Adrenal cysts—report, review and classification

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Summary: A case of a giant adrenal pseudocyst is reported. The clinico-pathological features are reviewed and a classification is proposed.

Introduction

Cystic lesions of the adrenal gland are uncommon but should be considered in the differential diagnosis of an abdominal mass. The first report of an adrenal cyst was attributed by Doran to Greiselia, a Viennese physician, in 1670.1

Case report

A 53 year old woman presented with a one year history of abdominal discomfort. Systematic enquiry was unremarkable apart from recent onset of vitiligo affecting hands and feet. There was no evidence of endocrinological disorder. She was obese (96 kg) and there was a large, fixed, smooth mass filling the left upper quadrant.

Routine laboratory investigations were normal, as were serum cortisol levels. Ultrasound showed a cystic mass displacing adjacent viscera but did not identify the organ of origin. Intravenous urography showed the left kidney displaced to lie over the bodies of L4 and L5 but renal outlines and collecting systems were normal. Computerized tomography (Figure 1) confirmed a cystic lesion, 22 × 15 cm, displacing the left kidney. The left adrenal was not visualized.

At laparotomy, a thick walled cyst was found, which arose from the posterior abdominal wall in the region of the tail of the pancreas. The splenic vein was incorporated in the cyst wall but it became apparent as dissection proceeded that the pancreas, though closely applied to the cyst, was itself intact. During dissection the cyst ruptured, releasing several litres of thick fluid, which was dark brown due to altered blood. The fluid was sterile on culture and had a normal amylase and electrolyte content. The cyst and spleen were removed. The patient made an uneventful recovery.

The cyst comprised a main locule plus a smaller one which contained clotted blood. It had a 1 cm thick fibrous wall with fat and large blood vessels externally. The inner surface was mostly smooth but with several groups of thrombosed, aneurysmal vessels and scattered areas of calcification.

Histologically (Figure 2) the cyst wall comprised fibro-elastic tissue with focal calcification. There was no epithelial or endothelial inner lining but the wall contained flattened islands of adrenal cortical tissue. There were numerous abnormal blood vessels: enlarged thick-walled arteries externally, distended intra-mural veins and thrombosed aneurysmal vessels with no elastica, which protruded into the cyst. There was no medullary tissue, tumour or hydatid disease. The diagnosis was giant adrenal pseudocyst.

Figure 1 Computed tomography at the level of the pancreas. The cyst (C) is smooth-edged, thin walled and low density with no contrast enhancement. The tail of the pancreas is pushed forward (arrowheads). Left adrenal not seen on any section.

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An alternative approach, if measured. Catecholamines are urinary metabolites found in Cushing's syndrome and are often elevated in patients with parathyroid adenoma. Some 250 adrenal cysts have been previously reported. Our case is one of the largest recorded (a cyst removed from a West African woman measured 33 cm). Most cysts are asymptomatic and are found incidentally. An estimate of true incidence, based on post-mortem studies, is 0.06%. They occur at all ages, with a peak incidence of symptomatic (operative) cases in the 4th-5th decades and a female: male ratio of about 3:1.

Adrenal cysts may cause abdominal discomfort due to their size and displacement of adjacent viscera. The diagnosis is suggested by peripheral curvilinear calcification on a plain radiograph. Ultrasonography and computed tomography offer the most reliable methods of pre-operative diagnosis. Hormone production causing Cushing's syndrome and an isolated association with a parathyroid adenoma have been reported. A small minority of cysts contain functioning medullary tissue with catecholamine levels high enough to suggest phaeochromocytoma; if this is suspected, urinary catecholamines and metabolites should be measured.

Surgical exploration is recommended, either trans-abdominally if the cyst is large, or via a loin approach, with or without rib resection if small; the latter approach avoids peritoneal contamination. An alternative treatment is ultrasound guided percutaneous drainage so long as cystic degeneration can be excluded using ultrasonography or computed tomography. This option was rejected in our case due to the large size of the cyst and doubt concerning its origin.

Pathological classification

A classification is proposed appropriate to pathologists and clinicians (Table I) based on that of Hodges & Ellis. True cysts have a definite inner endothelial or epithelial cellular lining whereas pseudocysts have not. Most endothelial cysts are of lymphatic type. These are regarded as a developmental anomaly and are usually solitary and small. True vascular endothelial cysts (haemangiomata) are rare. True cysts of epithelial type are uncommon and probably represent cystic adenomas; ciliated epithelial-lined developmental cysts were claimed in an early publication, but are disputed.

Pseudocysts, by definition, have no cellular lining. The common haemorrhagic type has a thick fibrous wall, often calcified or even ossified. The contents are frequently blood-stained. Adrenal cortical tissue is often found in the wall as flattened islands or a discrete mass. It has been thought that they arise from encapsulation and organisation of a haematoma within a normal or abnormal adrenal related to trauma, sepsis, hypoxia or neoplasia.

Two rarities must be considered: cystic degeneration in a malignant primary or a metastatic tumour and parasitic (echinococcal) cysts. These are distinct entities but we agree with Hodges & Ellis that they are most conveniently classified as pseudocysts.

It is useful to remember that the differential diagnosis generally lies between lymphatic endothelial true cysts and haemorrhagic pseudocysts, each group accounting for 40% of cases.

<table>
<thead>
<tr>
<th>Pathological classification of adrenal cysts</th>
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<tbody>
<tr>
<td>True cysts</td>
<td>(1) Endothelial – lymphatic</td>
</tr>
<tr>
<td></td>
<td>(2) Epithelial</td>
</tr>
<tr>
<td>Pseudocysts</td>
<td>(1) Haemorrhagic</td>
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<td>(2) Neoplastic</td>
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<td>(3) Parasitic</td>
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Figure 2 Microscopy of the cyst with adrenal cortical tissue (AC) in the wall and thrombosed vessels (TV) on the inner surface. Magnification × 22.
References

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