

This is an Open Access article licensed under the terms of the Creative Commons Attribution-NonCommercial 3.0 Unported license (CC BY-NC) ([www.karger.com/OA-license](http://www.karger.com/OA-license)), applicable to the online version of the article only. Distribution permitted for non-commercial purposes only.

## A Rare Adrenal Incidentaloma: Adrenal Schwannoma

Mine Adas<sup>a</sup> Filiz Ozulker<sup>b</sup> Gokhan Adas<sup>d</sup> Bora Koc<sup>d</sup> Tamer Ozulker<sup>b</sup>  
Ilknur Mansuroglu Sahin<sup>c</sup>

Departments of <sup>a</sup>Endocrinology, <sup>b</sup>Nuclear Medicine and <sup>c</sup>Pathology, Okmeydani Training and Research Hospital, and <sup>d</sup>Department of General Surgery, Bakirkoy Sadi Konuk Training and Research Hospital, Istanbul, Turkey

### Key Words

Incidentaloma · Schwannoma · Adrenalectomy · Positron emission tomography-computed tomography

### Abstract

Adrenal schwannoma is an extremely uncommon cause of incidentaloma. It originates from neural sheath Schwann cells of the adrenal gland. We report the case of a left adrenal schwannoma incidentally discovered in a 32-year-old woman during examination of bloated feeling and stomach ache. The patient was incidentally found to have a left adrenal mass of 9 cm on abdominal ultrasonography. Computed tomography (CT) of the abdomen and [<sup>18</sup>F] fluorodeoxyglucose positron emission tomography (PET) were also performed. Metabolic evaluation was unremarkable. Due to the large size of the tumor, left adrenalectomy was performed. The postoperative course was uneventful. Histological examination established the diagnosis of schwannoma. This diagnosis was supported by immunohistochemistry of S-100 and vimentin positivity. In conclusion, adrenal schwannoma is an extremely rare entity and can grow considerably in size. The present case report emphasizes that clinicians should be aware of the possibility of retroperitoneal schwannoma. Total excision of benign schwannoma is associated with a favorable outcome. To our knowledge, there are case reports of schwannoma with CT and magnetic resonance imaging findings in the literature, although this is the first schwannoma case with PET-CT imaging.

© 2013 S. Karger AG, Basel

Bora Koc  
Department of General Surgery  
Bakirkoy Sadi Konuk Training and Research Hospital  
TR-34510 Istanbul (Turkey)  
E-Mail [drborakoc@hotmail.com](mailto:drborakoc@hotmail.com)

## Introduction

Schwannomas are infrequent tumors originating from the neural crest cells and usually interpreted as slow-growing benign nerve sheath tumors [1, 2]. Visceral schwannomas are rare and usually discovered serendipitously. Visceral sites of this tumor include the gastrointestinal tract, liver, pancreas, kidney, brain, heart and retroperitoneum. Retroperitoneal schwannomas account for only 1–3% of all schwannomas and for only 1% of all retroperitoneal tumors [3]. The literature reports schwannomas detected using computed tomography (CT) and magnetic resonance imaging (MRI), but we could not find any case of schwannoma detected using positron emission tomography (PET)-CT imaging. Herein, we report a case of large adrenal schwannoma which was diagnosed incidentally and review the cases previously reported in an attempt to clarify the inconsistent features of this rare disease.

## Case Report

A 32-year-old female presented with new onset of intermittent right flank pain. During examination, a left adrenal mass was found incidentally on abdominal ultrasonography and she was referred to our institution for additional evaluation. Her medical and family histories were unremarkable. On physical examination the abdomen was soft, blood pressure was 130/85 mm Hg and pulse rate was 72 beats/min; sinus rhythm was normal. Blood count and biochemistry analysis were within normal limits.

A heterogeneously enhancing soft tissue mass on the medial aspect of the left adrenal gland measuring 65 × 95 mm was visualized and no additional mesenteric or retroperitoneal lymphadenopathy was identified on the abdominal CT scans. As the mass was approximately 10 cm in size, [<sup>18</sup>F] fluorodeoxyglucose (<sup>18</sup>FDG) PET-CT was also performed. It showed intense uptake within the left adrenal nodule with a standardized uptake value (SUV<sub>max</sub>) of 9.5, indicative of malignancy (fig. 1). Endocrinological examinations including serum electrolytes, cortisol, urinary metanephrine, normetanephrine, adrenalin, noradrenalin and vanillyl mandelic acid, as well as plasma renin and aldosterone were within normal range. Also, a dexamethasone suppression test showed values within the normal range at onset and at 30 min. Due to the large size of the tumor, standard transperitoneal adrenalectomy was performed without complications. Postoperative pathological evaluation revealed schwannoma in the adrenal gland, a hard, well-circumscribed, encapsulated mass 4 × 8 × 10 cm in overall dimensions that was grossly compressing the adrenal parenchyma. Histologically, the tumor consisted of spindle cells with alternating areas of compact hypercellularity with irregular streams and without atypia or mitosis. Immunohistochemical analysis demonstrated rare Ki67-positive cells (<5%) while SMA, desmin, CD34 and CD117 staining was negative in tumor cells. In contrast to these results, S-100 and vimentin staining were diffusely positive across the tumor. Thus, the evidences corresponded to a benign adrenal schwannoma type Antoni A (fig. 2, fig. 3, fig. 4, fig. 5). The patient was discharged uneventfully on the sixth day after surgery.

## Discussion

Schwannomas are slow-growing benign nerve sheath tumors whose principal components originate from crest cells, the vestibulocochlear nerve being the most frequent site [1,

4]. They are most often found within the head, the neck as well as the upper and lower extremities, and with a lesser frequency on the trunk, in the gastrointestinal tract, liver, pancreas, kidney, brain, heart and retroperitoneum [5]. These rare tumors account for only 1–5% of retroperitoneal masses and comprise only 1–3% of all schwannomas [6–8]. This incidence increases to 7% in patients older than 70 years [9]. Nevertheless only 0.7% of benign and 1.7% of malignant schwannomas occur in the retroperitoneum [8]. These tumors are usually characterized by a benign course, are encapsulated and vary from firm solitary masses to fluctuant cysts. Schwannomas occasionally occur in cases with Cushing syndrome and von Recklinghausen disease. Schwannomas were initially described by Verocay in 1908; after this description Antoni had performed histological subclassification into two patterns [10]. Histologically, these tumors consist of compact hypercellularity with irregular streams (Antoni type A) and loose hypocellularity with cystic spaces (Antoni type B). Investigators believe that Schwann cells of nerve fibers innervating the medulla are the source of these adrenal lesions [11–13].

Functional adrenocortical tumors can be easily revealed by characteristic hormone-related symptoms, blood chemistry or urinary tests. However, non-functional adrenocortical tumors present no characteristic lesions. Due to their non-secreting and asymptomatic properties, adrenal schwannomas are often diagnosed incidentally. Moreover, increasing medical examination health screening and advances in imaging modalities give rise to an increased incidental detection of retroperitoneal tumors. CT scan shows a well-circumscribed, homogeneous, round-oval mass with cystic degeneration or calcification [4]. MRI findings are usually non-specific, but helpful in the differential diagnosis of solid non-functional tumors. Solid tumors with a low signal intensity on T1WI and heterogeneously high intensity on T2WI are revealed on MRI [14]. <sup>18</sup>FDG PET has previously been suggested as valuable in discriminating benign from malignant adrenal masses. The concept of <sup>18</sup>FDG PET is based on an increased glucose uptake by malignant lesions. The quantitative analysis of FDG uptake is performed using standardized SUVs or by qualitative visual evaluation with respect to liver uptake [15]. The sensitivity of FDG-PET in identifying malignant lesions varied between 93 and 100% with a specificity between 80 and 100% [16–20]. The sensitivity of <sup>18</sup>FDG PET imaging is only moderate, however, for the diagnosis of small lesions, and false-positive results have to be considered. In our case, the huge size of the tumor and the location and also the characteristics of the lesion on imaging were strongly suspicious of a malignant adrenal neoplasm. Whereas some of the characteristics of retroperitoneal schwannomas, including heterogeneity and degeneration, may mimic malignancy on radiological interpretation, a preoperative diagnosis of adrenal schwannoma is almost impossible and usually should be confirmed by histopathological examination as seen in our patient. It should not be forgotten that the malignant form is frequently associated with von Recklinghausen disease and neurofibromatosis [21]. In consequence of significant risks, fine needle aspiration should be performed only in patients with a history of carcinoma or suspected metastasis [2].

Quite often postoperative recovery is uneventful. It should be noted that the diagnosis will remain unclear until after surgical intervention. Furthermore, physicians should consider the possibility of local recurrence and malignant formation in benign schwannomas despite a prior benign diagnosis [22]. Therefore, it is very important to remove the tumor completely. A non-secreting adrenal mass larger than 4 cm should be removed surgically with the added benefit of a definitive diagnosis. In cases of huge tumors as our case, the conventional approach can be preferred to laparoscopic resection.

Pathological, histological and immunohistochemical findings determine the definitive diagnosis. Histologically, neoplastic cells that simulate the appearance of differentiated Schwann cells organized as cellular areas with nuclear palisading (Antoni A) and loose,

hypocellular, myxoid lesions with microcystic spaces (Antoni B) are seen. In our case the specimen consisted of spindle cells with alternating areas of compact hypercellularity with irregular streams and without atypia or mitosis. Therefore the pathologist suspected benign adrenal schwannoma. However, the ultimate result should provide secondary confirmation of histology with immunohistochemical staining. Immunohistochemically, these lesions stain positively for S-100 antigen, vimentin, collagen IV and laminin and negatively for keratin, desmin, actin, CD34 and CD117 [2–9, 23]. Fine et al. [24] demonstrated that a positive stain for calretinin, the same protein family as S-100, which expressed for schwannoma but not for neurofibroma, can be used for the discrimination of these two diseases.

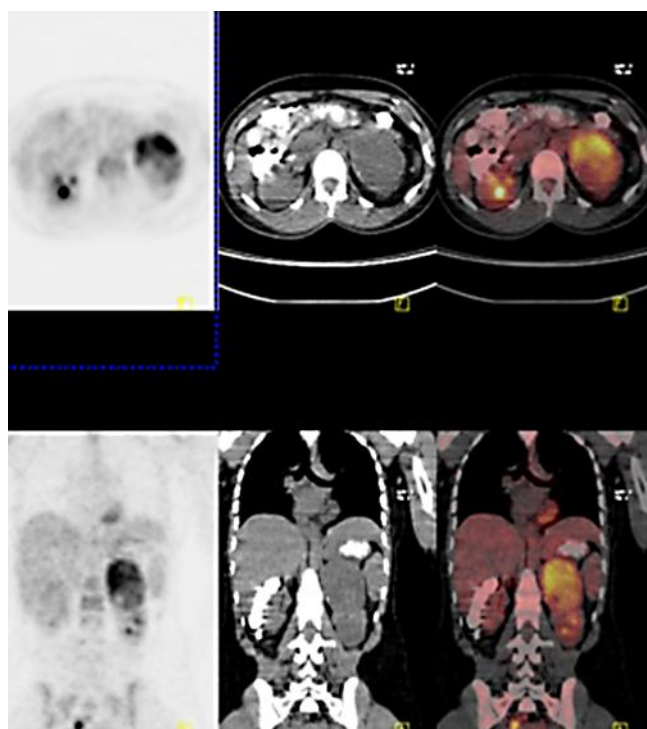
## Conclusion

We report the case of an incidentally discovered schwannoma difficult to diagnose in preoperative evaluation. Because of the lack of definitive non-histological diagnostic modalities in this rare tumor, surgical excision can provide the benefit of a definitive diagnosis. Consequently, a definitive diagnosis can only be made by histological and immunohistochemical evaluations.

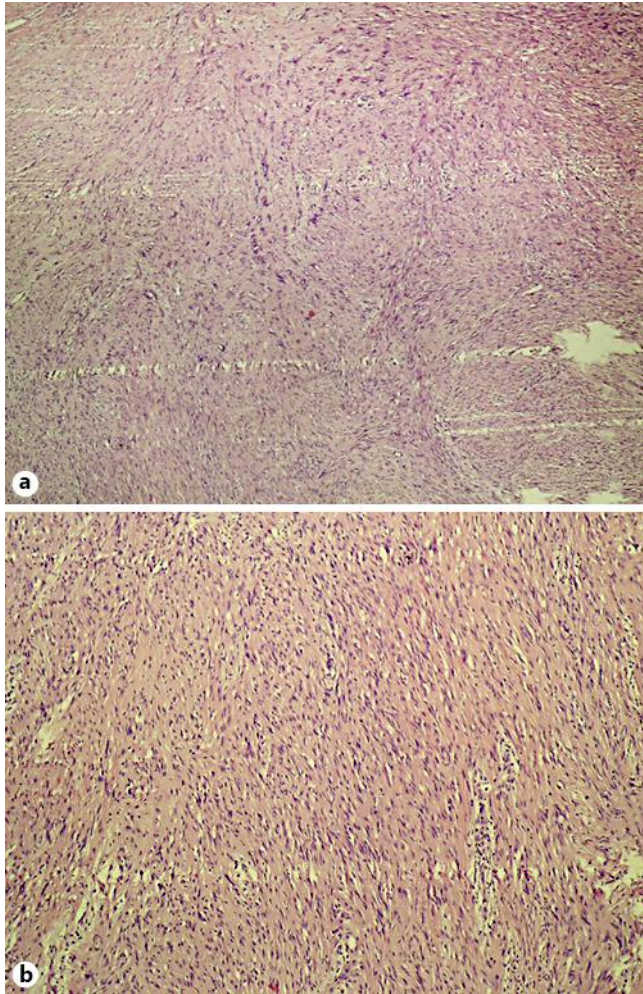
## References

- 1 Behrend M, Kaaden S, von Wasielewski R, Frericks B: Benign retroperitoneal schwannoma mimicking an adrenal mass. *Surg Laparosc Endosc Percutan Tech* 2003;13:133–138.
- 2 Quayle FJ, Spittle JA, Pierce RA, Lairmore TC, Moley JF, Brunt LM: Needle biopsy of incidentally discovered adrenal masses is rarely informative and potentially hazardous. *Surgery* 2007;142:497–504.
- 3 Gubbay AD, Moschilla G, Gray BN, Thompson I: Retroperitoneal schwannoma: a case series and review. *Aust N Z J Surg* 1995;65:197–200.
- 4 Târcoveanu E, Dimofte G, Bradea C, Moldovanu R, Vasilescu A, Anton R, Ferariu D: Adrenal schwannoma. *JLS* 2009;13:116–119.
- 5 Sharma SK, Koleski FC, Husain AN, Albala DM, Turk TMT: Retroperitoneal schwannoma mimicking an adrenal lesion. *World J Urol* 2002;20:232–233.
- 6 Scanlan DB: Primary retroperitoneal tumors. *J Urol* 1959;81:740–745.
- 7 Das Gupta TK, Brasfield RD, Strong EW, Hajdu SI: Benign solitary schwannomas (neurilemmomas). *Cancer* 1969;24:355–366.
- 8 Whitaker WG, Droulias C: Benign encapsulated neurilemoma: a report of 76 cases. *Am Surg* 1976;42:675–678.
- 9 Grumbach MM, Biller BM, Braunstein GD, Campbell KK, Carney JA, Godley PA, Harris EL, Lee JK, Oertel YC, Posner MC, Schlechte JA, Wieand HS: Management of the clinically inapparent adrenal mass ('incidentaloma'). *Ann Intern Med* 2003;138:424–429.
- 10 Korets R, Berkenblit R, Ghavamian R: Incidentally discovered adrenal schwannoma. *JLS* 2007;11:113–115.
- 11 Suzuki K, Nakanishi A, Kurosaki Y, Nogaki J, Takaba E: Adrenal schwannoma: CT and MRI findings. *Radiat Med* 2007;25:299–302.
- 12 Lau SK, Spagnolo DV, Weiss LM: Schwannoma of the adrenal gland. *Am J Surg Pathol* 2006;30:630–634.
- 13 Yang CY, Chou CW, Lin MB, Li CF: Schwannomas of the left adrenal gland and posterior mediastinum. *J Chin Med Assoc* 2009;72:83–87.
- 14 Onoda N, Ishikawa T, Toyokawa T, Takashima T, Wakasa K, Hirakawa K: Adrenal schwannoma treated with laparoscopic surgery. *JLS* 2008;12:420–425.
- 15 Terzolo M, Stigliano A, Chiodini I, Loli P, Furlani L, Arnaldi G, Reimondo G, Pia A, Toscano V, Zini M, Borretta G, Papini E, Garofalo P, Allolio B, Dupas B, Mantero F, Tabarin A; Italian Association of Clinical Endocrinologists: AME position statement on adrenal incidentaloma. *Eur J Endocrinol* 2011;164:851–870.
- 16 Boland GW, Goldberg MA, Lee MJ, Mayo-Smith WW, Dixon J, McNicholas MM, Mueller PR: Indeterminate adrenal mass in patients with cancer: evaluation at PET with 2-[F-18]-fluoro-2-deoxy-D-glucose. *Radiology* 1995;194:131–134.
- 17 Erasmus JJ, Patz EF Jr, McAdams HP, Murray JG, Herndon J, Coleman RE, Goodman PC: Evaluation of adrenal masses in patients with bronchogenic carcinoma using <sup>18</sup>F-fluorodeoxyglucose positron emission tomography. *AJR Am J Roentgenol* 1997;168:1357–1360.

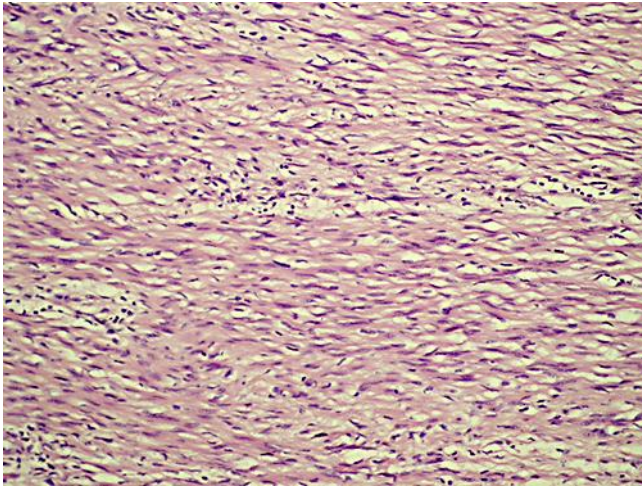
- 18 Maurea S, Mainolfi C, Bazzicalupo L, Panico MR, Imparato C, Alfano B, Ziviello M, Salvatore M: Imaging of adrenal tumors using FDG PET: comparison of benign and malignant lesions. *AJR Am J Roentgenol* 1999;173:25–29.
- 19 Yun M, Kim W, Alnafisi N, Lacorte L, Jang S, Alavi A: <sup>18</sup>F-FDG PET in characterizing adrenal lesions detected on CT or MRI. *J Nucl Med* 2001;42:1795–1799.
- 20 Tenenbaum F, Groussin L, Foehrenbach H, Tissier F, Gouya H, Bertherat J, Dousset B, Legmann P, Richard B, Bertagna X: 18F-fluorodeoxyglucose positron emission tomography as a diagnostic tool for malignancy of adrenocortical tumours? Preliminary results in 13 consecutive patients. *Eur J Endocrinol* 2004;150:789–792.
- 21 Garcia G, Anfossi E, Prost J, Ragni E, Richaud C, Rossi D: Benign retroperitoneal schwannoma: report of three cases. *Prog Urol* 2002;12:450–453.
- 22 Nishio A, Adachi W, Igarashi J, Koide N, Kajikawa S, Amano J: Laparoscopic resection of a retroperitoneal schwannoma. *Surg Laparosc Endosc Percutan Tech* 1999;9:306–309.
- 23 Hettiarachchi JA, Finkelstein MP, Schwartz AM, Johnson GB, Konno S, Choudhury MS: Benign retroperitoneal schwannoma presenting as a giant adrenal tumor. *Urol Int* 2003;14:356–360.
- 24 Fine SW, McClain SA, Li M: Immunohistochemical staining for calretinin is useful for differentiating schwannomas from neurofibromas. *Am J Clin Pathol* 2004;122:552–559.



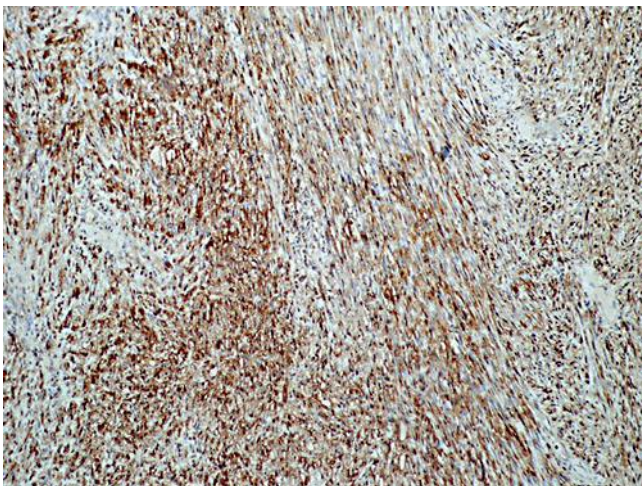
**Fig. 1.** <sup>18</sup>F-FDG PET-CT showed intense uptake within the left adrenal nodule with a standardized SUV<sub>max</sub> of 9.5, indicative of malignancy.



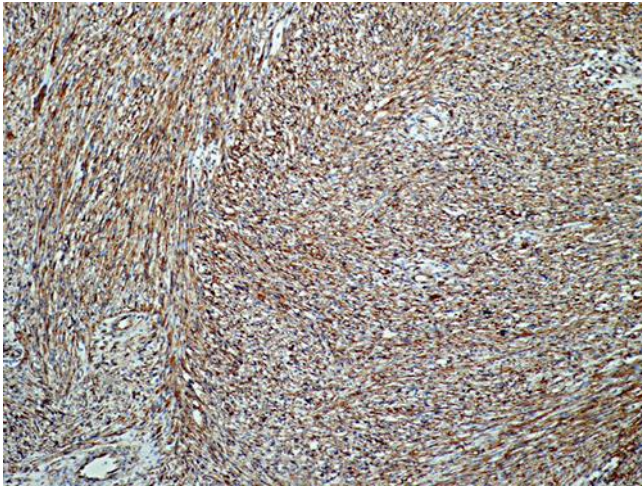
**Fig. 2.** Spindle cells in an Antoni type A area; large hyperchromatic nucleus, Antoni B areas consisting of spindle cells within a loose myxoid matrix (**a** H&E,  $\times 40$ ; **b** H&E,  $\times 100$ ).



**Fig. 3.** Spindle cells in myxomatous matrix (H&E, ×200).



**Fig. 4.** Positive S-100 immunohistochemical staining (×100).



**Fig. 5.** Positive vimentin immunohistochemical staining ( $\times 100$ ).