Vascular Adrenal Cysts

A Brief Review of the Literature

Eleni Carvounis, MD, FRCPC; Athanasios Marinis, MD; Nikolaos Arkadopoulous, MD; Theodosios Theodosopoulos, MD; Vassilios Smyrniotis, MD

Adrenal cysts are rare and form a heterogeneous group of lesions. Their incidence in autopsy series has been reported as 0.06%. However, the frequency with which they are discovered appears to be increasing because of improved radiologic imaging techniques.

Historically, the first case of a ruptured adrenal cyst was described by Griselius in Vienna in 1670. Rayers reported the second case in France in 1837. Classification of adrenal cysts was first attempted in 1906 by Terrier and Lecene. In 1959, Abeshouse reclassified adrenal cysts based on 155 cases, and in 1966 Foster modified and classified histologically 220 adrenal cysts into 4 types: (a) parasitic cysts (7%), (b) epithelial cysts (9%), (c) pseudocysts (39%), and (d) endothelial cysts (45%). The relationship between the latter 2 types of adrenal cysts (pseudocysts and endothelial cysts) has been investigated by immuno- and electron microscopy, and the evidence is in favor of the vascular origin of the pseudocyst. Indeed, in the recent literature, adrenal pseudocysts and endothelial cysts are considered as variants of vascular adrenal cysts.

Cysts of parasitic (mainly echinococcal) and epithelial origin are very rare. Vascular adrenal cysts account for the majority of the cases of adrenal nonneoplastic cysts.

Patients with vascular adrenal cysts are in their fifth to sixth decade, and women outnumber men with a ratio of 3:1. The clinical presentation varies and is independent of the cyst size. The most common clinical presentation is abdominal pain (35%), followed by incidental findings (32%). A few patients had hypertension that resolved following removal of the tumor. There are reports in the literature of patients presenting with acute abdomen and shock due to a ruptured adrenal vascular cyst. Rarely, the clinical presentation is with pyloric stenosis. Abdominal imaging studies show a solid or cystic suprarenal mass (Figure 1). Calcifications are detected in 15% to 30% of adrenal vascular cysts and are peripheral (mural) or speckled throughout the lesion. Rarely, the calcifications may be massive and central in location. These imaging features of adrenal vascular cysts are not specific. Although the cystic nature and liquid content of these cysts is demonstrated on ultrasonographic scans, computed tomographic scans, and magnetic resonance images, their histologic type cannot be predicted. Fine-needle aspiration biopsy on a reported case of adrenal vascular cyst yielded hemorrhagic material that was nondiagnostic. Subsequent histologic evaluation of the excised specimen following fine-needle aspiration biopsy may show papillary endothelial hyperplasia in the lesion. Grossly, both types of adrenal vascular cysts (endothelial and pseudocysts) are well circumscribed and surrounded by a capsule (Figure 2). Their size ranges from 1.4 to 33 cm in diameter. The majority of cases are unilateral. Bilateral involvement has been reported in 8% of patients. Pseudocysts are usually thick-walled and unilocular and contain blood and yellow-brown amorphous material. Endothelial vascular cysts are usually thin-walled and multilocular and contain yellow-tinted serous fluid. Microscopically, pseudocysts have a wall of dense, hyalinized connective tissue that may have focal calcifications or metaplastic bone formation. Entrapped cortical cells are identified in the cyst wall. In some cases, smooth muscle is detected within the wall of the cyst, and it seems to be continuous with the smooth muscle of the adrenal vein. Similarly, in a few cases there are irregular endothelial-lined spaces within the residual cortex and the cyst wall itself. No endothelial lining is detected in the cyst wall.
The contents of the cyst are clotted blood and fibrinous material that may contain islands of viable or necrotic cortical cells. Unusual variants of adrenal pseudocysts have been reported in the literature, with intracystic mature adipose tissue, myelolipomatous metaplasia, or intracystic metastatic breast carcinoma. Endothelial cysts are lined by flattened cells resembling normal endothelium (Figure 3). Immunohistochemical studies using antibodies to CD31 and factor VIII antigen confirm the endothelial nature of the lining cells. Staining results similar to those of factor VIII antigen have been obtained in residual foci of flattened cells lining the inner wall of some pseudocysts, lending support to the notion that pseudocysts are of vascular origin. The lining of some of the endothelial cysts reported in the literature reacted only weakly for factor VIII-related antigen, but it stained strongly for collagen type IV. This staining pattern, along with the lack of hemorrhage in endothelial cysts, suggests lymphatic differentiation. Immunohistochemical staining for epithelial markers (EMA, AE1/AE3, CAM 5.2) has been negative in both types of adrenal vascular cysts. Various theories have been suggested concerning the pathogenesis of adrenal vascular cysts, including origin from a preexisting vascular hamartoma, from ectasia of lymphatic channels, or secondary to intraparenchymal hemorrhage. Anomalous vascular channels have been identified within and surrounding most vascular cysts of the pseudocyst type and a case of focal nodular hyperplasia of the liver (a lesion thought to be a result of vascular malformation) coexisting with an adrenal vascular pseudocyst has been reported in the literature, suggesting that vascular abnormalities are important in the formation of these lesions. Other entities to be considered in the differential diagnosis of adrenal vascular cysts include cystically degenerated adrenal neoplasms, primary (pheochromocytoma, adrenal cortical adenoma, and adrenal cortical carcinoma) or metastatic. Thorough gross examination of the specimen with multiple sections from the cyst wall will disclose the presence of residual neoplastic cells, and close examination of the cyst contents will reveal...
the presence of necrotic neoplastic cells in the case of a cystically degenerated neoplasm. Surgical excision when possible by laparoscopic approach is the treatment of choice for adrenal vascular cysts.\(^1,14\) The prognosis is excellent.\(^1\)

References


