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CASE REPORT

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Retroperitoneal bronchogenic cyst masquerading clinically and radiologically as a phaeochromocytoma

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Abstract Bronchogenic cysts are relatively rare congenital anomalies that represent malformations of the embryonic foregut and are morphologically expressed as maldevelopments of the respiratory system. Anatomically, they can be positioned at any location along the central axis of the respiratory system, but are more commonly discovered in the thorax. Infradiaphragmatic bronchogenic cysts are rare and retroperitoneal ones distinctly unusual. We report a retroperitoneal bronchogenic cyst clinically masquerading as a phaeochromocytoma.

Key words Cyst · Retroperitoneum · Bronchogenic Cyst · Phaeochromocytoma

Introduction

Bronchogenic or bronchial cysts are usually discovered as incidental findings on chest radiographs, during exploratory chest surgery or postmortem examination [4, 5, 12–14]. When they come to clinical attention, it is usually because of symptoms related to their physical presence including obstruction of the tracheobronchial tree, especially in neonates, or due to secondary manifestations such as infection, perforation or haemorrhage [6, 14].

The cysts are of endodermal foregut origin and are are often associated with maldevelopment in the upper digestive tract [1]. They represent abnormal or supernumerary buds from the embryonic foregut and can be found along the central axis of the respiratory system or in the areas occupied by the lungs [4, 5, 12–14]. They occur most commonly in the subcarinal, parahilar or hilar areas in the middle mediastinum, but can also arise at or above the suprasternal notch and subdiaphragmatically [14]. Retroperitoneal locations are rare and we found only five previously published cases in the retroperitoneum [2, 7, 9, 17]. Our patient is unique from a clinical standpoint in that radiological imaging studies, laboratory findings and biochemical features suggested the presence of a phaeochromocytoma.

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Clinical history

A 44-year-old guidance counsellor came to the emergency room of his local hospital with a 4 day history of left upper quadrant pain. He was treated with ranitidium and Sucralfate with no success. Initial physical examination and chest radiograph were normal. An intravenous pyelogram showed mild extrinsic distortion of the superior pole of the left kidney. CT scan of the abdomen revealed a heterogeneous 8 cm×6 cm left adrenal mass with apparent intratumoral haemorrhage and a possible small haemorrhage adjacent to the left crus of the diaphragm.

The patient was taking gemfibrozil for hypertriglyceridaemia, ibuprofen for chronic low back pain and melatonin supplements. He had no history of hypertension and no family history suggestive of familial endocrinopathy. Sixteen years earlier, he had been the victim of vehicular trauma necessitating a splenectomy; a 4 cm accessory spleen was visible inferior to the greater gastric curvature on CT scan.

On physical examination, blood pressure was 136/80 mmHg, pulse 76 per min and regular, respirations 20 per min weight 75.1 kg and temperature 99–100.2° F. He appeared well-nourished and anxious. There was minor left costovertebral percussion tenderness as well as mild tenderness to deep palpation in the left hypogastrium.

A morning plasma cortisol was low at 1.8 µg/dl (normal 6–30 µg/dl). Twenty-four hour urine collection revealed mildly elevated levels of norepinephrine at 118.2 µg (normal 12–85.5 µg) and vanillylmandelin acid (VMA) at 7.58 mg (normal 1.4–5.5 mg), with normal levels of dopamine at 401.4 µg (normal 65–400 µg) and epinephrine at 2.23 µg (normal 1.7–2.4 µg). Serum chromogranin A was normal at 29.5 ng/ml (normal 10–50 ng/ml) and 24 h urine free cortisol was slightly high at 99 mg/total volume (TV) (normal 20–90 mg/TV). Full blood count, electrolytes and blood chemistry were all within normal limits.

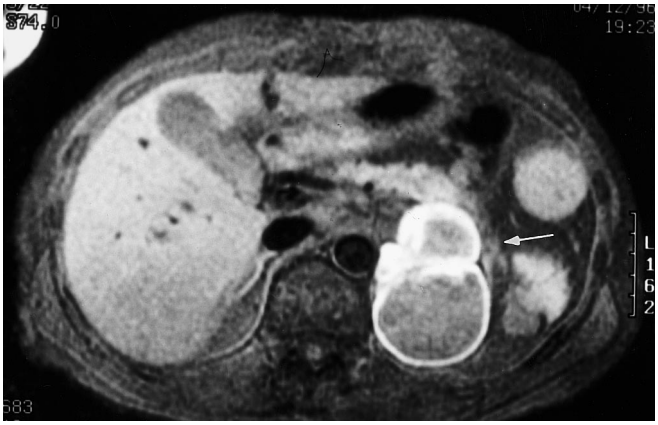

Fig. 1 MRI scan, T1 weighted image, demonstrates a large, lobulated left apparent adrenal mass with a hyperintense rim consistent with methaemoglobin blood products. The lesser signal intensity of the central component is suggestive of proteinaceous material or haemosiderin. The laterally displaced tissue (*arrow*) probably represents a limb of the left adrenal gland

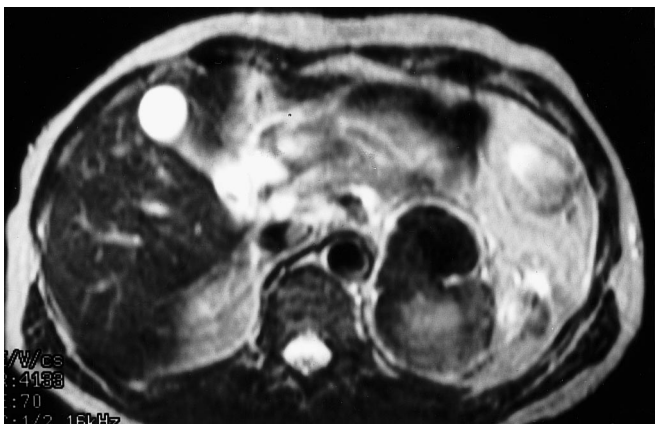

Fig. 2 MRI scan, T2 weighted image, demonstrates a hyperintense rim of a large, lobulated mass which appears to be within the left adrenal gland. The hypointense central components of the mass are consistent with haemosiderin-containing blood products

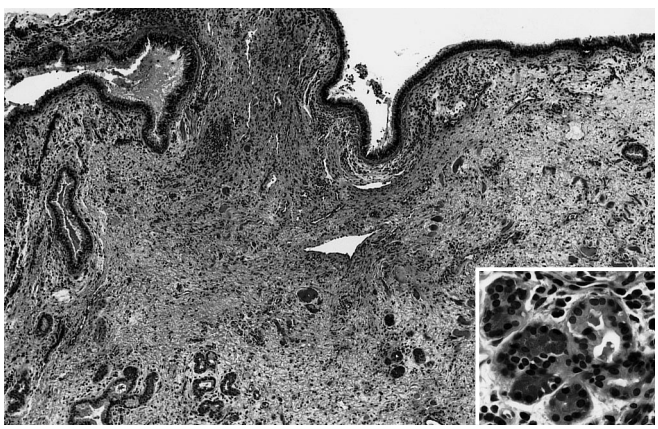

Fig. 3 Histological section of the bronchogenic cyst. The lumen is lined by pseudostratified, ciliated columnar epithelium and the lamina propria contains chronic inflammatory cells and seromucinous glands, (×60). *Inset*, (×300)

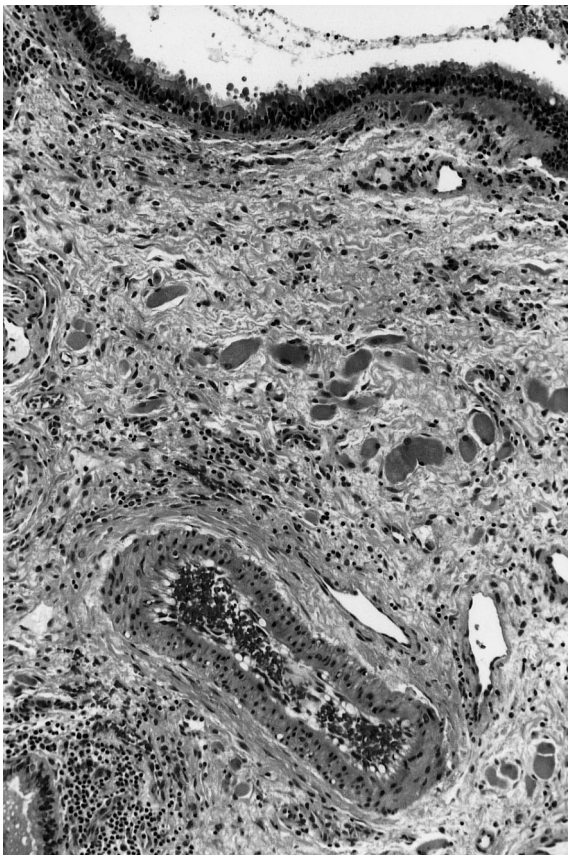

Fig. 4 Scattered smooth muscle fibres are present in the lamina propria of the cyst wall, (×150)

The patient also reported the recent onset of anxiety spells that awakened him from sleep and were accompanied by nausea, diaphoresis and a feeling of impending doom. He reported more or less constant anxiety compared to his mood in the previous 2 years. An MRI demonstrated an internally haemorrhagic left adrenal mass which was suggestive of adrenocortical carcinoma or pheochromocytoma (Figs. 1, 2). In preparation for left adrenalectomy alpha blockade was instituted using phenoxybenzamine hydrochloride.

At surgical exploration the mass was vascular and friable and palpation produced immediately tachycardia and hypertension although the systolic blood pressure did not exceed 150 mmHg. Normal adrenal tissue was visible at the inferior and medial aspects of the mass, which was intimately adherent to the left hemidiaphragm. The tumour was resected en bloc, excising some posterior diaphragmatic fibres with the specimen. Cardiovascular lability improved after the adrenal vein was ligated. The patient experienced an uneventful recovery and was discharged home on the fourth postoperative day.

Pathological findings

The mass was oval-shaped, weighed and measured 170 g 10 cm×10 cm×6 cm with an adherent, normal appearing 7.0 cm×3.3 cm×2.0 cm adrenal gland. The tumour was focally covered by peritoneum, diaphragmatic skeletal muscle fibres and adipose tissue. The mass was cystic and filled with dark red, clotted and unclotted blood. The

wall of the cyst ranged 1–4 mm in thickness and was predominantly fibrous. The portion of the wall adjacent to the adrenal cortex was multiloculated with cysts ranging from 0.1 cm to 0.6 cm in diameter. No areas of calcification, dentigerous structures, bone, cartilage, hair or sebaceous material were noted.

Microscopically, the lumenal wall was lined by pseudostratified, ciliated, columnar epithelium with a well-defined basal cell layer on a distinct basement membrane. There was a lamina propria composed of loose, collagenous connective tissue that contained occasional clusters of mixed seromucinous glands, variable numbers of chronic inflammatory cells, and lymphoid aggregates (Fig. 3). Small arteries, veins, capillaries and nerves along with occasional smooth muscle fibres were also scattered throughout the lamina propria (Fig. 4). Indeed, with all components of a normal bronchus present, the cyst focally recapitulated a normal bronchus. Areas of recent, organizing haemorrhage were noted on the mucosal surface and within the wall of the cyst. The adrenal gland had a histologically normal capsule, medulla and cortex. The surrounding connective tissue focally exhibited evidence of previous haemorrhage with numerous siderophages, active fibroblastic proliferation and recent and remote neovascularization. No heterologous structures were noted. All structures present were consistent with derivation from entoderm. There was no evidence of malignancy.

Discussion

This bronchogenic cyst, located in the retroperitoneum, appeared to be an adrenal mass by CT scan and MRI. The mild elevation in 24 h urinary catecholamines and precursor metabolites and clinical symptomatology stimulated a phaeochromocytoma. We found five previously reported cases of bronchogenic cysts in the retroperitoneum occuring in adults in four cases with locations in the supra-adrenal area, superior to the tail of the pancreas, loosely attached to the superior aspect of the body of the pancreas, and one adjacent to the left adrenal [4, 8, 22, 24]. The one paediatric case was attached to the anterior surface of the pancreas in an infant with multiple congenital anomalies who died at 10 weeks of age. Roentgenographic studies had suggested the possibility of an adrenal neuroblastoma [17]. Ultrasound and CT scan suggested an adrenal mass in one of the adult cases, an asymptomatic 38-year-old male [24]. In none of these cases was there a clinical suspicion of phaeochromocytoma.

Phaeochromocytomas are renowned for their ability to mimic a number of other diseases, thereby obscuring clinical recognition in some instances. Symptomatic hypertensive attacks can be episodic or ephemeral in duration with intervening normotensive periods while only 50% of phaeochromocytoma patients have sustained hypertension [16]. However, this patient did manifest several clinical signs that are often associated with phaeochromocytoma: periods of “altered sensorium,” and anxiety attacks accompanied by feelings of impending doom, diaphoresis and nausea. He lacked a history of constipation, a very common clinical finding in phaeochromocytoma patients. A 24 h collection urine sample revealed slight increases in (VMA), norepinephrine and dopamine. Alterations in plasma and urine catecholamines may be caused by a number of factors including emotional and physical stress, Valsalva manoeuvre, drugs and alcohol [10, 11, 16]. A number of drugs have been noted to alter catecholamine metabolism or interfere with biochemical assay [16]. Although the patient was not on a specific medication noted for this, he was taking melatonin which has not been widely studied in this regard.

It was noted intraoperatively that palpation of the bronchogenic cyst with the closely applied left adrenal gland produced an increase in systolic blood pressure and heart rate. This phenomenon, presumably mediated through catecholamine release, conceivably could occur with sudden adjacent pressure from an expanding mass, or more rarely, recent haemorrhage into the bronchogenic cyst, which was documented. The patient’s postoperative 24 h urine norepinephrine and VMA levels were completely normal.

This case confirms previously published reports of retroperitoneal bronchogenic cysts and further emphasizes the need to include bronchogenic cysts in the differential diagnosis of those cysts that can occur in the retroperitoneum.

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