

## Oncology

# Laparoscopic excision of a large Adrenal Ganglioneuroma masquerading as Pheochromocytoma- A case report & review of literature

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## ABSTRACT

Adrenal Ganglioneuromas are rare. Evaluation for hormone secretion followed by complete excision is the treatment of choice for such masses. We present our experience of a symptomatic large Adrenal Ganglioneuroma, which was removed with laparoscopic procedure.

A 44-year gentleman presented with a symptomatic adrenal tumor. Under the impression of Adrenal Pheochromocytoma, tumor was excised laparoscopically without any untoward event. And to our surprise Adrenal Ganglioneuroma was detected in biopsy. Hence,

Ganglioneuromas should be considered as a differential for a functional or symptomatic adrenal mass. And by following proper technique even such large functional adrenal masses can be removed laparoscopically.

## Introduction

Ganglioneuromas are rare, well-differentiated tumors of neural crest origin. They are usually found in the paravertebral sympathetic plexus, while an Adrenal source is sporadic. The majority of Adrenal Ganglioneuromas (AGN) are functionally inert and are serendipitously detected in abdominal imaging for other reasons. Presently, definitive biochemical and imaging features to differentiate them from other adrenal tumors are not defined. Adrenal Ganglioneuromas are managed on the lines of an adrenal incidentaloma. Workup for hormone secretion guides in pre-operative optimization. Laparoscopic adrenalectomy is the gold standard treatment for a symptomatic AGN amenable to laparoscopy, and definitive diagnosis is by immunohistochemical examination. Here we present our experience of a symptomatic large Adrenal Ganglioneuroma clinically presenting as a pheochromocytoma.

## Case report

A 44-year old gentleman presented with a history of decreased appetite and early satiety, resulting in a progressive weight loss of 7 kg over 12 months. There was no prior history of any co-morbidity. His physical examination was unremarkable, except for hypertension. The

initial sonographic screening revealed a  $9.8 \times 5.6$  cm lesion in the right suprarenal region with internal vascularity.

Hemogram, renal function tests, and serum electrolytes were normal. A CECT abdomen confirmed the presence of a  $10.2 \times 8 \times 6.4$  cm mass (10 HU) in the right suprarenal location with a variable enhancement of 10–20 HU (Fig. 1. a,b). MRI noted a hyperintense mass on T2WI sequences with no loss of signal intensity in out of phase images indicating a lack of intracellular fat (Fig. 1.c,d).

Plasma renin activity (0.21 ng/ml/hr in supine position), aldosterone levels (10.7 ng/dl in supine position) and plasma cortisol (11.89 mcg/dl) were normal. However, Plasma aldosterone/renin ratio was found to be elevated (50.95). Plasma free metanephrines (262.4 pg./ml) and 24 hr urine metanephrines were elevated (1110.1mcg/24 hrs) indicating a medullary origin.

Tc-99 m HYNIC-TOC SPECT showed tracer uptake in the mass, implying an expression of somatostatin receptor, likely of neuroendocrine origin-Pheochromocytoma (Fig. 1.e,f).

With these findings, a diagnosis of Pheochromocytoma was reached. Blood pressure was optimized with Alpha blockade (Prazosin 2.5mg once a day) along with correction of the contracted intravascular volume. There was no requirement for beta-blockade.

Using the three standard ports, laparoscopic right adrenalectomy

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was done with specimen extraction through a small infra-umbilical incision (Fig. 2-a). In view of hemodynamic instability after pneumoperitoneum, low-pressure at 8–10 mm Hg was maintained till the adrenal vein was clipped, to reduce the risk of catecholamine storm. The post-operative period was uneventful with no requirement for post-operative antihypertensive medications. The patient was discharged on post-operative day 3. On follow up after three months, he had no complaints, and abdominal imaging was normal.

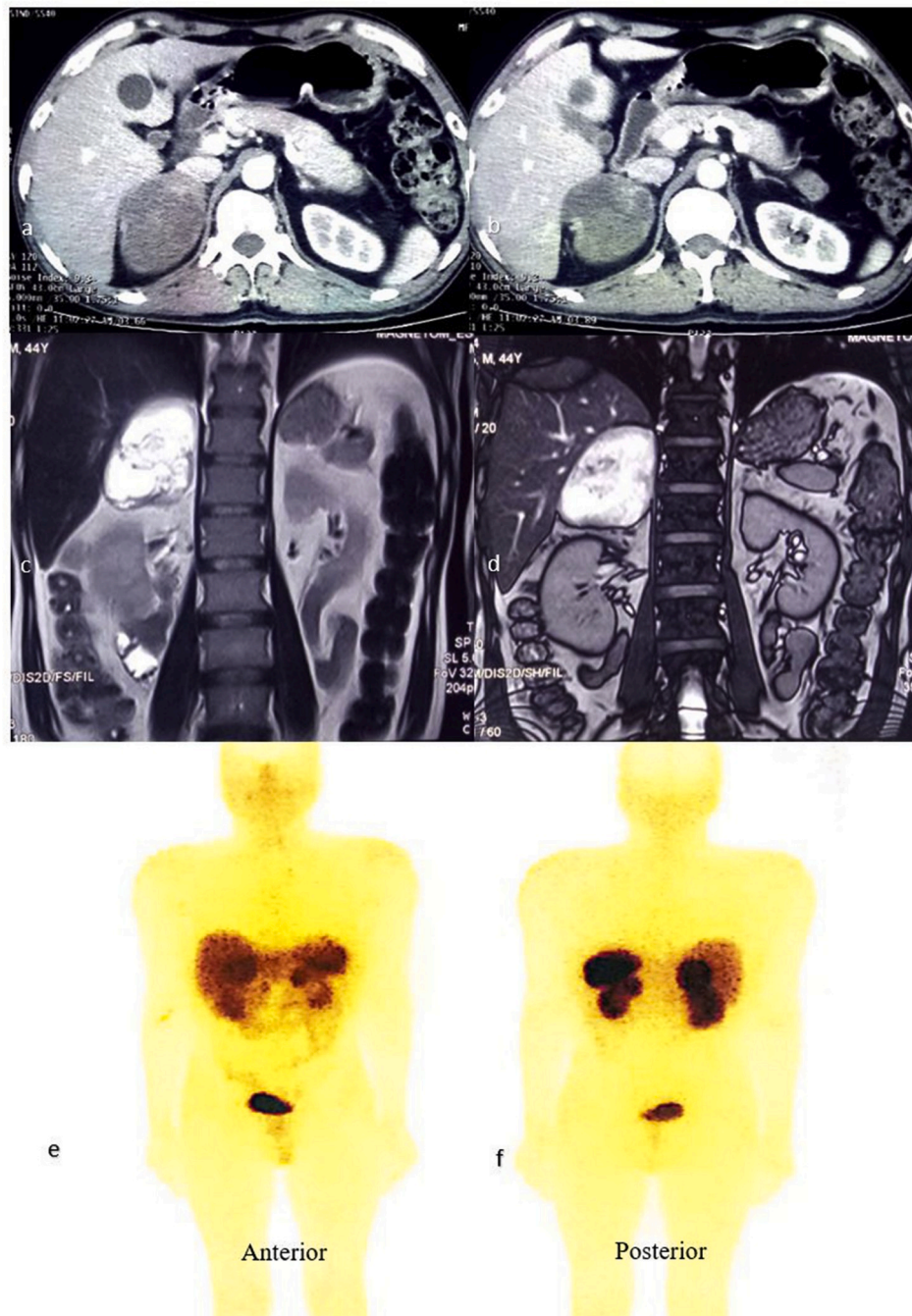
Gross specimen revealed a well-circumscribed Gray-white colored ovoid tumor of size  $10.2 \times 6$  cm with a fibrous capsule having a firm yet resilient texture (Fig. 2-b). Microscopic examination showed Schwann cells and ganglionic components (Ganglioneuroma) with peripherally

pushed adrenocortical cells (Fig. 3. a-f).

## Discussion

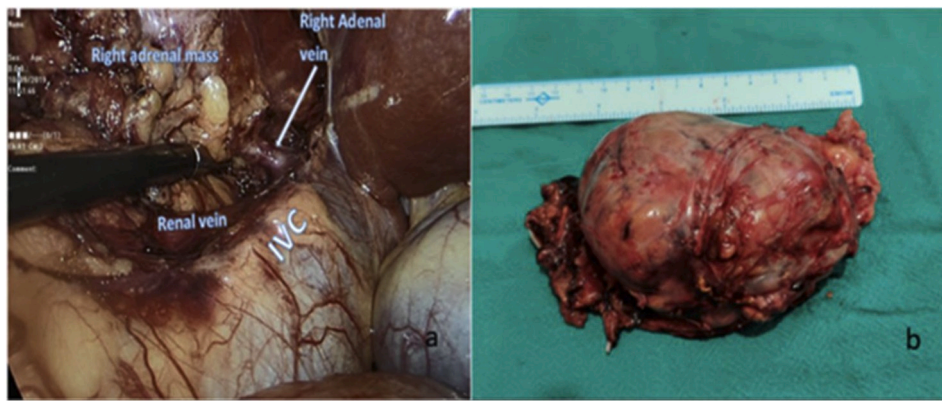
Adrenal “incidentalomas” are reported in about 4% of the abdominal cross-sectional imaging. Adrenal adenomas and metastatic deposits in the adrenal gland are the commonest; Pheochromocytoma, adrenocortical carcinoma, and myelolipoma being the other usual causes.<sup>1</sup>

Characterization of such incidentalomas is imperative to their treatment. CT attenuation number, size, and contrast washout time by a dedicated adrenal protocol help in determining the nature of the mass and provides excellent anatomical delineation.<sup>2</sup> MRI gives a better soft

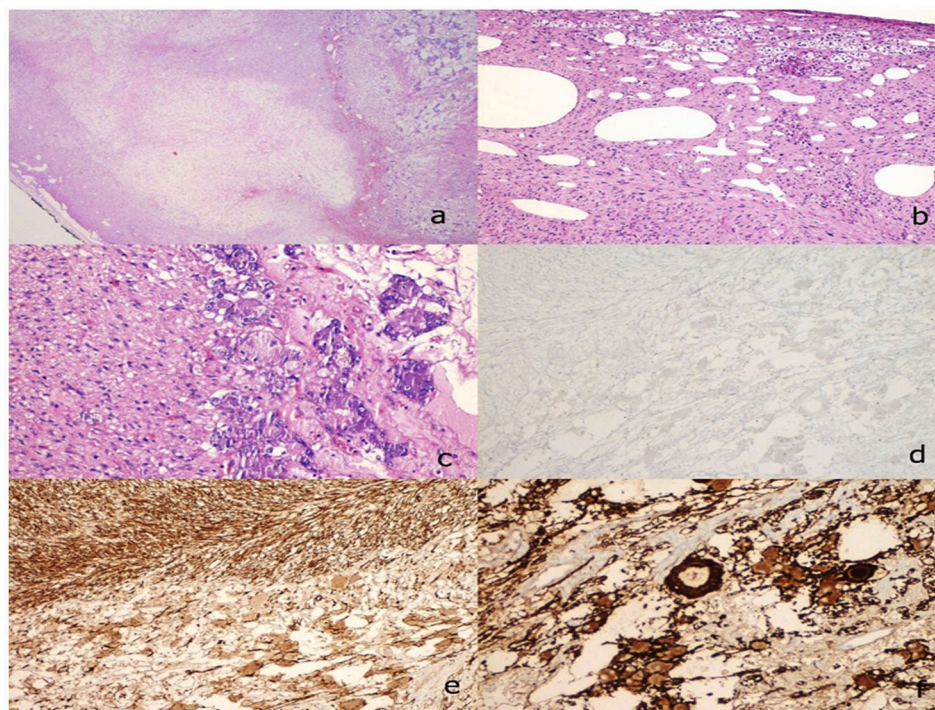


**Fig. 1.** a-b: Contrast enhanced CT scan showing  $10.2 \times 8.2 \times 6.4$  cm mass in the right suprarenal location with a variable enhancement of 10–20 HU. c: MRI abdomen showing hyperintense lesion on T2WI fat-suppressed sequence. d: No loss of signal intensity in “out of phase” images of chemical shift imaging. e-f: Tc-99 m HYNIC-TOC SPECT imaging shows tracer uptake in the right supra renal gland likely of neuroendocrine origin- Pheochromocytoma.





**Fig. 2.** 2-a: Intraoperative picture showing the right adrenal mass, right adrenal vein, right renal vein and the infra hepatic vena cava (IVC). 2-b: Gross specimen of the excised adrenal mass.



**Fig. 3.** a: The scanning view photomicrograph of the ganglioneuroma highlighting the schwannian and ganglionic component (Hematoxylin and Eosin, 20x).

b: The periphery showing entrapped adrenocortical cells (Hematoxylin and Eosin, 40x)

c: The morphology of spindle shaped Schwannian cells and polygonal ganglion cells in higher magnification, the latter with eccentric nuclei and prominent nucleoli and focal neuromelanin cytoplasmic granules giving brownish hue to the ganglion cells (Hematoxylin and Eosin, 200x)

d-f: Immunohistochemistry; Negativity for Melan-A (d, 200x), neural tissue positivity for S100 (e, 200x), and ganglionic positivity for Synaptophysin (f, 400x).

tissue delineation, and should always include both T1W and T2W sequences with “Chemical shift imaging.”<sup>2</sup> Laboratory parameters like serum Cortisol, serum Metanephrines or 24-h urine Metanephrines, overnight Dexamethasone suppression test and serum potassium estimation should be done to know the functional status.<sup>3</sup> Nuclear imaging is essential to determine the functional status of the tumor, to detect any concurrent extra-adrenal functional tumor, or to rule out undetected metastasis. FNAC may be required when an adrenal secondary is suspected after ruling out Pheochromocytoma.<sup>3</sup> Adrenal tumors that are more than 6 cm have a more than 90% chance of harboring a malignancy while those less than 4 cm have a shallow risk of malignancy.<sup>4</sup> Hence all tumors more than 6 cm are resected, whereas those less than 4 cm can be observed safely. While all symptomatic or functional tumors need surgical resection after optimization, the management of asymptomatic tumors of 4–6 cm size is controversial.

Adrenal ganglioneuromas are rare tumors arising from the Neural crest cells. They fall towards the differentiated end of the spectrum, while Neuroblastomas and Ganglioneuroblastomas are less differentiated. They are composed of mature Schwann cells and ganglion cells,

admixed in a fibrous stroma. The majority of these are non-functional, allowing them to attain large sizes without detection. Though the majority are incidentally detected (17%), vague abdominal pain (9%) is the most frequent symptom.<sup>5</sup> Patients may also complain of headaches, dizziness, weakness, virilization, and hypertension. Malignant transformation, though rare, has been reported, usually into Neuroblastoma and Malignant peripheral nerve sheath tumor. TNM staging is the primary determinant of prognosis and laparoscopic adrenalectomy is the gold standard treatment whenever feasible.

There is no difference in the malignancy recurrence rates by open or laparoscopic adrenalectomy techniques when standard oncological principles are followed. Pheochromocytomas are notoriously known to cause an intraoperative sympathetic storm due to manipulation of the gland. The laparoscopic approach reduces this risk; however, the pneumoperitoneum exerts pressure on the adrenal glands and may in turn precipitate a sympathetic storm. Meticulous dissection under low-pressure pneumoperitoneum is imperative to prevent the uncontrolled release of catecholamines into the bloodstream.<sup>5</sup> The classical teaching of early adrenal vein ligation has been refuted in recent studies and is

not necessary.<sup>5</sup> Appropriate selection of patients and the expertise of the surgeon are the most crucial factors dictating the outcome.

## Conclusion

Ganglioneuroma may mimic Pheochromocytoma and should be considered as a differential while evaluating a functional or symptomatic adrenal mass. Even large functional adrenal masses can be excised laparoscopically by following proper techniques. And to establish a diagnosis, immunohistochemical staining is a must to differentiate neuroblastic tumors from Pheochromocytomas.

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