

# Pure cystic adrenal space-occupying lesion: always rule out cystic pheochromocytoma

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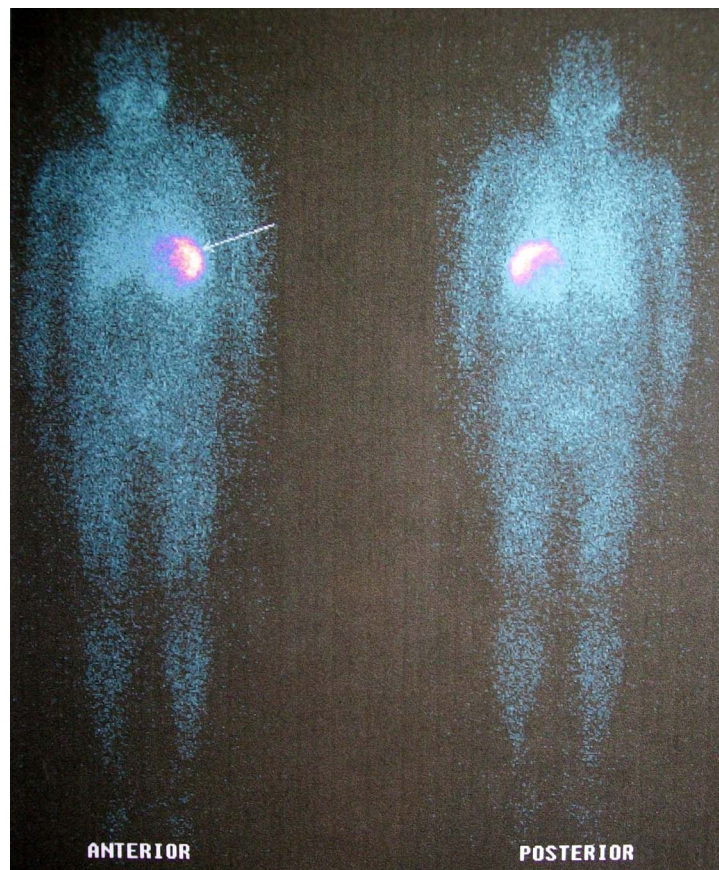
## DESCRIPTION

A 57-year-old woman was referred for preoperative fitness before planned laparotomy for left adrenal cystic space-occupying lesion (SOL). She had been



**Figure 1** Large purely cystic left adrenal space-occupying lesion in precontrast CT scan.

suffering from ill-defined epigastric pain for the preceding 6 months and was found to have a well-defined rounded left suprarenal cystic SOL (11.6 cm×9.4 cm×9 cm) seen on abdominal ultrasonography. A contrast-enhanced CT scan revealed a purely cystic SOL (12.1 cm(CC)×11.3 cm (TR)×10 cm(AP)) with hyperdense enhancing thin wall in the left suprarenal area and non-visualisation of the left adrenal gland (figure 1). She was found to have elevated blood pressure (150/100 mm of Hg) and preoperative evaluation revealed impaired fasting glucose (fasting plasma glucose 110 mg/dL) and normal plasma catecholamines (epinephrine 59.1 pg/mL (reference <125 pg/mL); norepinephrine 303.7 pg/mL (reference <600 pg/mL)). She did not report headache, palpitation, sweating, paroxysmal spells or any of the typical signs and symptoms of glucocorticoid excess. We could not find any previous record of her blood pressure measurements. A relook into the CT scan documented that the precontrast



**Figure 2** I-131-metaiodobenzylguanidine (MIBG) scan showing solitary MIBG avid lesion in left adrenal.

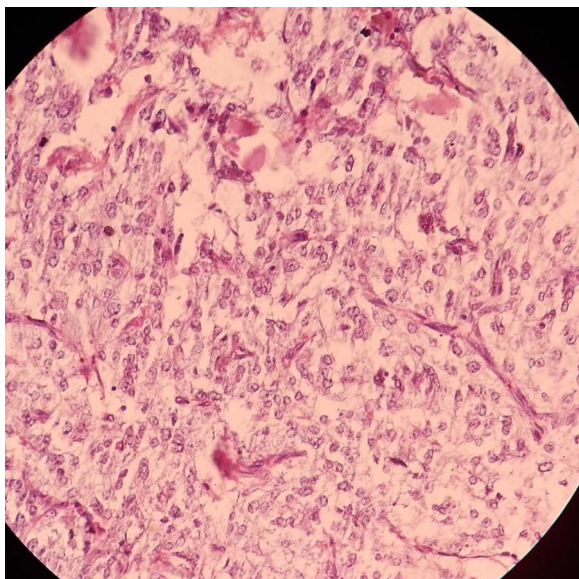


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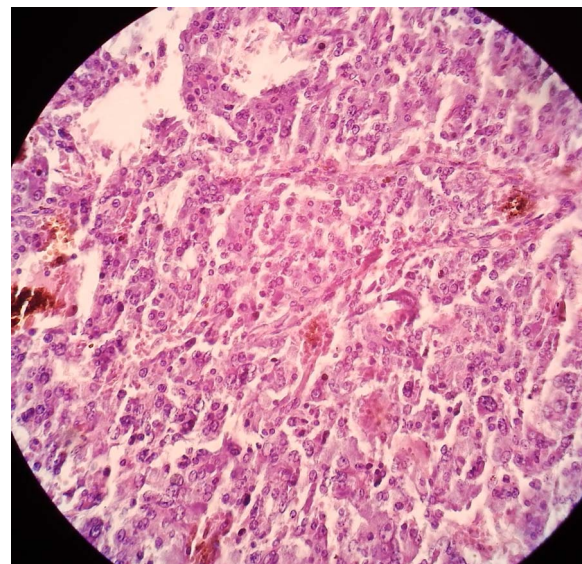
Hounsfield unit (HU) of the cystic component was 22 (pre-contrast HU of <10 is suggestive of lipid-rich benign adenoma). A thorough and systemic work-up for adrenal incidentaloma revealed the following: serum cortisol after 1 mg overnight dexamethasone suppression test: 2.1 µg/dL; dehydroepiandrosterone sulfate: 78.9 µg/dL (reference 15–200 µg/dL). Despite plasma catecholamines being within the reference range, we proceeded for estimation of 24 hours urinary fractionated metanephrines and the values were grossly elevated (metanephrines 5297.6 µg/day (reference <350 µg/day); normetanephrines 2598.4 µg/day (reference <600 µg/day)). I-131-metaiodobenzylguanidine (MIBG) scan documented a single MIBG concentrating mass in the left suprarenal area without any other MIBG avid lesion anywhere in the body (figure 2). A purely cystic solitary left adrenal pheochromocytoma was diagnosed and the patient was put on prazosin and liberal oral salt for 2 weeks followed by propranolol 4 days prior to surgery. The left-sided adrenal mass was removed by laparotomy (figure 3) with normalisation of urinary metanephrines carried out 2 weeks following surgery. The histology of the resected



**Figure 3** Surgical specimen removed by laparotomy.



**Figure 4** Typical nested to solid pattern of monomorphic polygonal cells having adequate amphophilic cytoplasm (magnification×400).



**Figure 5** Areas of haemorrhage and pleomorphic cells with giant cell formation. Nested pattern is still maintained. Here cytoplasm is finely granular and eosinophilic. Pleomorphism and giant cell formation are not indicative of malignancy in pheochromocytoma (magnification×400).

mass revealed a typical nested pattern of monomorphic polygonal cells having adequate amphophilic cytoplasm with areas of haemorrhage (figures 4 and 5).

Pure cystic lesions of the adrenal glands having heterogeneity in aetiology and clinical manifestations are uncommon and majority of them represent benign endothelial cysts, epithelial cysts or pseudocysts. Less than 10% of adrenal incidentalomas have proved to be pheochromocytomas and CT findings consistent with (though not diagnostic of) pheochromocytomas include solid/heterogeneous appearance, increased attenuation on unenhanced CT scan, prominent vascularity of the mass and delayed washout of the contrast.<sup>1</sup> The exact incidence of pure cystic pheochromocytoma is largely unknown, but about one-fifth of these adrenal medullary tumours may have pure cystic appearance.<sup>2</sup> Patients with cystic pheochromocytoma are less likely to present with paroxysmal spells/typical symptoms, more commonly present with unusual manifestations like upper abdominal or lumbar pain and have negative biochemical screening tests. Urinary or plasma metanephrines are preferred over urinary/plasma catecholamines as the initial screening test in patients with suspected pheochromocytoma. The sensitivity of metanephrines are much higher than catecholamines as these catecholamine metabolites are produced continuously within these tumours independent of exocytotic catecholamine release, which in many cases is episodic or occurs at very low rates. The pathogenesis of cystic pheochromocytoma has largely been speculated and the inciting event could be the tumour outgrowing its vascular supply. Subsequent intralesional haemorrhage, necrosis and liquefaction within the mass eventually give the tumour a cystic appearance. The lesser tumour burden also makes them less likely to be symptomatic and to have elevated plasma or urinary catecholamines or its metabolites. The cystic component reflects necrosis and has a low attenuation on CT and hyperintense signal on T2-weighted MRI images. However, the characteristic radiological appearance is a relatively thick wall having persistent contrast enhancement which was also present in this patient.

## Learning points

- ▶ Purely cystic adrenal lesions with non-specific clinical manifestations may at times be associated with catecholamine excess, which greatly mimic benign lesions and carry detrimental clinical consequences if misdiagnosed.
- ▶ All adrenal lesions, even asymptomatic, require a thorough and systemic endocrinological evaluation irrespective of their imaging appearance.
- ▶ Considering the poor sensitivity of plasma/urinary catecholamines, screening biochemical testing for suspected pheochromocytoma should include measurements of plasma-free metanephrines or urinary fractionated metanephrines.

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**Patient consent** Obtained.

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