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Short Report

Adrenal Schwannoma Treated with Laparoscopic Adrenalectomy: A Case Report and Literature Review

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ABSTRACT

Adrenal schwannoma (AS) is extremely rare. We report on a 36-year-old female patient who presented with right upper abdominal pain. Preoperative computed tomography (CT) showed a 5-cm right adrenal mass. The metabolic work-up was normal. She underwent laparoscopic right adrenalectomy without complications. Microscopy and immunohistochemistry demonstrated a cellular schwannoma. One year after surgery, the patient was fine and without clinical recurrence or metastasis. Usually, the pre-operative diagnosis of AS is challenging and pathologic examination with immunohistochemistry is the only way to make a definite diagnosis.

Keywords: adrenalectomy; adrenal mass; adrenal schwannoma; cellular schwannoma

INTRODUCTION

Schwannoma, which is also known as neurilemmoma, is a common benign peripheral nerve sheath tumor that derives from the Schwann cells of the myelinated neural sheath[1]. The tumor can arise in any area of neural tissue; however, the common locations are the head, neck, mediastinum, and retroperitoneal space[2]. Adrenal schwannoma (AS) is rare and frequently misdiagnosed as adrenal cortical carcinoma[1]. AS is usually a benign slow-growing tumor that is non-functioning; however, some patients can present with either abdominal or flank pain from spontaneous degeneration and hemorrhage[1,3]. We report the case of a 36-year-old female with AS who presented with right-side abdominal pain.

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CASE REPORT

A healthy 36-year-old Thai female presented with right upper abdominal pain radiating to the right flank with no other associated symptoms for 4 days. She reported no history of fever or weight loss. She did not have familial diseases in her family. Her blood pressure was within the normal range, and the general examination was unremarkable. On abdominal examination, there was tenderness at the right upper abdomen. A complete blood count revealed leukocytosis. The white blood cell count was 15,760 cells/μL with a predominance of polymorphonuclear neutrophils (84%). An abdominal computed tomography (CT) demonstrated a 4.6×5.3×4.7 cm enhancing mass near the right adrenal gland with some necrotic areas and several subcentimeter lymph nodes at the para-aortic, aortocaval, retrocaval, and mesenteric regions (Figure 1). A metabolic work-up was performed and the results were negative. The patient underwent transperitoneal laparoscopic right adrenalectomy under general anesthesia. A skin incision of approximately 2 cm was made at the right midclavicular line to place the first trocar (12 mm) as the camera port. The second and third (12-mm and 5-mm) working trocars were placed at the right subcostal line. The fourth 5-mm working trocar was inserted at the right posterior axillary line for liver retraction. The procedure was started by incising the hepatocolic ligament and the peritoneal membrane vertically along the side of the inferior vena cava. The right adrenal vein drains directly to the inferior vena cava, which is a landmark during surgery. This landmark must be identified, controlled, ligated, and cut. The adrenal tumor was dissected from the fat that surrounded the superior and lateral borders. Multiple collateral adrenal vessels were cut using an ultrasonic vessel sealing device (Harmonic®). The tumor capsule was then completely mobilized. The specimen was removed in a bag (Figure 2). No postoperative complications occurred, and the patient was discharged within 4 days.

Pathology demonstrated a schwannoma arising from the right adrenal gland. The surgical specimen was 5.7×5.5×3.8 cm. The cut surfaces were rubbery to firm with a glistening homogenous yellow-white appearance. The histologic features of the well-defined adrenal mass were composed of spindle cells with some degree of nuclear atypia. The mitotic activity was assessed and was found to be low (0–1/10 HPF). Mitotic activity is a measure of the growth rate of a mass, and a high rate is an indication of cancer. Focal secondary reactive lymphoid follicles and peripheral lymphoid cuffs were noted. Hemorrhage and necrosis were absent (Figure 3). An immunohistochemical panel was used for confirmation. S100 showed diffuse and strong positivity. CD117, DOG-1, CD34, desmin, and smooth muscle actin (SMA) were negative (Figure 4).

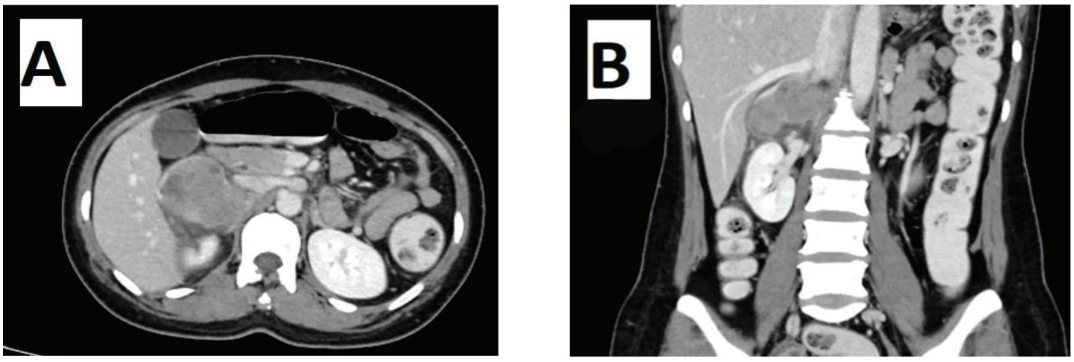


Figure 1 Contrast-enhanced computed tomography: (A) axial image and (B) coronal image show a well-defined enhancing mass epicentered at the anterior to right adrenal gland with some necrotic areas and perilesional fat stranding

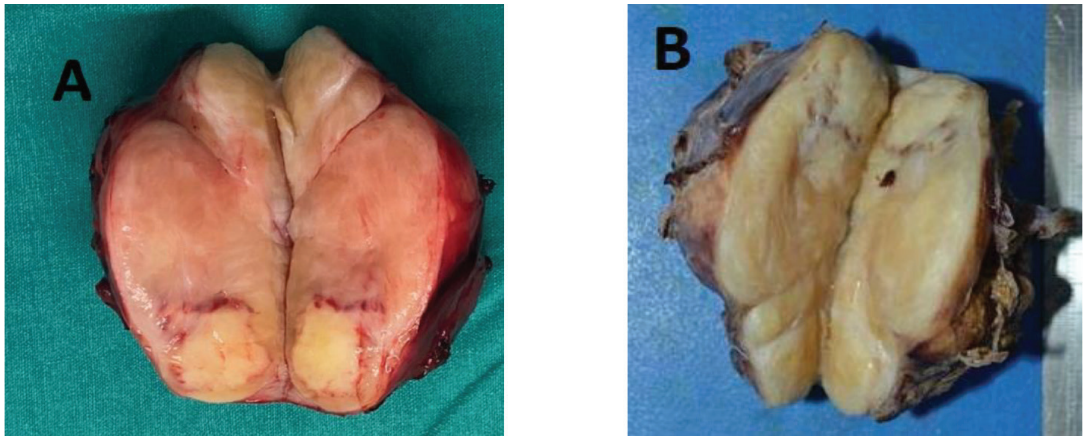
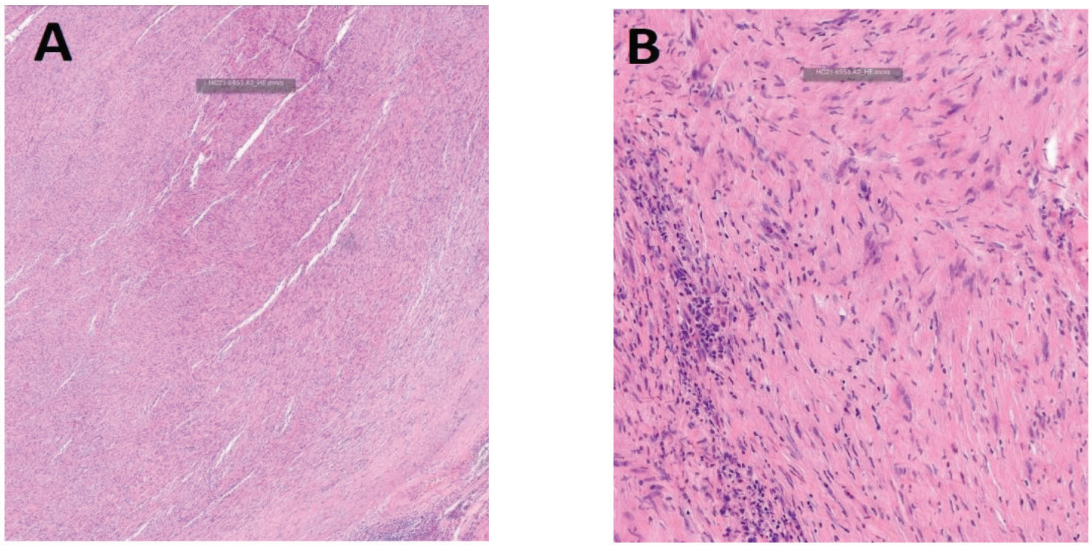


Figure 2 (A) Immediate postoperative fresh tumor mass. (B) Cut surface of the mass showing solid glistening areas



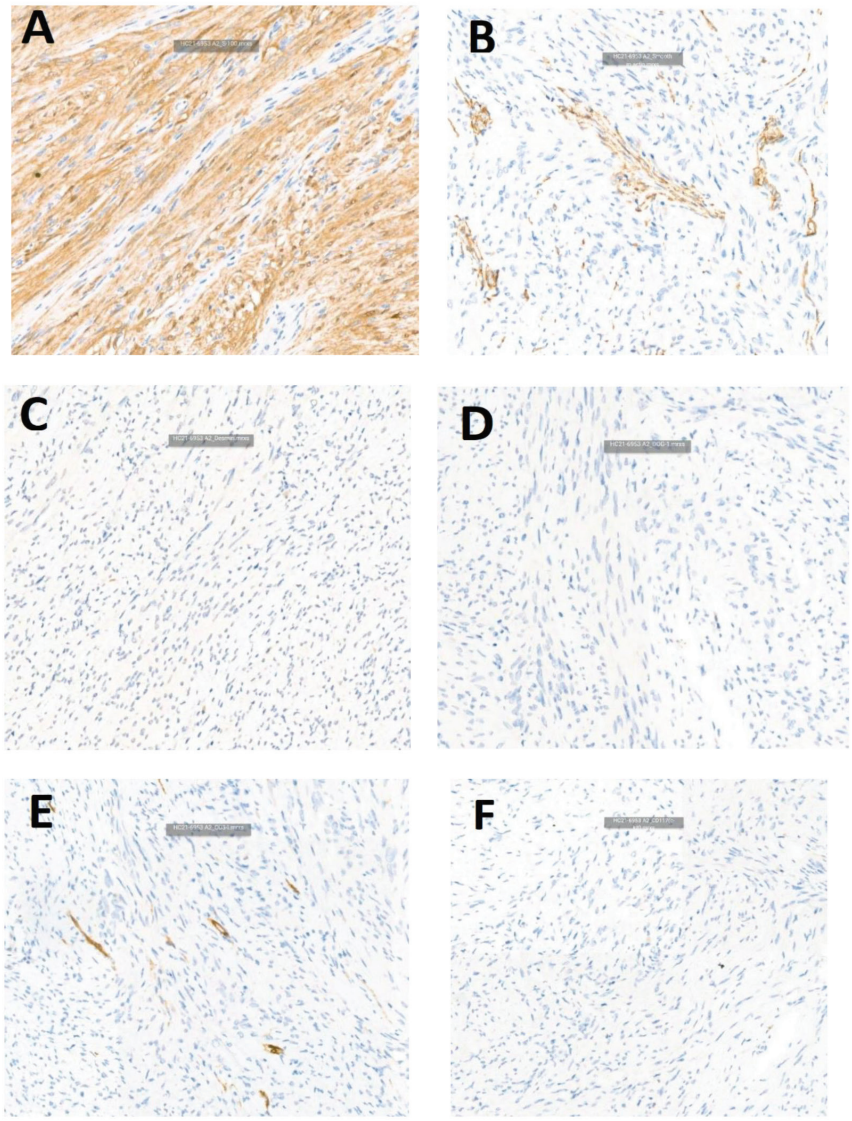
H&E=hematoxylin and eosin

Figure 3 (A) The well-circumscribed mass was composed of loose fascicles of spindle cells with peripheral lymphoid cuff (H&E, 10×). (B) The spindle cells show tapering nuclei with occasional mild nuclear atypia and wavy eosinophilic cytoplasm (H&E, 40×)

DISCUSSION

Adrenal schwannoma is a rare peripheral nerve sheath tumor that generally occurs between the third and sixth decades of life, and is more common in women, with an approximate female-to-male ratio of 1.2:1.[1] A schwannoma rarely occurs in the retroperitoneal space. The majority of cases are sporadic. Genomic analyses have defined neurofibromatosis type 2 and schwannomatosis or Carney complex,[4,5] which have various morphologic appearances and very rarely undergo malignant transformation[5]. About 80 cases of AS have been reported in the literature[2]. AS patients are usually diagnosed with adrenal lesions while imaging for other diseases. Some patients suffer from abdominal pain due to the tumor size or degeneration.

Common incidental findings of AS on CT scans show a well-defined border, homogeneous, an ovoid or spherical



IHC=immunohistochemical staining; DOG-1=discovered on gastrointestinal stromal tumors protein 1; CD=cluster of differentiation

Figure 4 Immunohistochemical findings: (A) diffusely strong S100 positivity (IHC, 40×); (B) negative smooth muscle actin (SMA) (IHC, 40×); (C) negative desmin (IHC, 40×); (D) negative DOG-1 (IHC, 40×); (E) negative CD 34 (IHC, 40×); and (F) negative CD 117 (IHC, 40×)

mass with cystic degeneration or calcification but no invasion or obstruction of the adjacent structures. However, these findings may also be suggestive of malignancy[6,7]. The difference between AS and adrenal adenoma or myelolipoma are the attenuation on unenhanced CT. The average CT attenuation is 30.2 Hounsfield Units (HU) but the CT attenuation is less than 10 HU for adrenal adenoma and myelolipoma. AS demonstrates an early mild heterogeneous contrast enhancement in the arterial phase, and progressive enhancement during the venous phase, with an average CT attenuation of 60 HU. However, there is no consensus criteria for preoperative diagnosis of AS from CT. On magnetic resonance imaging (MRI), AS reveals low signal intensity on T1-WI and heterogeneous high signal intensity on T2-WI. AS has no intracellular lipid or fat tissue. Therefore, AS usually depicts no significant drop

of signal intensity in the out-phase image. From this finding, AS could be mistaken for cysts, pheochromocytomas or adrenocortical carcinoma.

The clinical diagnosis of AS is extremely difficult. The final diagnosis requires a histopathologic evaluation. There is no pathognomonic sign for the diagnosis of AS. The typical characteristics can be evaluated by gross examination, hematoxylin and eosin (H&E) staining, and immunohistochemistry. An AS specimen on gross examination is a well-circumscribed mass that varies in size and is round- to oval-shaped, appearing as a lobulated mass. The outermost layer is an external capsule that has a tan-yellow or grayish-white appearance. The cross-sections are pink or brownish-grey and slightly transparent. The texture is solid and rough with a spiral structure. In some specimens, jelly-like necrosis in focal areas is apparent. Upon H&E staining, spindle cells can be found and may be complicated by hemorrhage, cystic lesions, calcification, hyaline degeneration, and alternating hypercellular Antoni A and hypocellular Antoni B areas. An immunohistochemistry analysis demonstrates strong and diffuse S100, Leu7, laminin, and myelin basic protein. The tumor cells can express CD34, glial fibrillary acidic protein, and cytokeratins, but not epithelial membrane antigen, desmin, or actin[1,10]. Mitotic figures are common, but the mitotic count is <5 mitosis/mm^2 in most cases.

Surgical intervention with minimally invasive surgery is the treatment of choice. However, open surgery is required in some cases when there is a complex mass that mimics malignancy[11,12]. Schwannomas are usually benign tumors, and the prognosis after removal is very good with a very low risk of recurrence. Follow-up care is recommended after removal of the tumor[13].

CONCLUSION

We treated a rare case of AS in a 36-year-old female patient who presented with right abdominal pain. The pre-operative diagnosis of AS is challenging and pathologic examination with immunohistochemistry is so far the only way to make a definite diagnosis. AS is rare; thus, the differential diagnosis includes pheochromocytomas, adrenocortical or lipid-poor adrenal adenoma. Although, AS has benign biological behavior and good prognosis, long-term follow-up of AS is necessary.

CONFLICT OF INTEREST

None of the authors have any disclosures or conflicts of interest to report.

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