Case report - Thoracic non-oncologic

Esophageal cyst producing CA19-9 and CA125

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1. Introduction

Esophageal cyst is known as a congenital esophageal malformation mainly formed in children [1]. In our case, the serum and intracystic CA125 values were significantly elevated. Although a few cases of CA19-9-producing esophageal cyst have been reported, CA125-producing esophageal cyst is extremely rare [2]. Herein, we report a resected case of esophageal cyst producing CA19-9 and CA125.

2. Case report

The patient was a 59-year-old woman in whom computed tomography revealed a posterior mediastinal cyst and ovarian cystoma at a medical check-up in March 2007. Blood tests showed high CA19-9 and CA125 levels. She underwent left adnexectomy for ovarian cystoma in July 2008. Pathological examination showed that the cystoma lumen contained hair and was lined by epidermis-like stratified squamous epithelium. No cartilage or bronchial glands were identified. These findings led to a diagnosis of esophageal cyst. On immunohistochemical staining, the cyst-lining epithelial cells were positive for CA19-9 and CA125. The serum and intracystic CA125 values were significantly increased, and then the cyst was resected completely. The CA19-9 fluid from the cyst. Firmly adherent portions were cauterized during the dissection, resulting in the ejection of greenish fluid. The firm and thickened wall of the cyst was firmly attached to the esophagus that its wall was ruptured. The cyst contained greenish fluid with CA19-9 and CA125 contents of 65,000 and 78,000 U/ml, respectively. Histologically, the cyst had a thickened wall, which contained two muscle layers, and was lined by squamous and pseudostratified ciliated epithelium. No cartilage or bronchial glands were identified. These findings led to a diagnosis of esophageal cyst. On immunohistochemical staining, the cyst-lining epithelial cells were positive for CA19-9 and CA125. The serum CA19-9 and CA125 levels returned to normal two months after surgery. We report a resected case of esophageal cyst producing CA19-9 and CA125.

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Fig. 1. Radiologic and macroscopic findings. (a) Chest X-ray showed an ovoid, well-defined, regular-bordered mass shadow of 60 mm in diameter in the posterior mediastinum in the midline and slightly to the left. (b) Chest CT revealed a 60-mm-diameter cyst in contact with the esophagus and posterior surface of the pericardium. (c) Gross appearance of the resected specimen showing a well-encapsulated cyst with a wall thickened in places.

Fig. 2. Histologically, the cyst had a thickened wall, which contained two muscle layers and, in some areas, Auerbach’s plexus (Fig. 2a). No cartilage or bronchial glands were identified. The cyst was lined by pseudostratified ciliated and squamous epithelium (Fig. 2b and c). These findings led to a diagnosis of esophageal cyst. No epithelial atypia was present. On immunohistochemical staining, the cyst-lining epithelial cells were positive for CA19-9 and CA125 (Fig. 2d and e). The patient is now leading a normal healthy life. The serum CA19-9 and CA125 levels returned to normal two months after surgery. We plan to follow her postoperative course in the outpatient clinic.

3. Discussion

Esophageal cysts originate from the endodermal foregut and are often associated with maldevelopment [1, 3]. Among cysts of foregut origin, those containing cartilage or seromucinous respiratory glands are designated as bronchogenic cysts, while those containing two well-developed layers of smooth muscle without cartilage are designated as esophageal cysts [4, 5]. Bronchogenic cysts usually contain milky white material, while esophageal cysts are filled with greenish mucus, as in this case [5].

Benign disorders associated with CA125 serum elevation involve benign ovarian tumors, ectopic pregnancy, salpin-
gitis, uterine myoma, cirrhosis, active hepatitis, pancreatitis, renal failure, bronchitis, pneumonia, and diabetes [6]. In the present case, the elevation of CA19-9 and CA125 was initially thought to be due to the presence of the ovarian cystoma, but was found to be due to the presence of an esophageal cyst producing CA19-9 and CA125. Although a few cases of CA19-9-producing esophageal cyst have been reported, CA125-producing esophageal cyst is extremely rare [2].

The cyst fluid contained high levels of CA19-9 and CA125. On immunohistochemical staining, the ciliated epithelial cells were positive for CA19-9 and CA125, suggesting that the lining epithelial cells of the esophageal cyst produce CA19-9 and CA125. Leakage of the cystic fluids into the serum is responsible for increased levels in the serum. During surgery, the intracystic pressure was high, and rupture of the cyst resulted in the ejection of a large volume of cystic fluid. It appears that the cyst-lining epithelial cells produced an excessive amount of fluid that accumulated in the cyst, causing it to enlarge over time. After resection, the serum level of CA19-9 and CA125 returned to the normal range, and this also clearly indicated that the esophageal cyst had caused the increased serum level of CA19-9 and CA125 in this case.

Esophageal cysts should be surgically extirpated including the entire cyst wall [5]. The conventional method involves the enucleation of the esophageal cyst via a posterolateral thoracotomy. In recent years, some cases of video-assisted thoracoscopic surgery (VATS) for esophageal cyst have been reported [7, 8]. However, dissection by VATS is sometimes difficult for patients in whom the cyst wall is firmly attached to the surrounding tissue, as in this patient, making open chest surgery appropriate [9, 10].

In conclusion, we report a resected case of esophageal cyst producing CA19-9 and CA125.

References