Pulmonary sequestration is defined as a congenital malformation resulting in the development of abnormal lung tissue that does not communicate with the airway system and that receives its blood supply from aberrant systemic arteries. Pulmonary sequestration is divided into two types: intralobar and extralobar. Intralobar pulmonary sequestration is usually diagnosed in early adulthood and presents as recurrent infections, hemoptysis, or pleural effusion. The sequestrated tissue is typically located in the posterior basilar segment of the lower lobe, and it is present on the left side in 60% of cases. There have been reports on elevations in tumor markers such as CA19-9 or CA125 in patients with intralobar pulmonary sequestration. Among these reports, only 4 patients have presented with both elevated serum CA19-9 and CA125 levels; no reports have involved gynecological disease.

We herein report a patient with intralobar pulmonary sequestration and bilateral ovarian cysts who presented with elevated serum CA19-9 and CA125 levels. These tumor markers decreased to within their normal ranges after resection of the left ovarian cyst and pulmonary sequestration.

A 41-year-old woman was referred to her previous hospital because of elevated serum levels of the tumor markers CA19-9 and CA125. Whole-body examination revealed an intralobar pulmonary sequestration in the right lower lobe and bilateral cystic ovarian tumors (right: 20 mm, left: 60 mm in diameter, respectively). The left ovarian cyst was resected and diagnosed as an endometrioma. The right ovarian cyst was preserved because of its small size. However, the tumor marker levels remained elevated postoperatively. S10 segmentectomy of the right lung was subsequently performed. Immunohistochemical examination of the sequestrated lung demonstrated positive staining for CA19-9 in the bronchial and alveolar epithelia and mucus. After the pulmonary resection, the CA19-9 and CA125 levels decreased to their normal ranges.

Keywords: pulmonary sequestration, ovarian cysts, CA125, CA19-9, tumor markers
revealed consolidation in the right S10 region of the lung (Fig. 1A, arrow), a large cyst in the left ovary (diameter: 60 mm), and a small cyst in the right ovary (diameter: 20 mm). Three-dimensional contrast-enhanced CT showed an aberrant artery arising from the descending thoracic aorta (Fig. 1B, arrow), and a right intralobar pulmonary sequestration was diagnosed. Venous drainage was via the pulmonary venous system. The left ovarian cyst was resected by a gynecologist because of the possibility of malignancy and was diagnosed as an endometrioma (chocolate cyst). The right ovarian cyst was preserved because of its small size. However, the tumor marker levels remained elevated after the resection of the ovarian cyst (CA19-9, 241 U/mL; CA125, 231 U/mL).

The patient was then referred to our hospital, and S10 segmentectomy of the right lung was performed. An aberrant artery arising from the descending thoracic aorta and entering the right S10 segment through the pulmonary ligament was identified intraoperatively (Fig. 2, arrowhead) and was ligated proximally and transected with a stapler. The histological findings of the resected specimen were consistent with intralobar pulmonary sequestration. The microscopic findings of the sequestrated lung showed mucus-containing dilated bronchi with infiltration of neutrophils, macrophages, and polynuclear giant cells. Immunohistochemistry demonstrated positive staining of CA19-9 in the bronchial and alveolar epithelia and mucus (Fig. 3) and negative staining of CA125 in the sequestrated lung. The postoperative course was uneventful. Two months after the pulmonary resection, the CA19-9 and CA125 levels decreased to within their normal ranges (CA19-9, 18.4 U/mL; CA 125, 18.6 U/mL).

**Discussion**

In gynecology, high serum CA19-9 and CA125 levels are considered to be markers of malignant tumors. However, these levels are also elevated in patients with benign ovarian tumors, endometriosis, and ovarian chocolate cysts. A high level and/or rapid increase in the serum CA-125 level generates strong suspicion for the eventual development of ovarian cancer in patients with endometriosis, and surgical resection should be considered in these cases. In the present case, the ovarian cysts were first considered to be the cause of the elevated CA19-9 and CA125 levels with a possibility of malignancy. Therefore, the left ovarian cyst was resected. However, these levels did not decrease to their normal ranges, which led us to perform pulmonary resection under consideration of the possibility that the increased levels of these markers were a result of pulmonary sequestration.

There have been reports on elevations of tumor markers, including CA19-9 or CA125, in patients with intralobar pulmonary sequestration. Among these reports, there have been only 4 patients with both elevated serum CA19-9 and CA125 and no reports involving gynecological disease. Although the detailed mechanisms of the increase in these tumor markers in patients with pulmonary sequestration are unknown, these reports indicate...
that chronic inflammation may promote hyperplasia of epithelial cells of the sequestrated lung. Other reports have stated that neither CA19-9 nor CA125 is a specific tumor marker and that both are synthesized by normal bronchial epithelial cells. The serum CA19-9 level is occasionally elevated in patients with certain respiratory diseases. In the present case, the serum CA19-9 and CA125 levels normalized after resection of the pulmonary sequestration. The microscopic findings suggested that the cause of elevation of these tumor marker levels was chronic inflammation in the sequestrated lung. However, the left ovarian chocolate cyst may have contributed to the elevation in these tumor marker levels because the levels decreased only slightly after the resection and did not reach their normal ranges.

Conclusion

CA19-9 and CA125 are not specific for cancer, and it is important to note that other conditions such as respiratory disease or pulmonary sequestration elevate the levels of these tumor markers even in patients with known ovarian tumors, cysts, or endometriosis.

Disclosure Statement

The authors declare no conflict of interest.

References