Perivascular epithelioid cell tumor of the lung: A case report and literature review

Shaofu Yu1,2 | Shasha Zhai3 | Qian Gong4 | Xiaoping Hu5 | Wenjuan Yang6 | Liyu Liu2 | Yi Kong2 | Lin Wu2 | Xingxiang Pu2

1Department of Clinical Pharmacy, the Second People’s Hospital of Huaihua, Huaihua, Hunan, China
2The Second Department of Thoracic Medical Oncology, Hunan Cancer Hospital, Changsha, Hunan, China
3Department of Trauma Surgery, The First Affiliated Hospital of Hunan University of Medicine, Huaihua, Hunan, China
4Department of Clinical Pharmacy, Hunan Cancer Hospital, Changsha, Hunan, China
5Department of Pathology, Hunan Cancer Hospital, Changsha, Hunan, China
6Department of Radiotherapy, Hunan Cancer Hospital, Changsha, Hunan, China

Correspondence
Lin Wu and Xingxiang Pu, The Second Department of Thoracic Medical Oncology, Hunan Cancer Hospital, Tongzipo Road 283, Changsha, Hunan, 410000, China. Email: wulin@hnca.org.cn; puxingxiang@hnca.org.cn

Funding information
the Beijing CSCO Clinical Oncology Research Foundation, Grant/Award Number: Y-XD202001-0215; the Hunan Provincial Natural Science Foundation of China, Grant/Award Number: 2019jj80018

INTRODUCTION

The perivascular epithelioid cell tumor (PEComa) is a very rare mesenchymal neoplasm characterized by specific histological and immunohistochemical features. A primary PEComa commonly occurs in the uterus, kidney and liver, but not in the lung. The clear cell sugar tumor (CCST) is a subtype of PEComa that was first described in the lung by Liebow and Castleman in 1963. This study reports a single case of lung CCST observed at the Second Department of Thoracic Oncology of Hunan Cancer Hospital (China). We also conducted a literature review and discussed the overall findings.

CASE PRESENTATION

A 48-year-old man was admitted at the Second Department of Thoracic Oncology of Hunan Cancer Hospital on March 25, 2020 due to a lung mass. The lung mass was diagnosed 1 month before. The chest computerized tomography (CT) performed on March 27, 2020 showed enlarged mediastinal lymph nodes and a mass in the apical segment of the right upper lobe (Figure 1). No abnormalities were found using bone single-photon emission computed tomography (ECT), brain magnetic resonance imaging (MRI) (Figure 2), and tumor markers. The needle biopsy pathology of the lung mass performed on April 10, 2020 revealed an epithelioid tumor.
The immunohistochemical evaluation showed a positive immunoreactivity for PNL-2, MelanA, SMA, HMB45, and VIM, but not for SOX-10, S-100, CK, LCK, CK7, CK5/6, P40, TTF-1, Napsin A, Syn or CgA, which matched a PEComa.

On May 10, 2007, the patient underwent a resection of a left renal angiomyolipoma (AML). He reported a history of hypertension for 4 years, regularly treated with nifedipine (20 mg oral tablets, once per day). He reported no history of

**Figure 1** Chest CT images showed a mass in the apical segment of the right upper lung and enlarged mediastinal lymph nodes

**Figure 2** No abnormalities were reported with bone single-photon emission computed tomography or brain magnetic resonance imaging
smoking, alcohol consumption or tumor-related family history.

There were no contraindications to surgery at the preoperative examinations. The patient received surgical treatment on April 22, 2020. The mass (size 5.0 × 4.0 × 4.0 cm) in the right upper lung invaded the chest wall with no clear boundaries to the vessels. The mediastinal lymph nodes were enlarged. There was no pleural effusion. No other nodules were observed in the remaining lung parenchyma. During the surgical operation, the gross specimens were

**Figure 3** The needle biopsy of the lung mass showed an epithelioid tumor

**Figure 4** Postoperative examination revealed a CCST of the right upper lung

**Figure 5** The comparison of CT images obtained on July 10, 2020 (b and d) and March 27, 2020 (a and c) showed a larger mass in the apical segment of the right upper lung. The mediastinal lymph nodes in group 4R were enlarged as before. Small nodules appeared in both lower lungs.
removed and the adhesion between the lung mass and the chest wall was relieved. The lymph nodes in groups 3, 7, 8, 10 and 11 were cleaned. The lymph nodes in groups 2 and 4 were enlarged and invaded the superior vena cava, thus part of them were removed. The residual tumor tissues were cauterized and R2 resection was performed. After resection, the PEComa was diagnosed as stage IIIB (pT3N2M0).

The postoperative pathological examination performed on April 30, 2020 (Figure 4) revealed a CCST of the right upper lung. The tumor was a solitary nodule without an envelope, located at the periphery of the right upper lung. It was composed of large cells with clear cytoplasm, with eosinophilic granules containing glycogen. The nuclei were round or oval, centered and hyperchromatic, without mitotic figures. Most of the tumor cells were distributed in sheets around thin-walled vessels. The perivascular interstitium was characterized by hyaline degeneration or calcification. The expression of programmed cell death ligand 1 (PD-L1) was negative.

On June 4, 2020, the positron emission tomography-computed tomography (PET-CT) revealed thickened soft tissues of the right upper chest. The positron emission tomography (PET) found clumps of abnormal radioactive concentration in the corresponding area, suggesting the presence of a residual tumor. A nodular soft tissue density shadow was observed in the air space. An abnormal radioactive concentration shadow was observed in the corresponding area, suggesting the presence of lymph node metastases. There were no other abnormalities in the chest.

The comparison of CT images between July 10, 2020 and March 27, 2020 showed a larger mass in the apical segment of the right upper lung. The mediastinal lymph nodes in group 4R were enlarged as before. Small nodules
FIGURE 8  The CT images indicated a stable disease among March 10 (a, b, e, and f), June 10 (c, d, g, and h), September 16, 2021 (i, j, m, and n), and January 19, 2022 (k, l, o, and p)

FIGURE 9  The diagnostic and treatment history of the patient

TABLE 1  Types and features of PEComas

| PEComas                  | Location                                         |
|--------------------------|--------------------------------------------------|
| Angiomyolipoma           | Usually occurring in kidneys and other organs    |
| - Common subtypes: classic type, epithelioid type, lipomatoid type, myxomatous type |
| - Rare subtypes: epithelial cyst type, eosinophilic tumor type, sclerosis type |
| Lymphangioleiomyoma      | Usually occurring in lungs, followed by other organs |
| Lymphangioleiomyomatosis |                                                  |
| Clear cell sugar tumor   | Usually occurring in lungs                       |
| Malignant PEComa         | Rare                                             |

| First author (publication year) | Gender | Age | Tumor location | Tumor size (cm) | Symptoms | Pathology | Immunohistochemistry | Coexisting tumors | Resectability | Treatment | Prognosis |
|-------------------------------|--------|-----|----------------|----------------|----------|-----------|---------------------|------------------|---------------|-----------|-----------|
| ZH Wang (2021)               | F      | 56  | The lower lobe of the left lung | 6.2 × 4.5 cm | NM       | CCST      | Positive for HMB45, MelanA, CD34, and CD10; negative for PCK, EMA, CK8/18, SMA, DES, Caldesmon, S100, SOX10, and PAX8; the Ki-67 score was about 2% | No               | Yes          | Left lower lung mass resection with lymph node dissection | No recurrence or metastasis in the 6 months after surgery |
| HJ Huang (2021)              | M      | 46  | The lingual segment of the left upper lobe, partly invading the basal segment of the lower lung, adjacent to the pleura, the anterior and posterior thoracic wall and the diaphragm | Lung mass with a size of 17.0 × 14.0 × 6.0 cm; the other mass with a size of 11.0 × 7.0 × 6.0 cm at 2.0 cm from the incision margin of the lung bronchus and immediately adjacent to the visceral pleura | Cough and chest pain for more than 10 days | Malignant PEComa (85%) and adenocarcinoma, acinar subtype (15%) | Positive for VIM, HMB45, and TFE3; negative for CK, CD34, S-100, SMA, and desmin; the p53 gene mutations were about 90%; the Ki-67 score was about 90%; a small number of tumor cells forming a cribriform or glandular tubular shape were described with moderate atypia, positive for CK7 and TTF-1; negative for CK5/6 and P40; the Ki-67 score was about 15% | Adenocarcinoma, acinar subtype (15%) | Yes          | Left upper lung mass resection with lymph node dissection; six cycles of chemotherapy (epirubicin combined with ifosfamide) and afatinib targeted therapy for EGFR sensitive mutation | Stable condition at 6 months |
| LL Shen (2020)              | F      | 28  | The lower lobe of the left lung | About 49 nodules, the largest was around 2.8 cm in diameter | Left chest paroxysmal pricking | CCTL      | Positive for HMB45, CD34, and VIM; negative for cytokeratin, SMA, S-100, CD10, PAX8, desmin, and Myo-D1 | No               | Yes          | Left lower lobectomy and mediastinal lymph node dissection | No metastasis or recurrence after 6 months |
| M Wang (2019)               | M      | 61  | The lower lobe of the left lung | 0.7 × 0.7 cm | The left lower lung nodules were associated with no symptoms | PEComa    | Positive for CD34, HMB45, MelanA, S-100, and SMA; negative for AE1/3; the Ki-67 score was about 2% | No               | Yes          | Left lower lobe lobectomy and lymph node dissection | The patient is still followed up |
| JK Zhao (2019)              | M      | 54  | The middle lobe of the right lung | About 4.0 cm in diameter | NM       | Malignant PEComa | Positive for VIM, MelanA, and TFE3; negative for HMB45; the Ki-67 score was about 10% | Primary adenocarcinoma of the lower lobe of the left lung | Yes          | Tumor dissection of the middle lobe of the right lung, wedge resection of the lower lobe of the left lung and lymph node dissection; three cycles of chemotherapy (paclitaxel combined with carboplatin) | Stable conditions |
| M Sjniari (2019)            | M      | 74  | The apical portion of the right lung | About 2.8 cm in diameter | NM       | CCST      | Positive for CD10, pan-CK, MNF116, and CK7; negative for TTF-1 | No               | Yes          | Right lobectomy and mediastinal lymphadenectomy | No recurrence or metastasis after 4 years |
| EK Yeon (2018)             | M      | 58  | The lower lobe of the right lung | About 2.7 cm in diameter | NM       | CCST      | Positive for HMB-45, VIM, and CD34; negative for AE1/3 and EMA | No               | Yes          | Wedge resection of the lower lobe of the right lung | No recurrence or metastasis after surgery |

(Continues)
| First author (publication year) | Gender | Age | Tumor location | Tumor size (cm) | Symptoms | Pathology | Immunohistochemistry | Coexisting tumors | Resectability | Treatment | Prognosis |
|--------------------------------|--------|-----|----------------|----------------|----------|-----------|---------------------|------------------|---------------|-----------|-----------|
| DI Tsilimigras (2018)          | M      | 46  | The upper and middle lobe of the right lung | About 5.5 cm in diameter | Without symptoms of cough, hemoptysis, shortness of breath or voice hoarseness | COST | Positive for HMB45, MART-1, SMA, and desmin; negative for AE1/3, CK-7, CK20, and EMA | No | Yes | Right middle lobectomy and anterior upper segmentectomy | NM |
| M Chang (2018)                 | F      | 61  | The upper lobe of the left lung | 3.0 × 2.5 × 2.5 cm | Without symptoms of cough, hemoptysis or shortness of breath | COST | Positive for HMB-45 and CD34; negative for S-100, AE1/3, SMA, calponin, GFAP, desmin, TTF-1, P40, and PAX-8 | No | Yes | Thoracoscopic surgery wedge resection of the tumor | Stable conditions |
| YH Song (2017)                 | F      | 49  | The lower lobe of the right lung | 4.0 × 3.0 × 2.0 cm | Cough and chest pain | COST | Positive for HMB-45, MelanA, CD34, CD1a, and SMA; negative for CK, Syn, chromogranin S-100, TTF-1, SP-A, CD31, desmin, mucin, CK7, and CD117; the Ki-67 score was about 3–5% | No | Yes | Surgical thoracoscopic right lower lobectomy | No metastasis or recurrence after 6 months |
| A Chakrabarti (2017)           | M      | 36  | The upper and middle lobe of the right lung | 18.0 × 13.0 cm | Right-sided chest pain for 2 months and a history of hemoptysis | Malignant PEComa | Positive for TFE-3, desmin, and SMA; negative for CK, EMA, CD56, HMB-45, MelanA, S-100, myogenin, and MiTF | No | Yes | Right upper and middle lobectomy | A sensation of heaviness in the right thorax for 6 months after surgery; the CT images of the thorax showed a lung mass extending into the lower part of the neck up to the posterior paravertebral soft tissues, with erosion of the upper ribs and a metastatic lesion in the right head of the humerus |
| XY Shi (2016)                  | F      | 50  | The lower lobe of the left lung | Nodules of different sizes diffusely distributed in both lungs, with evident exudative shadows | Cough and dysnea for 60 days, hemoptysis for 40 days and fever for 7 days | Malignant PEComa | Positive for HMB45, VIM, and SMA; negative for MelanA, CDX-2, CD56, Syn, CgA, CK7, Napsin A, TTF-1, EMA, and CD10; the Ki-67 score was about 23% | No | NA | Declined further specific therapy | Rapid progressive respiratory failure, the patient died 2 weeks after the diagnosis |
| HY Kim (2016)                  | M      | 51  | The upper lobe of the right lung | About 1.0 cm in diameter | Without symptoms | PEComa | Positive for HMB-45 and MelanA; negative for S-100, CD56, Syn, CgA, TTF-1, surfactant, Napsin A, and cytokeratin; the Ki-67 score was below 2% | No | Yes | Wedge resection of the upper lobe of the right lung | NM |
| First author (publication year) | Gender | Age | Tumor location | Tumor size (cm) | Symptoms | Pathology | Immunohistochemistry | Coexisting tumors | Resectability | Treatment | Prognosis |
|-------------------------------|--------|-----|----------------|----------------|----------|-----------|--------------------|-----------------|--------------|-----------|-----------|
| HB Sun (2015)                 | F      | 78  | The lower lobe of the right lung | 3.0 × 2.5 × 2.5 cm | Without symptoms of cough, hemoptysis, chest pain, chest tightness or fever | CCST | Positive for VIM, Bcl-2, CD34, and Melan-A; negative for CK, HMB-45, SMA, and S-100; the Ki-67 score was about 1% | No | Yes | Thoracoscopic mass resection | No metastasis or recurrence after 6 months |
| WJ Liang (2015)               | M      | 63  | The upper lobe of the left lung and the anterior mediastinum | Left upper lobe mass with a size of 4.2 × 4.7 cm; mediastinal mass with a size of 6.7 × 9.8 cm | Chest pain for more than 2 months | Malignant PEComa | Surgical specimens from the left lung masses: positive for VIM, HMB-45, and Melan-A; negative for PAN-Ck, EMA, and S-100 | Mediastinal PEComa | Yes | Resection of the tumor in the left upper lung and mediastinum | The anterior mediastinal mass recurred, the metastatic tumor in the left rib enlarged in 3 months after surgery; the patient died of cardiopulmonary failure approximately 7 months after surgery |
| AH Olivencia-Yurvati (2015)   | F      | 39  | The upper lobe of the left lung | 1.1 × 1.0 cm | NM | CCST | NM | No | Yes | Wedge resection of the upper lobe of the left lung | NM |
| S Neri (2014)                 | M      | 38  | The middle lobe of the right lung | 1.8 × 1.5 × 1.3 cm | NM | CCST | Positive for HMB-45, VIM, and SMA; negative for S100, desmin, AE1/3, EMA, and CD117 | AML of the liver in 2005 | Yes | Wedge resection of the middle lobe of the right lung | No metastasis or recurrence after 13 months |
| L Deng (2013)                 | F      | 54  | The lower lobe of the right lung | 5.0 × 4.0 × 4.0 cm | Cough, hemoptysis, and chest tightness for more than 2 months | Malignant PEComa | Positive for HMB45, PNL2, and A013; negative for AE1/3, CAM5.2, and VIM | No | Yes | Resection of right lower lobe mass and mediastinal lymphadenectomy | The patient is still followed up, without metastasis |
| GX Wang (2013)                | M      | 38  | The lower lobe of the left lung | About 3.4 cm in diameter | Recurrent cough, blood-streaked sputum for 2 months, and left chest pain for 10 days | CCST | Positive for HMB45, VIM, CD34, and S-100; negative for CK, desmin, CD68, EMA, BCC, and TTF-1 | No | Yes | Wedge resection of the lower lobe of the left lung | No metastasis or recurrence after 12 months |
| B Yan (2011)                  | F      | 75  | The lower lobe of the left lung | 2.8 × 2.2 × 2.0 cm | Fever of unknown origin for 3 months | CCST | Positive for S-100, HHF35, HMB45, and VIM; negative for AE1/3, EMA, SMA, desmin, CD54, NSE, CgA, and Syn | No | Yes | Resection of left lower lobe mass | No metastasis or recurrence after 10 years |

(Continues)
| First author (publication year) | Gender | Age | Tumor location | Tumor size (cm) | Symptoms | Pathology | Immunohistochemistry | Coexisting tumors | Resectability | Treatment | Prognosis |
|-------------------------------|--------|-----|----------------|----------------|----------|-----------|----------------------|------------------|---------------|-----------|-----------|
| ZY Wang (2010)               | M      | 79  | The lower lobe of the left lung | 5.0 × 3.0 × 3.0 cm | Cough and sputum for 1 week | Malignant | PEComa | Positive for HMB45 and VIM; negative for LCA, CD138, S-100, CD34, EMA, CK, TTF-1, Syn, and NSE; the Ki-67 score was about 50% | No | Yes | Resection of left lower lobe and mediastinal lymphadenectomy; one cycle of chemotherapy (gemcitabine) after surgery, not continued because of poor tolerance | Extensive metastasis of both lungs, left pleura and lymph nodes at 3 months after surgery; the patient refused further treatment. The patient is alive with no clear symptoms at 5 months after surgery |
| T Ye (2010)                  | F      | 50  | The lower lobe of the right lung | About 4.0 cm in diameter | A sensation of chest tightness for almost 2 months | Malignant | CCST | Positive for HMB45, PN1L2, and A013; negative for VIM, AE1/3, and CAM5.2 | No | Yes | Resection of right lower lobe and mediastinal lymphadenectomy | NM |
| S Sen (2009)                 | F      | 44  | The upper lobe of the right lung | 4.0 × 3.0 cm | Headache and weakness | CCST | Positive for S-100 and HMB45; negative for CK and CD68 | No | Yes | Resection of the tumor at the right upper lung | No complication or recurrence occurred in the postoperative period |
| H G (2008)                  | F      | 54  | The lower lobe of the right lung | About 3.5 cm in diameter | Without symptoms | PEComa | No | Positive for HMB45, CD34, S-100, and Actin; negative for CK and EMA | No | Yes | Resection of right lower lung | No recurrence |
| HJ Kim (2008)              | M      | 64  | The upper lobe of the left lung | 1.2 × 1.0 cm | NM | CCST | Positive for HMB45 and S-100; negative for CK | No | Yes | Wedge resection of the tumor at the left upper lung | No metastasis or recurrence after 2 months |
| ML Policarpio-Nicolas (2008) | M      | 64  | The lateral basilar segment of the right lobe | 2.2 × 2.0 × 1.9 cm | Shortness of breath on exertion | CCST | Positive for HMB45 and MelanA; negative for EMA, AE1/3, RCC, and S-100 | No | Yes | Wedge resection of the tumor at the right lobe | NM |
| B Papla (2003)             | M      | 68  | The superior segment in the lower lobe of the right lung | About 1.2 cm in diameter | Without symptoms | CCST | Positive for HMB45, NSE, S-100, and ACT; negative for TTF-1, CgA, and CD117 | No | Yes | Wedge resection of a fragment of the right lower lobe | The postoperative course is without complications |
| ZH Ding (1996)            | M      | 34  | The posterior segment in the upper lobe of the left lung | 3.0 × 3.0 × 3.6 cm | Chest tightness, chest pain, and cough for 3 months | CCST | NM | No | Yes | Resection of left upper lobe mass | NM |
| WP Harbin (1978)          | M      | 65  | The lower lobe of the right lung | About 2.0 cm in diameter | Denied hemoptysis, sputum production, fever or weight loss | CCST | NM | No | Yes | Resection of left lower lobe | No metastasis or recurrence after 18 months |

Abbreviations: AML, angiomyolipoma; CCST, clear cell sugar tumor; F, female; M, male; NA, not available; NM, not mentioned; PEComa, perivascular epithelioid cell tumor.
appeared in both lower lungs, which were considered tumor recurrence (Figure 5).

The patient received three cycles of chemotherapy, paclitaxel (albumin bound) 400 mg combined with cisplatin 130 mg on July 17, August 8, and August 30, 2020. Due to a gastrointestinal reaction, the patient was subsequently treated with paclitaxel (albumin bound) 400 mg combined with carboplatin 450 mg for three more cycles on September 30, October 26, and November 16, 2020. The patient received three-dimensional conformal intensity modulated radiotherapy from July 21 to August 28, 2020. The overall effect was evaluated as stable disease (Figure 6).

On December 29, 2020, the patient presented with cough and chest pain. The CT images revealed a radiation pneumonitis, improved after administration of anti-inflammatory medications (Figure 7). The CT images indicated a stable disease between March 10, 2021 and January 19, 2022 (Figure 8). The diagnostic and treatment history of the patient is described in Figure 9.

**DISCUSSION**

PEComas are mesenchymal tumors radially arranged around thick-walled vessels. They are characterized by cells with clear to weakly eosinophilic, granular cytoplasm, mainly expressing melanin markers, such as HMB45, and myogenic markers, such as SMA. PEComas include a series of tumors, such as angiomyolipoma, lymphangioleiomyomatosis, clear cell sugar tumor, and malignant PEComas. The types and features of PEComas are described in Table 1.

We searched PubMed, Embase, the Cochrane Library, the China National Knowledge Infrastructure, the Wanfang Data, and the China Science and Technology Journal database with the search terms “perivascular epithelioid cell tumor”, “PEComa”, “clear cell sugar tumor” and “CCST” from inception to January 22, 2022. We retrieved 29 case reports of primary lung PEComa, which are summarized in Table 2. They included 12 female patients and 17 male patients, ranging from 28 to 79 years old (mean age 54 years). Tumors ranged from 0.7 to 18.0 cm in diameter, with an average of 4.3 cm. Fourteen patients reported symptoms, including cough, chest tightness, chest pain, and hemoptysis. The other patients were asymptomatic. Eight tumors were malignant and the remaining 21 cases were benign. All patients underwent surgical resection except one patient who refused treatment. Four patients reported coexisting tumors: a tumor with 15% adenocarcinoma features, a single adenocarcinoma in a different lobe of the same lung, a single mediastinal PEComa, and a single metachronous hepatic angiomyolipoma. Six patients did not report the follow-up situation. In the other cases, 19 patients reported no recurrence, three patients had recurrence or metastases, and two patients died before the corresponding studies were published. One patient died of respiratory failure and the other one died of cardiopulmonary failure.

CCST is a rare tumor characterized by transparent cells rich in cytoplasm and glycogen. CCST usually occurs in middle-aged or elderly people, with no significant difference in incidence between men and women. It usually occurs in lungs, followed by other organs. CCST patients are usually asymptomatic, and only a few manifest chest pain and cough. CCST is usually found by accident in the lungs, like a “coin”, usually isolated and clearly defined without impairing the pleura.

According to the pathology, CCST generally distributes like nests or flakes, with clear boundaries between the tumor and the surrounding areas. Fibrous intervals are described between nests, with tumor cells arranged radially around blood vessels. In general, immunohistochemical markers of CCST are positive for HMB45, MelanA, and SMA and negative for CK and CD10.

The main treatment of CCST is surgical resection, with postoperative follow-up. Some tumors that cannot be surgically removed underwent postoperative adjuvant radiotherapy and chemotherapy. Some CCST patients are complicated with tuberous sclerosis complex (TSC), TSC1/2 gene mutation, andactivation of the mTOR signal transduction pathway, therefore mTOR inhibitors such as everolimus or sirolimus might be used for treatment.

**CONCLUSION**

CCST of the lung belongs to a rare subtype of PEComa, generally isolated and clearly defined, which usually occurs in middle-aged or elderly people. Neoplastic cells are transparent and characterized by cytoplasm rich in glycogen. CCST patients usually report no symptoms, with only a few complaining of chest pain and cough. The main treatment is a complete surgical resection, with postoperative follow-up. Adjuvant radiotherapy and chemotherapy can be used when necessary. Patients with TSC1/2 gene mutation are sensitive to mTOR inhibitors, including everolimus and sirolimus.

In our case report, the patient performed an R2 resection. The surface of the residual tumor was cauterized. Two months after surgery, the tumor reappeared in the right upper apical segment of the lung. It was slightly larger than before. After adjuvant radiotherapy and chemotherapy, the overall efficacy was evaluated as stable disease. There were no abnormalities regarding tumor markers. The radiotherapy caused a radiation pneumonitis, which improved after treatment with anti-inflammatory medications. During follow-up, the CT images indicated a stable disease.

**FUNDING INFORMATION**

This work was funded by the Beijing CSCO Clinical Oncology Research Foundation (No: Y-YD202001-0215) and the Hunan Provincial Natural Science Foundation of China (No: 2019jj80018).

**CONFLICT OF INTEREST**

The authors report no conflicts of interest related to this study.
CONSENT FOR PUBLICATION
A written informed consent was obtained from the patient for publication of his medical history and relative records. No information that would enable his identification has been provided.

AUTHORS’ CONTRIBUTIONS
S.Y., S.Z., Q.G., L.L., and Y.K. contributed to the design of the study, and the acquisition and analysis of data. S.Y. drafted and wrote the manuscript. X.H., W.Y., L.W., and X.P. critically reviewed the manuscript. All authors read and approved the final version of the manuscript.

ETHICAL APPROVAL
The study protocol was approved by the Ethics Committee of Hunan Cancer Hospital (Appendix S1).

ORCID
Xingxiang Pu https://orcid.org/0000-0002-2743-7080

REFERENCES
1. Folpe AL, Kwiatkowski DJ. Perivascular epithelioid cell neoplasms: pathology and pathogenesis. Hum Pathol. 2010;41(1):1–15.
2. Travis WD, Brambilla E, Nicholson AG, Yatabe Y, Austin JHM, Beasley MB, et al. The 2015 World Health Organization classification of lung tumors: impact of genetic, clinical, and radiologic advances since the 2004 classification. J Thorac Oncol. 2015;10(9):1243–60.
3. Liebow AA, Castlemann B. Benign “clear cell” tumors of the lung. Am J Pathol. 1963;43:13–4.
4. Wang ZH, Wang T, Zhang XJ, et al. Hyaline cell glycoma of the lung: a case report. Chin J Thorac Cardiovasc Surg. 2021;37(8):506–8.
5. Huang HJ, Ye WB, Wen YQ, et al. Pulmonary malignant perivascular epithelioid cell tumor mixed with lung adenocarcinoma components: a case report and literature review. Chin J Thorac Respir Dis. 2021;44(5):468–73.
6. Shen LL, Lin JX, Ren ZP, Wang B, Liu Y, Yuan J, et al. Clear cell tumor of the lung could be aggressive: a case report and review of the literature. J Cardiothorac Surg. 2020;15(1):177–81.
7. Wang M, Zhang XL, Wang B, et al. Perivascular epithelioid cell tumor of the left lower lung: a case report and literature review. Jiangsu Med J. 2019;45(11):1186–8.
8. Zhao JK, Teng HH, Zhao RY, Ding W, Yu K, Zhu L, et al. Malignant perivascular epithelioid cell tumor of the lung synchronous with a primary adenocarcinoma: one case report and review of the literature. BMC Cancer. 2019;19(1):235–9.
9. Sinjari M, Miele E, Stati V, di Cristofano C, Diso D, Pecoraro Y, et al. Early clear cell “sugar” lung cancer management: a case report and a brief literature review. Thorac Cancer. 2019;10(5):1289–94.
10. Yeon EK, Kim JI, Won KY, Lee HN. Growth pattern change of a renal cell “sugar” tumor of the lung: a serial imaging surveillance over seven years. Oncol Lett. 2018;15(6):8652–4.
11. Tsulimgar D, Bakopoulou A, Ntaniannis-Stathopoulos I, et al. Clear cell “sugar tumor” of the lung: diagnostic features of a rare pulmonary tumor. Respir Med Case Rep. 2017;23:52–4.
12. Chang M, Lim D, Genovesi M. Clear cell “sugar” tumor of the lung: a case report and review of the literature. AME Case Rep. 2018;2:40–2.
13. Song YH, Chen FF, Zhang CQ, Lin X. Spindle cell subtype of pulmonary clear cell tumor with prominent calcification and malignant potential. Thorac Cancer. 2017;8(5):330–4.
14. Chakrabarti A, Bandypadhyay M, Purkayastha B. Malignant perivascular epithelioid cell tumor (PEComa) of the lung – a rare entity. Innov Surg Sci. 2017;2(1):39–42.
15. Shi XY, Long F, Bing B, et al. Rapidly progressive pulmonary malignant perivascular epithelioid cell tumor: a case report and literature review. Chin J Tuberc Respir Dis. 2016;39(10):763–7.
16. Kim HY, Choi JH, Lee HS, Choi YJ, Kim A, Kim HK. Sclerosing perivascular epithelioid cell tumor of the lung: a case report with cytologic findings. J Thoral Dis. 2016;8(3):238–42.
17. Sun HB, Han XL, Wu SL, et al. Primary clear cell tumor of lung: a case report. Chin J Lab Diagn. 2015;19(7):1209–10.
18. Liang WJ, Xu SL, Chen F. Malignant perivascular epithelioid cell neoplasm of the mediastium and the lung: one case report. Medicine. 2015;94(22):e904.
19. Olivencia-Yurvati AH, Rodriguez AE. Clear cell “sugar” tumor of the lung: benign or malignant? Int Surg. 2015;100(3):924–6.
20. Neri S, Ishii G, Aokage K, Hishida T, Yoshida J, Nishimura M, et al. Multiple perivascular epithelioid cell tumors: clear cell tumor of the lung accompanied by angiomylipoma of the liver. Ann Thorac Cardiovasc Surg. 2014;20:453–6.
21. Deng L, Zhou LP, You C, et al. Malignant perivascular epithelioid cell tumor of the lung: a case report and literature review. Jpn J Thorac Surg. 2013;22(4):362–363,365.
22. Wang GX, Zhang D, Diao XW, Wen L. Clear cell tumor of the lung: a case report and literature review. World J Surg Oncol. 2013;11:247–51.
23. Yan B, Yao EX, Petersson F. Clear cell “sugar” tumour of the lung with malignant histological features and melanin pigmentation – the first reported case. Histopathology. 2011;58(3):498–500.
24. Wang ZY, Li H, Chen QR. Primary pulmonary perivascular epithelioid cell tumor: a case report and literature review. Cancer Res Clin. 2010;22(12):801–3.
25. Ye T, Chen HQ, Hu H, Wang J, Shen L. Malignant clear cell sugar tumor of the lung patient case report. J Clin Oncol. 2010;28(31):e626–8.
26. Sen S, Senturk E, Kuman NK, Fabuscu E, Kacar F. PEComa (clear cell “sugar” tumor) of the lung: a benign tumor that presented with thrombocytosis. Ann Thorac Surg. 2009;88(6):2013–5.
27. Gu HF, Gao LC, Kang SY, et al. Pulmonary pecomia: a clinicopathologic analysis of one case and literature review. Chin J Hemorh. 2008;18(3):434–7.
28. Kim WJ, Kim SR, Choe YH, Lee KY, Park SI, Lee HB, et al. Clear cell “sugar” tumor of the lung: a well-enhanced mass with an early wash-out pattern on dynamic contrast-enhanced computed tomography. J Korean Med Sci. 2008;23(6):1121–4.
29. Policarpio-Nicolas ML, Covell J, Bregman S, Atkins K. Fine needle aspiration cytology of clear cell “sugar” tumor (PEComa) of the lung: report of a case. Diagn Cytopathol. 2008;36(2):89–93.
30. Papla B, Demczuk S, Malinowski E. Benign clear-cell “sugar” tumor of the lung – a case report. Pol J Pathol. 2003;54(3):183–5.
31. Ding ZH, Li Q, Qu HS, et al. Primary benign clear cell “sugar” tumor of lung: a case report. Chin J Thorac Cardiovasc Surg. 1996;12(2):123.
32. Harbin WP, Mark GJ, Greene RE. Benign clear-cell “sugar” tumor (of the lung) of the lung: a case report and review of the literature. Radiology. 1978;129(3):595–6.
33. Liebow AA, Castlemann B. Benign clear cell (“sugar”) tumors of the lung. Yale J Biol Med. 1971;43(4–5):213–22.
34. Santana AN, Nunes FS, Ho N, et al. A rare case of hemoptysis: benign sugar (clear) tumor of the lung. Eur J Cardiothorac Surg. 2004;25(4):562–4.
35. Gaffey MJ, Mills SE, Zarbo RJ, Weiss LM, Gown AM. Clear cell tumor of the lung. Immunohistochemical and ultrastructural evidence of melanogenesis. Am J Surg Pathol. 1991;15:1144–53.
36. Xuan LL, Wei JG, Liu HG. Pathological diagnosis and new progress of perivascular epithelioid cell tumor. Chin J Thorac Surg. 2021;50(3):282–7.
37. Bisleri JJ, Franz DN, Frost MD, Belousova E, Bebin EM, Sparagana S, et al. The effect of everolimus on renal angiomylipoma in pediatric patients with tuberous sclerosis being treated for subependymal giant cell astrocytoma. Pediatr Nephrol. 2018;33(1):101–9.
38. Brakemeier S, Vogt L, Adams I, Zukunft B, Diederichs G, Hamm B, et al. Treatment effect of mTOR-inhibition on tissue composition of renal angiomylipomas in tuberous sclerosis complex (TSC). PLoS One. 2017;12(12):e0189132.
39. Sanfilippo R, Jones RL, Blay JY, le Cesne A, Provenzano S, Antoniou G, et al. Role of chemotherapy, VEGFR inhibitors, and mTOR inhibitors in advanced perivascular epithelioid cell tumors (PEComas). Clin Cancer Res. 2019;25(17):5295–300.

**SUPPORTING INFORMATION**
Additional supporting information can be found online in the Supporting Information section at the end of this article.