

A CASE OF RETROPERITONEAL BRONCHOGENIC CYST

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A 59-year-old hypertensive man was referred to our hospital with a retroperitoneal cystic tumor, measuring 6 cm in diameter that was detected by an ultrasound examination during routine check-up 2 years before coming to our department. During the 2-year follow-up, the cystic tumor gradually increased in size. The patient also became hypertensive with slightly elevated urine levels of noradrenaline and dopamine, while the plasma catecholamines and their metabolites in the urine were within the normal range. Computed tomographic scanning and magnetic resonance imaging revealed a dumbbell-shaped retroperitoneal cyst with dense fluid, measuring 7×3.5×3 cm in diameter, in the left supra-adrenal and sub-diaphragmatic regions. He underwent extirpation of the cystic tumor with suspicion of adrenal endocrine cystic tumor. The histopathological diagnosis was a bronchogenic cyst, which is an extremely rare developmental anomaly in the retroperitoneal space. We herein report this rare case of retroperitoneal bronchogenic cyst and present a brief review of the previously reported 30 Japanese cases.

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INTRODUCTION

A bronchogenic cyst is an unusual developmental anomaly and very rarely presents as a retroperitoneal cystic tumor^{1,2)}. Bronchogenic cysts usually develop on the posterior side of the mediastinum along the right paratracheal wall or in the pulmonary parenchyma, and are occasionally found in unusual sites such as the skin, subcutaneous tissue, pericardium, diaphragm, and intra-dural region. This anomaly is genetically considered as a developmental abnormality of the primitive foregut, which arises commonly from an accessory lung bud in the posterior part of the mediastinum after the third week of embryonic development, but it rarely migrates into the retroperitoneum.

This cystic tumor is unusually encountered in the retroperitoneal space, and there have been only 30 case reports in Japan since 1978 as well as only about 20 case reports in the English literature. We herein report this extremely rare case of a retroperitoneal bronchogenic cyst in an adult man and present a brief review of the previously reported 30 Japanese cases including the present case^{1,3–9)}

CASE REPORT

A 59-year-old hypertensive man was referred to our department for a left retroperitoneal cystic tumor, which possibly was derived from the adrenal gland or pancreas. This cystic tumor was disclosed without any symptoms by ultrasound examination during a

routine health checkup 2 years ago, and was followed up without treatment. However, the cystic tumor increased slightly in size along with an increase in his blood pressure to around 150/90 mmHg. The plasma and urine catecholamines and their metabolites, aldosterone and cortisol were measured to determine whether or not this cystic tumor had hormone-producing activity. The urinary catecholamines measurement showed increased daily output of nor-adrenaline (152 µg/day) and dopamine (1,200 µg/day). On the other hand, the plasma concentrations of these catecholamines and their metabolites in urine were within the normal levels as well as aldosterone and cortisol. The other biochemical examinations revealed mildly elevated serum levels of GOT (83 IU/l), GPT (91 IU/l), γ-GTP (185 IU/l) and LDH (235 IU/l) because he had a past history of liver dysfunction of unknown etiology.

Abdominal computed tomography (CT) scanning revealed a dumbbell-like cystic tumor, measuring about 7 cm in diameter, between the aorta and splenic hilus in the retroperitoneal space (Fig. 1). In a plain study, the cystic tumor showed high-density fluid contents, as if the fluid seemed to include mineral deposits or highly concentrated proteins. Contrast enhancement was seen slightly in the parenchymal portion on the medial side of the cyst wall. Magnetic resonance imaging (MRI) showed a cystic tumor in the left supra-adrenal region, the intensity of which was slightly lower than that of the liver and almost the

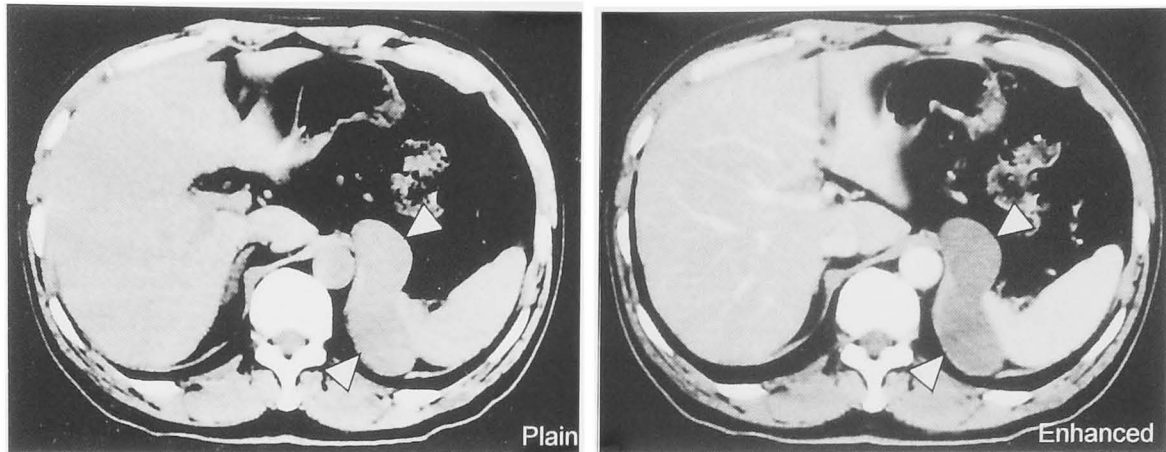


Fig. 1. Computed tomography scanning of the abdomen revealed a dumbbell-shaped cyst (arrow head), measuring about 7 cm in diameter, between the aorta and the hilus of spleen in the retroperitoneum. In a plain study, the cyst included high-density fluid contents. The cyst wall on the medial side was slightly enhanced by a contrast medium administered intravenously.

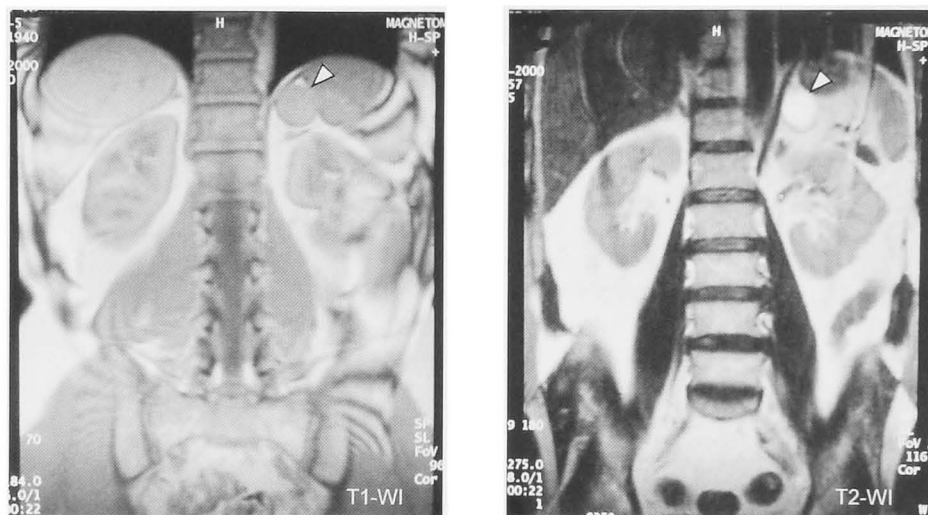


Fig. 2. Magnetic resonance images showing a cystic tumor in the left supra-adrenal region, with low and high signal intensities on T1-weighted and T2-weighted imaging, respectively. The postero-medial margin of the cyst was adjacent to the left adrenal gland, and abutted the tail of pancreas and stomach.

same as that of the skeletal muscle on T1-weighted imaging, but was much higher than that of the fat signal intensity on T2-weighted imaging (Fig. 2). The left adrenal gland was adjacent to the postero-medial margin of the cyst, which elevated the tail of pancreas and abutted the stomach anteriorly. Diagnostic imaging confirmed a retroperitoneal uncommon cyst and, therefore, there was some suspicion for concomitant malignancy and hormonal activity of this cystic tumor, probably derived from the adrenal gland, due to the above-mentioned clinical findings.

A transperitoneal exploration of this cystic tumor was performed through a sub-costal incision in the supine position. The cystic tumor was found between the left kidney, aorta and the diaphragm. It



Fig. 3. Gross appearance of the extirpated specimen that measured $7 \times 3.5 \times 3$ cm in size and showed a gourd-shaped cyst with a smooth surface including 30 ml of amber, turbid and high-viscosity fluid.

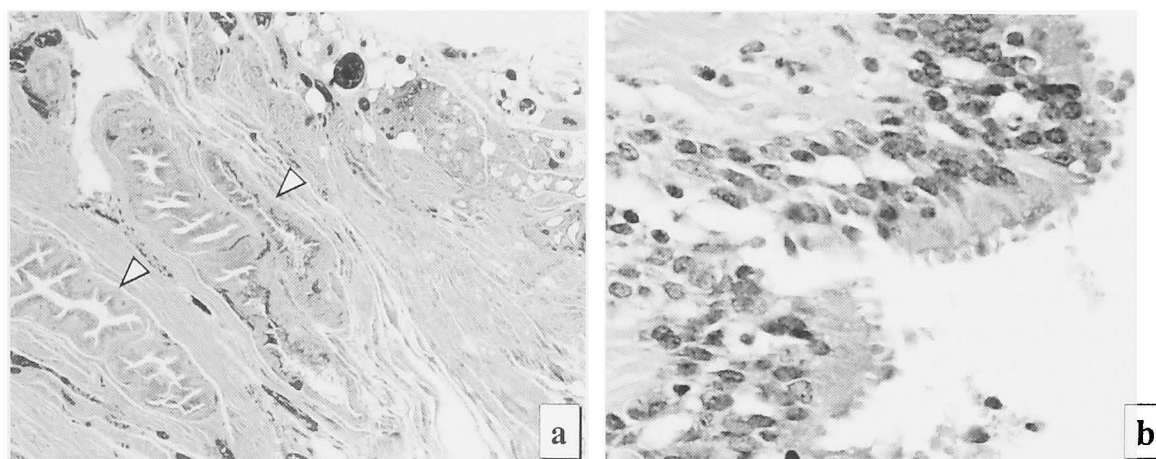


Fig. 4. Microscopically, sero-mucous bronchial glands (arrow head) surrounded by thickened smooth muscle cells were seen in the parenchymal cyst wall (a : original magnification $\times 20$, HE). The cyst wall was lined with columnar ciliated cells (b : original magnification $\times 400$, HE).

was dissected easily from the aorta and diaphragm, and was solely excised, while the adrenal gland was left in place. Macroscopically, the cystic tumor appeared as a gourd-shaped cyst with a smooth surface, measuring $7 \times 3.5 \times 3$ cm in size (Fig. 3). The cyst contained about 30 ml of amber, turbid and high-viscosity fluid. The cytological examination and bacterial culture of the fluid were negative. Microscopically, a number of bronchial glands were seen partly forming a cystic change. Mixed sero-mucous glands surrounded by a thickened smooth muscle cell layer were present in the cyst wall, resembling those in the bronchus (Fig. 4a). No hyaline cartilage was observed in the histological sections. The inner layer of the cyst wall was lined with columnar ciliated cells (Fig. 4b). On the other hand, analyzing the fluid components revealed that each catecholamine level was not significantly as high as the corresponding normal plasma concentration, and that only amylase was elevated to 456 IU/U, being mildly higher than the serum level. Now, 3 years after the operation, he maintained a spontaneous remission of hypertension without any local recurrence or complications, or any abnormal increase in the daily urinary output of catecholamines.

DISCUSSION

Bronchogenic cyst is a congenital anomaly usually developing in the mediastinum or in the pulmonary parenchyma. This entity is derived from the endodermal foregut as well as the esophagus, and develops from an abnormal bud of the primitive tracheal anlage or the bronchial tree during the first 3-6 weeks of embryonic life. If the abnormal bud migrates into the sub-diaphragmatic region before fusion of the diaphragm's components, this cyst exists incidentally and rarely in the retroperitoneal space.

In the 30 cases reported, in Japan, we found no

gender difference in the incidence of bronchogenic cysts (15 of men and women, each). The patient's age ranged from 17 to 74 years with a peak in the 40's and 50's. The mean maximum diameter was 6 ± 3 cm (ranging from 3 to 14 cm). Most bronchogenic cysts (86% of the reviewed cases) developed in the left retroperitoneal region, and this regional deviation can be explained on an anatomical basis as the liver usually disturbs the migration of the abnormal bud, which receives poor blood supply in the right para-aortic region. Although the localizations of bronchogenic cysts have been described using a variety of anatomical representations, the most common site was basically adjacent to the crus of the diaphragm. Exceptionally, several bronchogenic cysts developed in the retrovesical region or in the gastro-esophageal wall. The thoraco-abdominal type of bronchogenic cyst represents a unique dumbbell shape for its penetration through the narrow diaphragmatic slit. There were symptoms associated with this cyst in 69% of these reviewed cases, and they were mainly nausea, vomiting, and pain or discomfort in the abdomen or flank region, whereas the remaining 31% of cases had asymptomatic incidental cysts. Hypertension was a rare symptom and the catecholamines were measured in only a few cases^{2,7)} including the present case. Accordingly, the possible causality of the abnormal increase in plasma catecholamine levels and daily urinary output was not investigated or explained clearly. However, it was speculated that hypertension and elevated catecholamines might be associated with psychological stress or physical symptoms, such as pain and discomfort, before the operation. Therefore, the hypertensive symptom and abnormal catecholamine data actually disappeared postoperatively.

In our present case, plain CT imaging revealed a high-density cyst, but bronchogenic cysts showed

various patterns of CT attenuation values, which were often incompatible with the typical cyst with serous fluid density, as if the fluid seemed to include mineral deposits or highly concentrated proteins due to inflammation and hemorrhage^{3,10}. Although the differences in the CT density patterns between the bronchogenic cysts were speculated to be dependent on the fluid components, the fluid components had not been investigated in detail in the previous reports. A high level of amylase was often observed in the fluid of retroperitoneal cysts, but this was not specific for the bronchogenic cyst. On the other hand, bronchogenic cyst occasionally showed elevation of several tumor markers, such as CEA and CA19-9, in the fluid as well as elevations of amylase, calcium and iron. Unfortunately, in our present case, biochemical analysis of the fluid components was not fully performed including these tumor markers.

Surgical treatments were employed for all reported cases since the retroperitoneal cyst was not definitively diagnosed as a bronchogenic cyst until histopathological examination was done on the resected specimens. It is still difficult to correctly differentiate among several kinds of retroperitoneal cystic lesions, for example, those originating from the adrenal gland and pancreas, and to clarify their origin or coexisting malignancy only by the preoperative clinical findings. Therefore, the histological proof of the ciliated columnar epithelial cells and bronchus-like glands is necessary for a definitive diagnosis. On the other hand, the hyaline cartilage in the bronchogenic cyst was not always observed, and was only reported in less than 10% of the cases.

The incidence of malignancy in the retroperitoneal bronchogenic cysts was less than that in the thoracic bronchogenic cysts, but Sullivan et al.⁶⁾ reported a case of retroperitoneal bronchogenic cyst with malignancy. In the present case, the enhancement observed in the parenchymal portion of the cyst wall on CT scanning suggested the presence of concomitant malignancy as well. Therefore, although percutaneous ablation should be considered as a simple and minimally invasive therapeutic modality for benign cystic diseases in the retroperitoneum, it is still reasonable to treat this cystic disease with

surgical procedures including the less invasive endoscopic surgery^{4,9)}, because of the difficulty in the differential diagnosis by preoperative imaging studies, the possibility of concomitant malignancy, and the significant clinical manifestations caused by growth or localization of this cyst.

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和文抄録

後腹膜気管支原性嚢胞の1例

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症例は59歳, 男性. 2年前の健康診断時の腹部超音波断層検査で, 左後腹膜腔に長径 6 cm の嚢胞性腫瘍を指摘されたが, 無症状のため経過観察されていた. 今回, 腫瘍の増大に伴い高血圧と尿中カテコラミンの軽度高値がみられ, 精査加療を目的に当科へ紹介された. 腹部 CT および MRI で, 左副腎 横隔膜間に

径 7×3.5×3 cm で, 高密度の内容液を含むダンベル型の嚢胞性腫瘍を認めた. 内分泌活性を有する嚢胞性副腎腫瘍の疑いもあり開腹術にて摘除したが, 病理組織診断は後腹膜腔では稀な気管支原性嚢胞であった. 本邦報告30例に若干の文献的考察を加えて報告する.

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