Retroperitoneal bronchogenic cyst resembling an adrenal tumor with high levels of serum carbohydrate antigen 19-9

A case report

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Abstract

Rationale: Retroperitoneal bronchogenic cysts without specific clinical manifestations are extremely rare and difficult to diagnose preoperatively and are easily misdiagnosed as left adrenal or pancreatic tumors.

Patient concerns: A 48-year-old woman with the chief complaint of obscure epigastric pain for 1 month and with no other gastrointestinal symptoms and no significant medical history. The patient had signed informed consent for publication of this case report.

Diagnosis: The serum level of carbohydrate antigen 19-9 (CA 19-9) in the patient was >1200 U/mL, which far exceeded the normal level of <37 U/mL. Computed tomography (CT) initially suggested the presence of an adrenal tumor. However, endoscopic ultrasound (EUS) showed that the adrenal gland had an intact capsule and that the mass originated in the retroperitoneal space and did not involve the paranephros.

Interventions: Surgical resection was performed on the patient.

Outcomes: Histopathological examination demonstrated that the mass was a retroperitoneal bronchogenic cyst. At the 2-month postoperative follow-up, the level of CA 19-9 had returned to normal.

Lessons: EUS appears to be superior to CT because it clearly delineated the mass from the surrounding structures of the retroperitoneal region. EUS-fine needle aspiration can be used for diagnosis or determining whether the mass is malignant or benign.

Measurement of serum CA 19-9 may be helpful in the diagnosis of retroperitoneal bronchogenic cysts. However, this was a rare case, and the mechanism behind CA 19-9 elevation is not clear and needs further investigation.

Abbreviations: 17-OHCS = 17-hydroxycorticosteroid, ACTH = adrenocorticotropic hormone, AFP = alpha-fetoprotein, CA 125 = carbohydrate antigen 125, CA 19-9 = carbohydrate antigen 19-9, CEA = carcinoembryonic antigen, COR = cortisol, CT = computed tomography, EUS = endoscopic ultrasound, EUS-FNA = EUS-guided fine needle aspiration, VMA = vanillylmandelic acid.

Keywords: adrenal tumor, carbohydrate antigen 19-9, endoscopic ultrasound, retroperitoneal bronchogenic cyst

1. Introduction

Bronchogenic cysts are congenital abnormalities of the tracheobronchial bud originating from the embryonic foregut. They are typically found in the thoracic cavity (especially in the mediastinum, posterior to the carina) or embedded in the pulmonary parenchyma.\textsuperscript{[1]} Bronchogenic cysts in the retroperitoneal region were first reported by Miller et al in 1953 and are extremely rare. There is no specific imaging method for preoperative diagnosis of retroperitoneal bronchogenic cysts, which can be misdiagnosed as adrenal tumors in preoperative imaging.

Carbohydrate antigen 19-9 (CA19-9) is a colorectal antigen detected by a specific monoclonal antibody. Because its levels are frequently elevated in patients with gastrointestinal carcinomas, pancreatic carcinomas, or lung adenocarcinomas, it is used as a tumor marker. However, few studies have examined the relationship between CA 19-9 levels and bronchogenic cysts.\textsuperscript{[2–4]} To our knowledge, a serum level of CA 19-9 >1200 U/mL in a patient with retroperitoneal bronchogenic cyst has not been reported before. Here, we report a case of a retroperitoneal...
bronchogenic cyst that presented as a left adrenal tumor and was accompanied by high serum CA 19-9 levels.

2. Case report

A 48-year-old woman was admitted to the People’s Hospital of Nanchuan with the chief complaint of obscure epigastric pain for 1 month. She had no other gastrointestinal symptoms and no significant medical history (e.g., hypertension, pancreatitis, or clinical signs of hypercortisolism). The results of the physical examination were unremarkable.

In laboratory tests, the serum levels of CA 19-9 and carcinoembryonic antigen were >1200 U/mL and 3.06 ng/mL, respectively (Table 1). Biochemical and other serological parameters were all within normal ranges, and endocrine evaluation for adrenal hypersecretion showed no abnormalities.

Computed tomography (CT) of the abdomen showed a hypodense mass 8 × 6 × 5.5 cm in size, adjacent to the left adrenal gland (Fig. 1). The margins of the mass and the left adrenal gland could not be clearly discriminated at the left para-adrenal level. There was some calcification in the mass, which showed no contrast after contrast medium injection and had no septations. Radiologically, the mass appeared to originate in the left adrenal gland and was diagnosed as a benign tumor.

Endoscopic ultrasound (EUS) revealed that the mass was hypoechoic, partly calcified, and adjacent to the left adrenal gland. However, the left adrenal gland and pancreas were not involved, and the tumor originated in the retroperitoneal region. Cytopathology and histopathology of the specimen obtained via EUS-guided fine needle aspiration (FNA) revealed that the tumor was filled with red blood cells, without any malignant cells.

Because the patient’s CA 19-9 levels were very high, it was possible that the mass was malignant, and laparoscopy was, therefore, performed. During the operation, the boundaries of the mass and the left adrenal gland were not clearly distinguishable. No involvement of the pancreas, stomach, diaphragm, or aorta was observed.

The mass was filled with thick hematoma and secreted materials. The entire mass was sectioned for examination. Histopathologically, the wall of the cyst contained submucosal bronchial type glands, and the cyst lining was composed of respiratory epithelium and fully developed cartilage (Fig. 2). The

Table 1

| Parameter       | Concentration | Normal range       |
|-----------------|---------------|-------------------|
| White blood cells | 4.8 × 10⁹/mL  | 4–10 × 10⁹/mL     |
| Protein         | 43.1 g/L      | 40–55 g/L         |
| CA19-9          | >1,200 U/mL   | 0–39 U/mL         |
| CEA             | 3.06 ng/mL    | 0–3.4 ng/mL       |
| AFP             | 1.13 ng/mL    | <7.8 ng/mL        |
| CA125           | 19.4 U/mL     | 0–35 U/mL         |
| 17-OHCS         | 7.86 mg/24 h (urine) | 2–8 (women) and 3–10 (men) mg/24 h |
| COR             | 419.4 nmol/L (8 AM) | 171–536 (7–10 AM) and 64–327 (6–8 AM) nmol/L |
| VMA             | 5.31 mg/24 h (urine) | 0–13.6 mg/24 h (urine) |
| ACTH            | 4.84 pmol/L   | 1.6–13.9 pmol/L   |

17-OHCS = 17-hydroxycorticosteroid, ACTH = adrenocorticotropic hormone, AFP = alpha-fetoprotein, CA19-9 = carbohydrate antigen 19-9, CEA = carcinoembryonic antigen, CA125 = carbohydrate Antigen 125, COR = cortisol, VMA = vanillylmandelic acid.
patient’s recovery was uneventful. At the 1-month postoperative follow-up visit, the level of CA 19-9 had declined to 75.36 U/mL; at the 2-month follow-up visit, it had attained normal levels. The patient had known that the clinical data (including images) would be reported and signed informed consent for publication.

3. Discussion

Bronchogenic cysts are rare developmental anomalies that result from aberrant budding of the primitive foregut. They are usually found in the diaphragm, especially in the posterior part of the carina, and rarely in the retroperitoneum. Only 62 cases of retroperitoneal bronchogenic cysts have been reported, and only 30 of those represent true retroperitoneal bronchogenic cysts according to strictly applied anatomopathological criteria.[5] Retroperitoneal bronchogenic cysts occur almost equally in men and women, mostly (82% of cases) on the left side of the retroperitoneal region.[6]

Bronchogenic cysts in deep structures, such as the posterior pelvic peritoneum, usually have no specific clinical manifestations, and thus are difficult to diagnose preoperatively and are easily misdiagnosed.[7] However, if they become large enough to compress adjacent organs, symptoms such as nausea, vomiting, and abdominal pain can occur, which facilitates diagnosis via physical examination. CT and magnetic resonance imaging are useful diagnostic modalities for retroperitoneal bronchogenic cysts, although definitive diagnosis is more difficult when the cysts have a high protein or calcium content, are large and compressive, or are accompanied by infection.[8,9] In the case presented here and in the study by Jae et al, CT initially suggested the presence of an adrenal tumor, and immediate surgical resection was not recommended owing to the absence of specific clinical symptoms.

EUS is very safe, has a low complication rate, and has been shown to be useful for preoperative diagnosis of paraesophageal bronchogenic cysts. Therefore, to further examine our patient, we performed EUS, which showed that the adrenal gland had an intact capsule and that the mass originated in the retroperitoneal space and did not involve the paranephros. As determined via examination of the EUS-FNA specimen, the mass did not contain malignant cells. Surgical resection was performed. Postoperative histopathological examination demonstrated that the mass was a retroperitoneal bronchogenic cyst.

The tumor-associated antigen CA 19-9 is not a diagnostic marker in all cancer types. However, it has high sensitivity and specificity in the diagnosis of pancreatic malignancies. The relationship between CA 19-9 levels and bronchogenic cysts is largely unknown. To the best of our knowledge, retroperitoneal bronchogenic cysts with significantly elevated serum CA 19-9 have not been reported. In our case, the preoperative serum level of CA 19-9 was >1200 U/mL, which far exceeded the normal level of <37 U/mL. Preoperative serum levels of CA 19-9 were also elevated (albeit to a much lesser extent) in patients with bronchogenic cysts in the report by Han et al.[10] (156.3 U/mL). CA 19-9 levels returned to normal following resection of the cyst in our study; postoperative CA 19-9 levels were not measured in the study by Han et al.[10]

4. Conclusion

In summary, imaging techniques may not be particularly useful when bronchogenic cysts are deeply embedded in the left upper abdomen and large enough to significantly compress adjacent organs. Moreover, differentiating bronchogenic cysts from other masses, such as hemorrhagic cysts and pancreatic, adrenal, and pseudomyxoma peritonei tumors, via a single technique is challenging. Histopathological examination is the most reliable diagnostic method for retroperitoneal bronchogenic cysts, and EUS and EUS-FNA appear to be superior to CT as they can clearly delineate the mass from the surrounding structures and provide histopathological examination. Measurement of the serum level CA 19-9 may be helpful in the diagnosis of retroperitoneal bronchogenic cysts. However, this was a rare case, and the mechanism behind CA 19-9 elevation is not clear. Further studies are needed before definitive conclusions can be made.

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