Case Report

A case series of two cases of juxta-adrenal schwannoma presenting as adrenal mass lesion and review of the literature

Shivashankar Damodaran, Griffin Mahimairaj, Kamaraj Velaichamy

Department of Urology, Madras Medical College, Rajiv Gandhi Government General Hospital, Chennai, Tamil Nadu, India

Abstract

Schwannomas are rare tumors in the retroperitoneal location. They can pose a diagnostic dilemma when presenting as an adrenal mass lesion due to their imaging characteristics. We report two cases of juxta-adrenal schwannomas presenting as symptomatic adrenal mass lesions. In both the cases, the clinical examination and functional evaluation was unremarkable and the radiological examination revealed a mixed intense adrenal mass lesion in one case with predominantly hyperintense areas and a very hyperintense lesion in another, in T2-weighted images, mimicking a adrenocortical malignancy and a pheochromocytoma respectively. Both cases were treated by surgical excision. Histopathological examination established the correct diagnosis of schwannoma, which was confirmed by immunohistochemical staining. Juxta-adrenal schwannoma is rare tumors of the retroperitoneum, which should also be borne in mind whenever encountering large nonsecreting adrenal tumors. We report a unique imaging characteristic, which helps in preoperative identification these rare lesions.

Key Words: Adrenal mass lesions, juxta-adrenal schwannoma, symptomatic

Address for correspondence:
Dr. Shivashankar Damodaran, Department of Urology, Madras Medical College, Rajiv Gandhi Government General Hospital, Park Town, Chennai - 600 003, Tamil Nadu, India. E-mail: siva.shankar993@gmail.com
Received: 22.08.2014, **Accepted:** 13.01.2015

INTRODUCTION

Schwnnomas are rare tumors, usually benign and arising from Schwann sheath of the peripheral or cranial nerves. Retroperitoneal and juxta-adrenal schwannomas are extremely rare and may appear preoperatively as giant adrenal tumors, and only a few cases have been reported in the literature.[1] They usually present as asymptomatic mass lesions. In this case series, we report two cases that presented as symptomatic adrenal mass lesions. We also mention about a new imaging feature, which helps in preoperative identification of these lesions.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/0974-7796.152940

CASE REPORT

Case 1

A 26-year-old female came with dull aching right loin pain. Clinical examination was noncontributory. An abdominal ultrasound showed eight by nine cm mixed echogenic mass in left suprarenal region. Metabolic evaluation including plasma metanephrines, cortisol, serum electrolytes was within normal limits. Magnetic resonance imaging (MRI) showed mixed intense solid mass with predominantly hyperintense areas, of size eight by nine cm in the left adrenal gland [Figures 1 and 2]. Based on the clinical and radiological features we had a strong suspicion of an adrenocortical carcinoma and did an open adrenalectomy [Figures 3-6]. Postoperative period was uneventful. Histopathological examination Showed features

of schwannoma [Figure 7] in a juxta-adrenal location. Immunohistochemistry (IHC) confirmed the diagnosis and was positive for S-100 [Figure 8] and negative for vimentin.

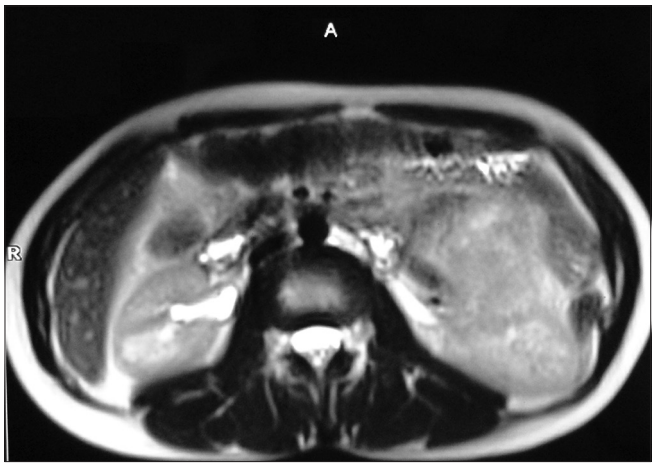

Figure 1: Magnetic resonance imaging showing left adrenal mass lesion

Case 2

A 50-year-old female came with dull aching left loin pain for 2 months. Clinical examination was noncontributory. An

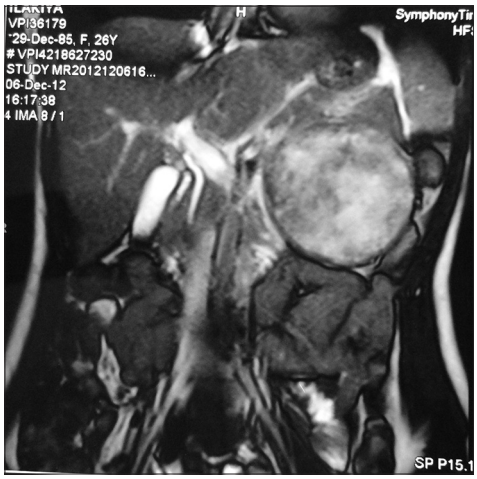

Figure 2: Coronal section of the adrenal mass showing mixed intense mass lesion

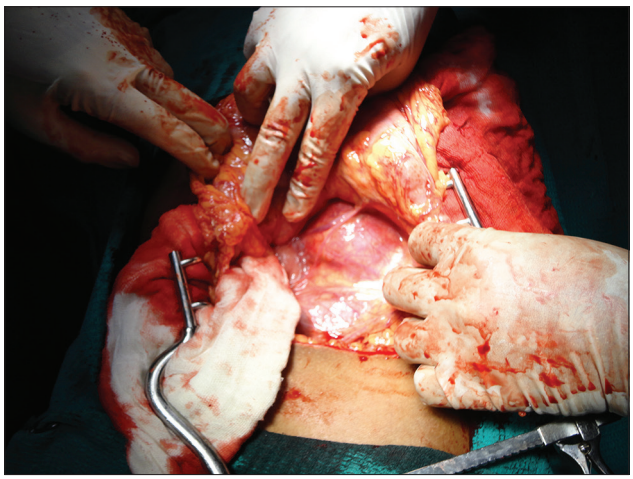

Figure 3: Left suprarenal mass exposed

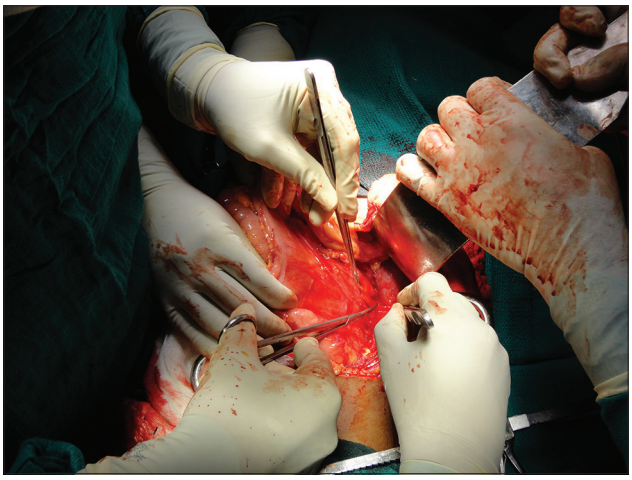

Figure 4: Dissection of the left suprarenal mass, after ligating the feeding vessels

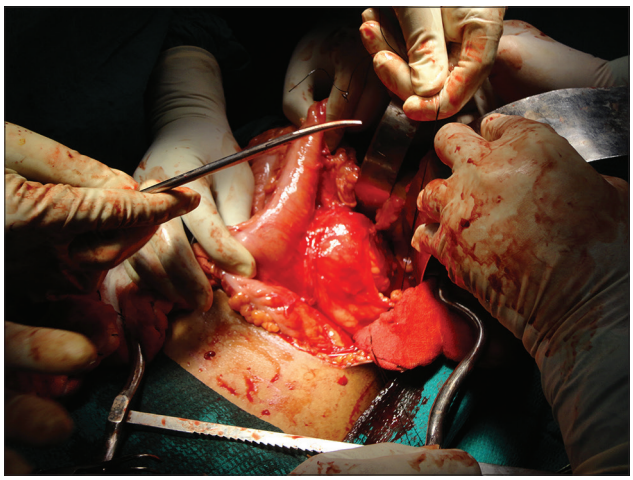

Figure 5: Dissection of the suprarenal mass, after separating adherent bowel loops

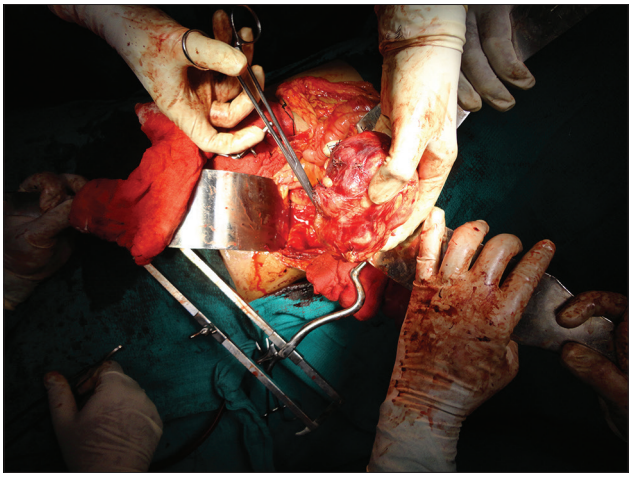

Figure 6: Left suprarenal mass removed in toto

abdominal ultrasound showed a left supra renal mass lesion, which was uniformly hypoechoic. Metabolic evaluation including serum electrolytes, plasma metanephrines and serum cortisol were within normal limits. MRI showed uniformly hypointense mass in T1-wieghted images and a very hyperintense mass lesion on T2-weighted images (light bulb sign), which made us to suspect a pheochromocytoma [Figures 9 and 10]. So we prepared the patient for surgery anticipating a pheochromocytoma. The tumor was removed by open approach [Figure 11], and the postoperative period was uneventful. But to our surprise, the lesion turned out to be juxta-adrenal schwannoma arising from nerve sheaths on histopathological examination [Figure 12]. IHC confirmed the diagnosis and was positive for S-100.

DISCUSSION

Schwannomas are rare tumors, usually benign, originating from the Schwann sheath of the peripheral or cranial nerves. They rarely present as retroperitoneal mass lesions. If they present in the suprarenal region, they may be confused with other adrenal mass lesions, particularly given that some case reports highlight uptake of metaiodobenzylguanidine by these lesions.[1]

Their cell of origin is from neural crest, comprised of differentiated Schwann cells in a background of the collagen stroma. Antoni *et al.* in 1920 classified them into two distinct histologic patterns, namely compact cellular areas named Antoni A areas and paucicellular areas named Antoni B areas. Thus, they are basically Encapsulated biphasic nerve sheath tumors derived from Schwann cells with highly ordered cellular component (Antoni A) that palisades (verocay bodies), plus myxoid component (Antoni B). Small tumors may be all Antoni A. There appear to be no racial or sex predilection for schwannomas.[2] Schwannomas have been reported to occur over a large age range.[3-9] IHC is a useful technique for detecting tumors originating from Schwann cells because they stain positive for S-100 antigen, collagen IV and laminin, and show the absence of reactivity for keratin, desmin, Vimentin.

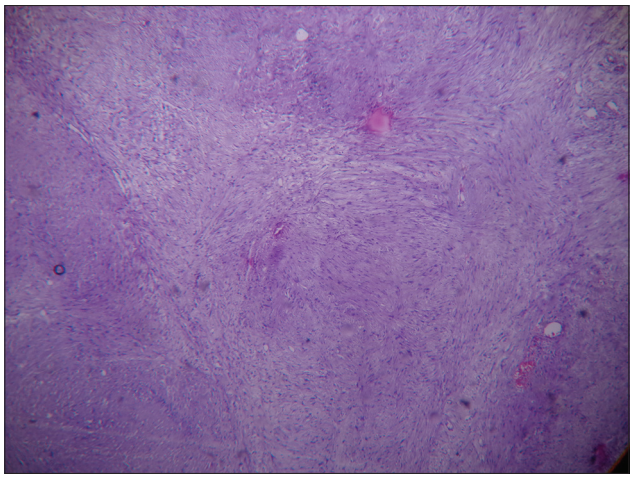

Figure 7: Whorled pattern of smooth muscle cells, features suggestive of schwannoma

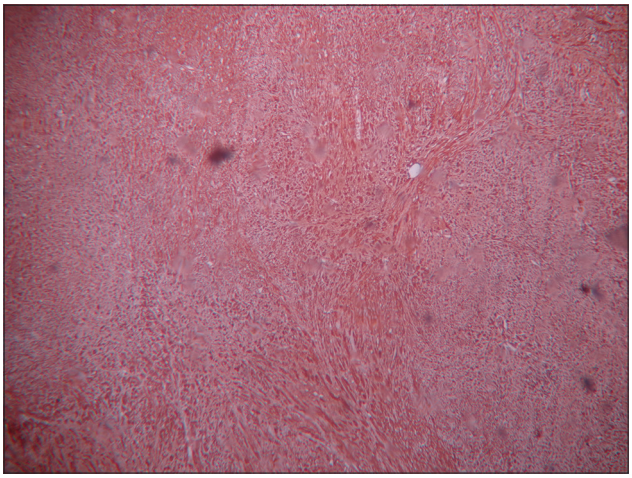

Figure 8: Immunohistochemistry staining positive for S-100 antigen

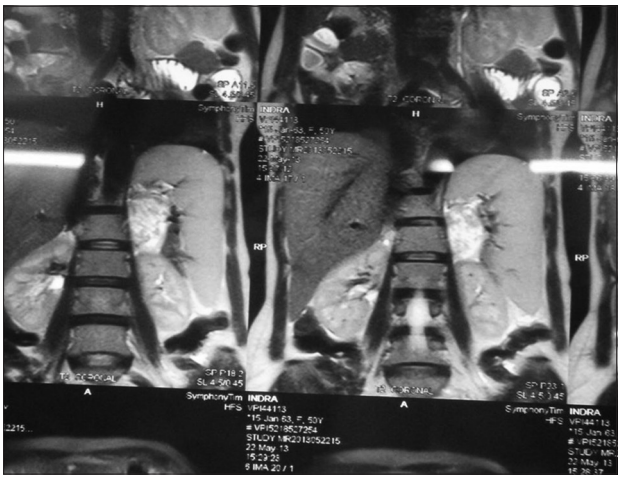

Figure 9: Coronal T2-weighted magnetic resonance imaging showing left suprarenal mass

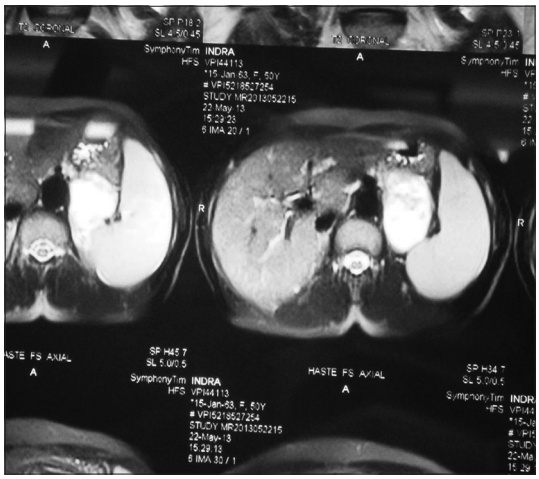

Figure 10: Hyper intense signals in T2-weighted images? Light bulb sign

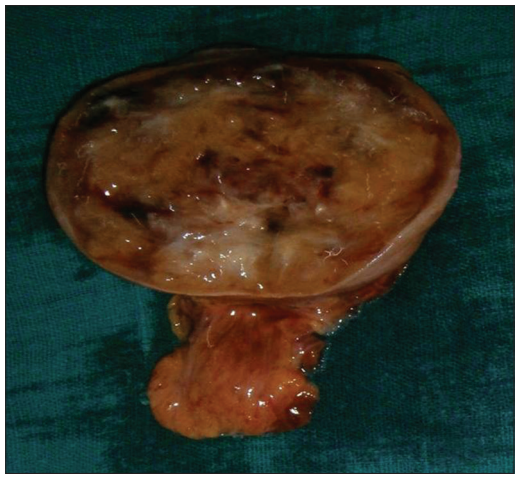

Figure 11: Left suprarenal mass, postoperative specimen

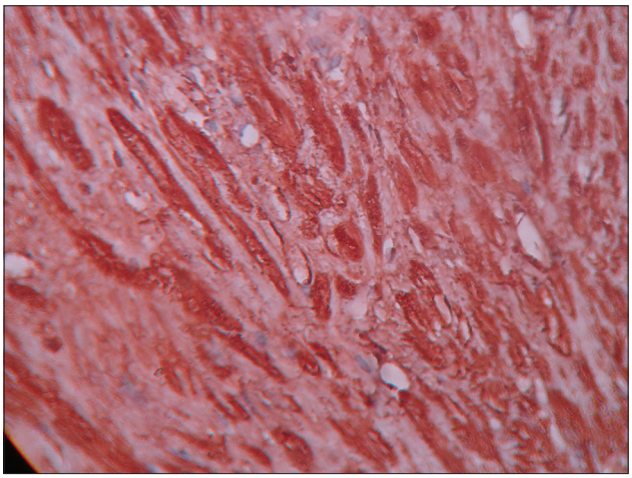

Figure 12: Immunohistochemistry showing positive staining for S-100

Antoni type A neurilemoma has elongated spindle cells arranged in irregular streams and is compact in nature, and type B tissue has a looser organization, often with cystic spaces intermixed within the tissue. The presence of cystic changes within a retroperitoneal tumor is relatively frequent in schwannomas and may suggest its diagnosis.[4] In both of our cases, we found a predominance of Antoni B areas with occasionally admixed Antoni A areas, which provided a unique diagnostic interface, which we discuss subsequently.

They are usually asympommatic and can attain massive dimensions before producing any symptoms. In both our cases, the tumors had grown to giant proportions while producing only vague dull aching pain because of their size.

In retrospective analysis, we made a note of a fascinating and unique imaging characteristic of this lesion in both of our cases. In the first case, we noted a mixed intense mass in MRI, with predominantly hyperintense areas in T2-weighted imaging. In the second case, there was a diffuse hperintense signal, mimicking the "light bulb" sign of pheochromocytoma. On correlating the radiological characteristic with Histopathological pictures, we found that this feature was due to the predominance of Antoni B areas in both our cases. According to a series of cases by Liu *et al*.[10] hypointense signal capsules in T2-weighted images in MRI and delayed enhancement of the tumor in triple phase contrast-enhanced computed tomography can be used to preoperatively diagnose juxta-adrenal schwannomas. However in our series, we observed that the simple but reliable feature of hyperintense signals in T2-weighted images could help in identification of these lesions. However, it is not mentioned in the literature about the percentage of lesions, with a predominance of Antoni B areas. Until this percentage is published, we cannot conclusively use this feature for preoperative diagnosis.

Hence at this point of time, the preoperative diagnosis of juxta-adrenal schwannomas remains elusive, with the definitive diagnosis often being established with postoperative IHC only. In selected cases, percutaneous biopsy can be used preoperatively, if the mass is nonfunctioning and there is a high suspicio of scvhwannoma.

To the best of our knowledge, only one case series of juxta-adrenal schwannoma has been published so far, although many individual case reports have been published.[10] This paper brings forward one unique imaging characteristic of these masquerading lesions, which may be used for reliable preoperative diagnosis of these rare lesions and serve as a trigger for percutaneous biopsy, which will be very safe given the hypovascular nature of these lesions. In our series, both cases presented as symptomatic mass lesions, which is also quite rare.

REFERENCES

1. Amirchetty S, Donaldson P, Etheridge C, Driver I, Parkinson C. Juxta-adrenal schwannoma presenting as 'Giant' adrenal adenoma. Endocr Abstrs 2011;25:60.
2. Yang CY, Chou CW, Lin MB, Li CF. Schwannomas of the left adrenal gland and posterior mediastinum. J Chin Med Assoc 2009;72:83-7.
3. Bedard YC, Horvath E, Kovacs K. Adrenal schwannoma with apparent uptake of immunoglobulins. Ultrastruct Pathol 1986;10:505-13.
4. Gonzalez Gonzalez A, Perea R, Palacios Llopis S, de Diego JA. Benign adrenal schwannoma. Med Clin (Barc) 2000;115:518-9.
5. Igawa T, Hakariya H, Tomonaga M. Primary adrenal schwannoma. Nihon Hinyokika Gakkai Zasshi 1998;89:567-70.
6. Ikemoto I, Yumoto T, Yoshino Y, Furuta N, Kiyota H, Oishi Y. Schwannoma with purely cystic form originating from the adrenal area: A case report. Hinyokika Kiyo 2002;48:289-91.
7. Pittasch D, Klose S, Schmitt J, Roessner A, Ridwelski K, Lippert H, *et al*. Retroperitoneal schwannoma presenting as an adrenal tumor. Exp Clin Endocrinol Diabetes 2000;108:318-21.
8. Lau SK, Spagnolo DV, Weiss LM. Schwannoma of the adrenal gland: Report of two cases. Am J Surg Pathol 2006;30:630-4.

9. Târcoveanu E, Dimofte G, Bradea C, Moldovanu R, Vasilescu A, Anton R, *et al*. Adrenal schwannoma. JSLS 2009;13:116-9.
10. Liu QY, Gao M, Li HG, Lin XF, Huang SQ, Liang BL. Juxta-adrenal schwannoma: Dynamic multi-slice CT and MRI findings. Eur J Radiol 2012;81:794-9.

How to cite this article: Damodaran S, Mahimairaj G, Velaichamy K. A case series of two cases of juxta-adrenal schwannoma presenting as adrenal mass lesion and review of the literature. Urol Ann 2015;7:254-8.

Source of Support: Nil, **Conflict of Interest:** None.