, mimicking diagnoses of adenoma [19,20], adrenocortical carcinoma [11,21,22], pheochromocytoma [21], and distant metastasis [4,7]. To the best of our knowledge, we encountered the first case with nonspecific lymphadenopathy and pulmonary nodules. Lymph node and lung metastasis could not be ruled out, and thus our suspicion for adrenal carcinoma was high.

On MRI, adrenal schwannomas show low intensity on T1-weighted images and heterogeneously high intensity on T2-weighted images; however, these findings are not specific to schwannomas and are common to tumors of neural origin [1,13,23]. Cystic components may be seen on MRI, CT, and ultrasound [15,24,25].

Laparoscopic adrenalectomy is the preferred approach except for large tumors. Some surgeons have successfully removed large tumors laparoscopically, while others have reported conversions to open surgery due to complications [26,27]. Large tumor size is therefore not an absolute contraindication to a laparoscopic approach [28]. Our surgical team converted to an open procedure due to the risk of bleeding from an involved splenic vein. We are aware of only one case where a patient managed non-operatively due to poor surgical candidacy [4].

Adrenal schwannomas are typically well-circumscribed rounded masses, sometimes with a fibrous capsule. Their appearance has been described as tan-yellow to gray-white, often with a homogenous consistency [1]. In correlation with imaging findings, longstanding schwannomas may have cystic areas, calcification, or hemorrhage [19,25,29].

Schwannomas stain positively for S-100, which provides a near-definitive diagnosis in addition to histological features [1]. Desmin, CD34, HMB-45, synaptophysin, chromogranin, and cytokeratin are not expressed [1].

Conclusion

We present a case of adrenal schwannoma with nonspecific local lymphadenopathy, which was initially suspicious for metastatic adrenal carcinoma. Despite its rarity, schwannoma should be considered in the differential for adrenal incidentalomas due to the lack of precise preoperative diagnostic features.

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