

## CASE REPORT

# A rare combination of giant right retroperitoneal schwannoma and right adrenal oncocytoma as an incidental finding

Michael Hollis<sup>1,\*</sup>, Chibueze Onyemkpa<sup>1</sup> and Tolutope Oyasiji<sup>1,2</sup>

<sup>1</sup>Department of General Surgery, Michigan State University, Lansing, MI 48912, USA, and <sup>2</sup>Department of Surgical Oncology, Barbara Ann Karmanos Cancer Institute at McLaren Flint, Wayne State University, Flint, MI 48532, USA

\*Correspondence address. Department of General Surgery, Michigan State University, Lansing, MI 48912, USA. E-mail: Michael.Hollis@hc.msu.edu

## Abstract

Adrenal oncocytomas and retroperitoneal schwannomas are two groups of very rare tumors with distinct histologic features. Both tumors are usually incidentally found and are mostly benign. Optimal management is usually with surgical resection. We report a unique case of a 74-year-old woman who was incidentally found to have these two exceptionally rare tumors at the same time. She was successfully managed with surgical resection of the retroperitoneal schwannoma and adrenalectomy for the adrenal oncocytoma and both diagnoses were confirmed with histopathology and immunochemistry.

## INTRODUCTION

The retroperitoneum is a complex space containing multiple structures and as a result, it is prone to a wide range of tumors of different origins [1]. Retroperitoneal schwannomas and adrenal oncocytomas are examples of such tumors. A review of the published literature showed that <200 cases of adrenal oncocytoma have been reported and they are estimated to constitute ~1.8% of adrenal tumors [2, 3]. Retroperitoneal schwannomas also make up <2% of all schwannomas and ~1% of retroperitoneal neoplasms [1]. These tumors are, therefore, very uncommon and although they have been discussed in the literature, no reports of both tumors existing together have been reported. We present a case of these rare tumors occurring at the same time in a patient.

## CASE REPORT

A 74-year-old female was referred to the surgical oncology clinic following an incidental finding of a right retroperitoneal

mass initially identified on transthoracic echocardiogram. As part of the work up for this mass, a computerized tomography scan of the abdomen and pelvis was obtained. The CT scan revealed an 18 cm × 14 cm mass in the right retroperitoneum. The mass was noted to displace the right kidney inferiorly and medially (Fig. 1). An additional 6.1 cm × 4.2 cm ovoid solid lesion was also seen posterosuperior to the larger retroperitoneal mass and reported as a possible right adrenal mass or exophytic hepatic mass (Fig. 2).

A full biochemical panel for adrenal incidentalomas was done and the tumor was found to be non-functioning. Subsequent to this, a CT-guided biopsy was performed for the masses. The larger, right retroperitoneal mass returned a diagnosis of neurilemmoma (schwannoma). The posterosuperior mass was also biopsied and the findings were reported as patchy hepatocyte atypia which was inconclusive of a definitive diagnosis.

Intra-operatively, significant findings included a huge right retroperitoneal mass which displaced the right kidney infero-

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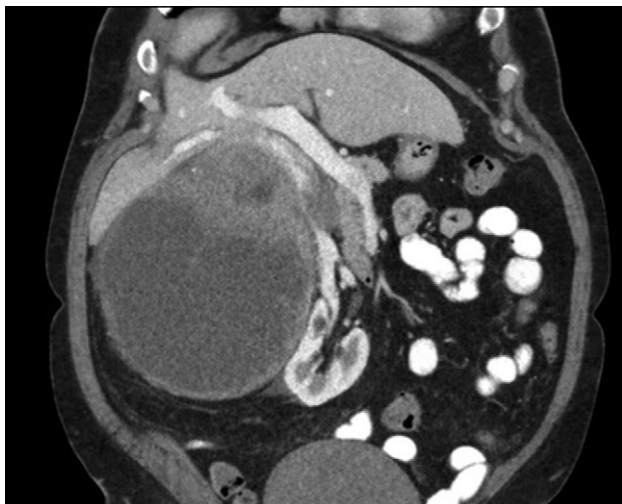


Figure 1: CT abdomen and pelvis showing the right retroperitoneal mass displacing the right kidney inferomedially.

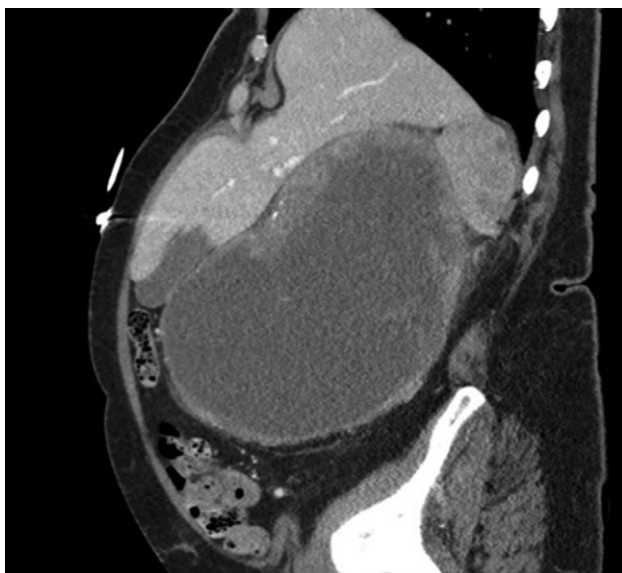


Figure 2: CT abdomen and pelvis showing the additional mass which is posteriosuperior to the large retroperitoneal mass.

medially. This mass was adherent to the right lateral aspect of the IVC and the right renal vein coursed on its anteromedial surface. The mass was carefully mobilized away from the IVC without any violation of its capsule. The right renal vessels were also salvaged thereby ensuring preservation of kidney. The mass was successfully removed with its capsule intact. A completely separate and encapsulated mass was identified posteriosuperior to the larger mass. This mass had vessels directly communicating with the IVC.

Pathologic analysis revealed that the large right retroperitoneal mass measured 22 cm × 15 cm × 7 cm and weighed 2289 g (Fig. 3). Histologically, the tumor showed spindle cells with focal nuclear palisading, nests of foamy histiocytes, patchy chronic inflammation and rare enlarged polymorphic nuclei. No mitosis was identified (Fig. 4). With immunohistochemical staining, the tumor cells were positive for S-100



Figure 3: Gross picture of the right retroperitoneal schwannoma.

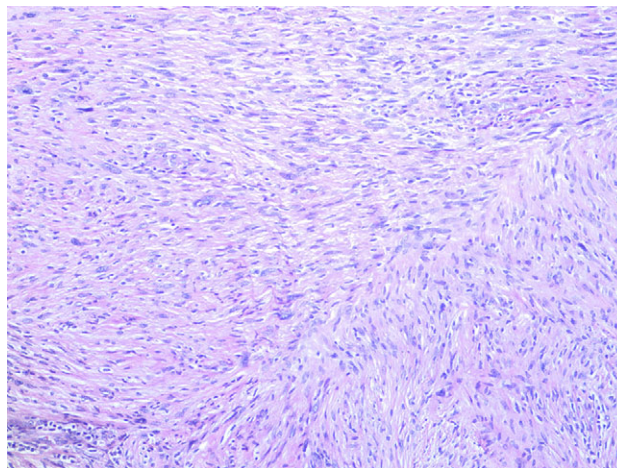


Figure 4: Histology picture for right retroperitoneal schwannoma.

(Fig. 5) and vimentin but negative for CD34, CD117, SMA, desmin and pancytokeratin. Both morphology and immunostains were consistent with neurilemmoma (schwannoma).

The smaller right retroperitoneal tumor weighed 177 grams and measured 10 cm × 8 cm × 5 cm (Fig. 6). Histopathology showed an oncocytic neoplasm with focal Zellballen growth pattern, abscess formation and recent hemorrhage (Fig. 7). The tumor cells were strongly positive for NSE, focally positive for S-100 but were negative for inhibin, Hepatic Specific Antigen (HSA), pancytokeratin, PAX8, EMA, chromogranin-A and synaptophysin. Ki-67 stained ~5% of tumor nuclei (Fig. 8). The final diagnosis was an oncocytic variant of adrenal cortical adenoma (oncocytoma).

## DISCUSSION

We report a case of a very rare combination of a giant right retroperitoneal schwannoma and right adrenal oncocytoma.

Schwannomas are typically benign, slow-growing tumors comprised of Schwann cells of the peripheral nerve sheath [1, 4]. These tumors mostly affect patients between 20 and 50 years old, with greater preponderance for the female gender [1]. Commonly found in the head, neck and flexor surfaces, occurrence in the retroperitoneum is rare and accounts for <2% of all

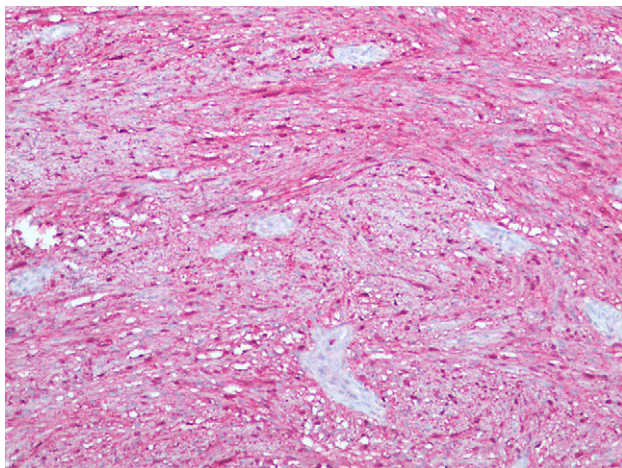


Figure 5: High power view confirms uniform cells with oncocytic features (H&E; original magnification  $\times 40$ ).

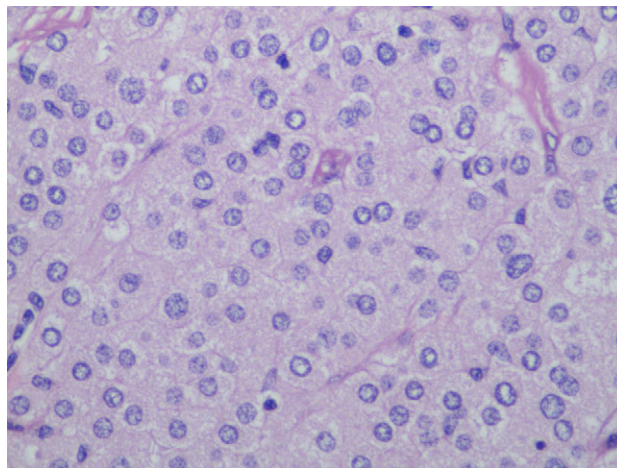


Figure 7: Histology picture for right adrenal oncocytoma.

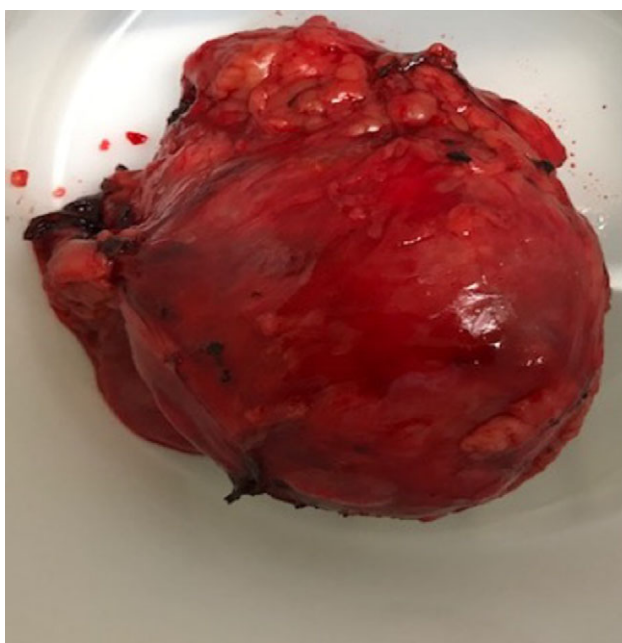


Figure 6: Gross picture of the right adrenal oncocytoma.

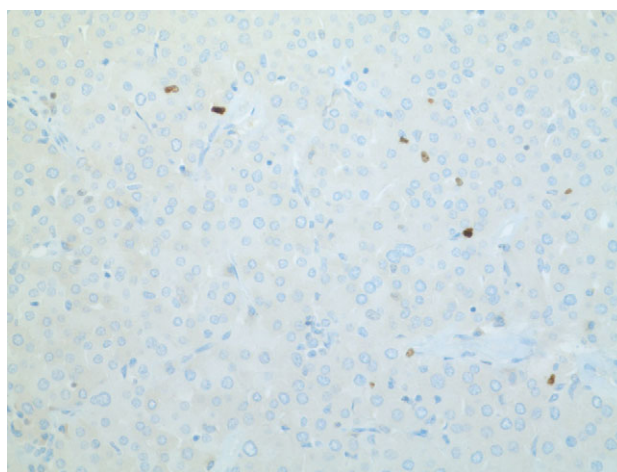


Figure 8: A Ki-67 immunostain shows sparse positive nuclear staining confirming the low proliferative index of adenoma (Ki-67; original magnification  $\times 20$ ).

Schwannomas [4]. These tumors are typically asymptomatic in the retroperitoneum and are usually incidental findings [1, 4]. Symptoms, when present, are vague and non-specific. [4].

Oncocytomas, on the other hand, are neoplasms made up of epithelial cells with abundant granular cytoplasm due to abnormal mitochondrial accumulation [3, 5]. They most commonly occur in the kidney, salivary glands and thyroid gland, but have also been reported in the parathyroid, pituitary, liver, ovary and small intestine [3, 5]. Given the rarity, with <200 cases reported in literature, the true epidemiology of adrenal oncocytomas is unknown [3]. These tumors arise from the adrenal cortex and are typically large, benign and non-functional [2]. However, data suggest that ~10–30% are functional and may release one or more adrenal hormones [2].

Although various syndromes include schwannomas, none to the best of our knowledge includes retroperitoneal

schwannoma and adrenal oncocytoma as part of the syndrome. However, it is noteworthy to mention Carney syndrome—a rare and dominantly inherited syndrome associated with schwannomas, and endocrine tumors [6]. Carney syndrome, however, does not include adrenal oncocytoma. Other significant components of the syndrome were conspicuously absent in the index case [6]. Retroperitoneal schwannomas are commonly found in Neurofibromatosis type I, however, adrenal oncocytoma is not a feature of this disease process. To this end, this patient was approached as one with two isolated rare tumors.

To facilitate diagnosis, various imaging modalities are utilized. While MRI is the imaging modality of choice for schwannomas, adrenal oncocytomas have no characteristic radiological features [4, 5]. On MRI, schwannomas possess characteristic ‘fascicular sign’, which refers to the appearance of bundles along the nerve sheath, and the ‘target sign’, caused by a hypointense center surrounded by a hyperintense periphery on T2 weighted MRI images [4]. It is pertinent to note that these distinguishing features are usually not seen in schwannomas found in the retroperitoneum and in the index case, these distinguishing features were not identified on MRI [4].

In addition to histologic analysis, immunohistochemical stains are also used to characterize both tumors. As in the patient discussed, retroperitoneal schwannomas stain positively for S-100 and negatively for CD 34 on immunohistochemistry [1]. The tumor in the index patient also stained positively for vimentin. Adrenal oncocytomas on the other hand are usually positive for vimentin, synaptophysin, alpha inhibin and melan-A, but negative for chromogranin-A and S-100 [3].

Both tumors are mostly benign in behavior. Malignant transformation in schwannoma is rare—occurs in ~5%—and are usually observed in individuals with neurofibromatosis type 1 [7]. In these situations, they behave like high-grade sarcomas. Malignant features in adrenal oncocytomas are determined using the Lin-Weiss-Bisceglia criteria [2, 3]. In either case, the presence of a malignant feature worsens prognosis. Histologically, our patient presented with benign features and in this instance, surgical resection is the treatment of choice with no need for adjuvant treatment [2, 7]. Schwannomas are generally treated by surgical resection while adrenal oncocytomas are treated with adrenalectomy [1, 5].

Although the index patient was incidentally found to have two rare tumors, there is a possibility this by chance and a very rare occurrence. In our review of literature, this is the first case documenting the simultaneous presence of both tumors and no prior syndromic associations have been noted. Since no genetic associations were suspected, the patient did not undergo any genetic testing.

## ACKNOWLEDGEMENTS

None.

## CONFLICT OF INTEREST STATEMENT

None of the authors have any forms of conflicts of interest.

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## ETHICAL APPROVAL

This case report was approved by the institute's Institutional Review Board as per its policy.

## CONSENT

Appropriate consent was obtained from the patient.

## GUARANTOR

Tolutope Oyasiji.

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