Clinical Case Report

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Primary clear cell carcinoma of the trachea

A CARE-compliant case report

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Abstract

Rationale: Primary clear cell carcinoma of the lung is a rare condition, and presentation as an endotracheal lesion is even more unusual. In this report, we present a patient with clear cell carcinoma occurring in the trachea, which obstructed the tracheal lumen and lead to the respiratory distress.

Patient concerns: A 60-year old female patient was admitted due to a 6-month history of dyspnea with worsening symptoms for 1 month. Chest CT scan revealed a smooth nodular shadow with homogeneous density on the wall of upper trachea.

Diagnosis: Bronchoscopy therapy and surgical removal of the tumor were performed. The histopathological diagnosis revealed clear cell carcinoma.

Intervention: Surgical removal of the clear cell carcinoma was performed.

Outcomes: The patient recovered well after the surgery and is now being followed-up after hospital discharge.

Lessons: Bronchoscopy is an essential tool for diagnosis of tracheal clear cell carcinoma. Surgical removal should be performed if possible.

Abbreviations: APC = argon plasma coagulation, CT = computer tomography, NP = Navelbine and cisplatin.

Keywords: airway obstruction, clear cell carcinoma, trachea

1. Introduction

Clear cell carcinoma of the lung is a rare pulmonary tumor that originates from perivascular epithelioid cells, which was initially described by Liebow and Castleman in 1963.[1] According to the WHO classification of the tumor, it is identified as a subtype of undifferentiated pulmonary large cell carcinoma.[2] To date, only sporadic cases of the primary lung clear cell carcinoma have been reported and its presentation as a tracheal lesion is even rarer. Herein we present a case of a patient with dyspnea because of upper airway obstruction caused by clear cell carcinoma.

2. Case presentation

A 60-year old female patient with no history of smoking was admitted with a 6-month history of dyspnea presenting with worsening symptoms for 1 month. There was no obvious cause for patient's symptoms and no other symptoms like cough, sputum production, or chest pain were observed. Likewise there was no particular finding on physical examination. Blood tests were normal. Computer tomography (CT) scan of chest revealed a smooth nodular shadow with homogeneous density on the wall of trachea 40 mm below the glottis, measuring 16 × 17 mm and obstructing the greater part of the tracheal lumen (Fig. 1).

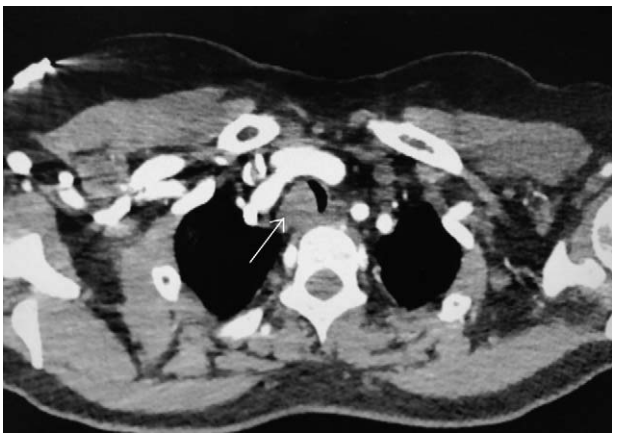

Figure 1. Contrast axial CT image shows an enhancing soft tissue density mass arising from the right posterolateral wall of the trachea. The arrow shows the tumor in the trachea.

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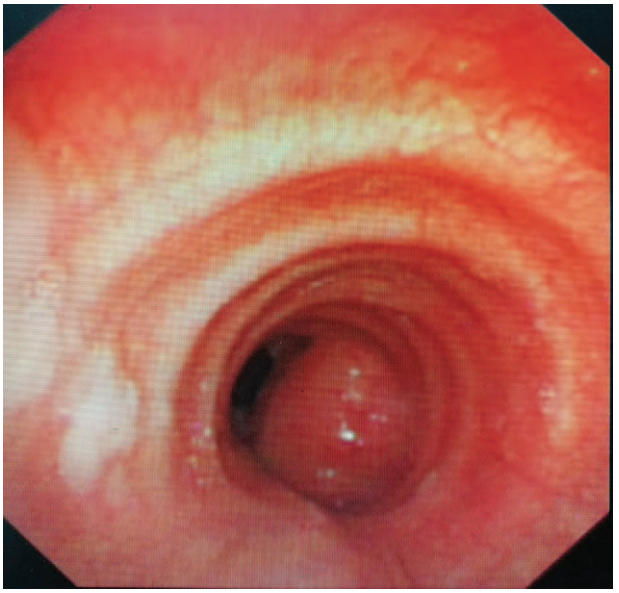

Figure 2. Bronchoscopy revealed a round, smooth-surfaced mass in the posterior wall of trachea.

Bronchoscopy was performed, and it revealed a round, smooth-surfaced, reddish mass on the posterior wall of the trachea, obstructing the tracheal lumen by approximately 70% (Fig. 2). Bronchoscopic partial resection of the tumor was performed with high frequency electric cauterization and snare excisions to obtain the pathological sample, and to relieve the obstruction. The surrounding area was coagulated by argon plasma coagulation (APC). Dyspnea of the patient disappeared after the bronchoscopic treatment. Histopathological examination demonstrated diffused growth of oval cells with abundant clear cytoplasm and distinctive cell border. The diagnosis of clear cell carcinoma was made (Fig. 3). Color Doppler ultrasonography of thyroid and abdominal CT were performed to rule out thyroid tumor or abdominal tumor, such as renal clear cell carcinoma. Both were negative. The patient was transferred to the department of thoracic surgery to receive the tracheal segmental resection. The patient recovered with satisfactory results after the surgery and now is being followed-up after discharge from the hospital.

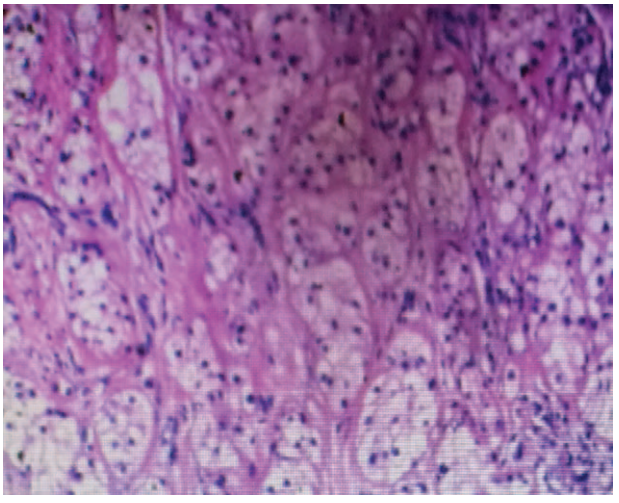

Figure 3. Histopathological examination demonstrated diffuse growth of oval cells with abundant clear cytoplasm and distinctive cell border (HE staining × 40).

A written informed consent for the case report was obtained from the patient and the consent procedure was approved by the Ethics Committee of the First Affiliated Hospital of Kunming Medical University.

3. Discussion

Primary clear cell carcinoma of the lung is a rare pulmonary tumor, which accounts for 0.3% to 3.4% of the lung cancer.[3] Presentation of primary clear cell carcinoma as an endotracheal lesion is even more unusual. To our knowledge, this is the second report about the tracheal clear cell carcinoma since Kung's report in 1984. In that case, hemoptysis was the leading clinical symptom of the patient.[4] As for the clear cell tumor in lung, most of the patients were clinically silent. Only a few showed some nonspecific symptoms like cough, dyspnea, or hemoptysis, which are not distinctive enough to differentiate from other entities. Most of the cases were discovered incidentally by routine chest radiographs or CT scan.[5,6] In our case, the tumor occurred in trachea, which obstructed the tracheal lumen and led to respiratory distress. Bronchoscopic partial resection of the tumor was hence performed to remove the central airway obstruction and to obtain the specimen for pathological examinations.

Primary tracheal tumor is relatively rare and they are rarely benign in adults; most of them being malignant, such as squamous cell carcinoma, adenocarcinoma, or adenoid cystic carcinoma.[7] Out of all the benign tracheal tumors, the most common are polyps, papillomas, lipomas, and fibromas.[8] Clear cell carcinoma of the trachea has hardly ever been reported previously and the relevant clinical data is very limited, making it more crucial to understand the biology of this rare tumor.

Typically, the biological behavior of the tumor is considered to be benign. Several literatures reported good prognosis of the clear cell carcinoma after surgical resection.[1,3,4–6,9] However, Duan et al[10] reported that in case of early blood bound metastasis of lung clear cell carcinoma, the prognosis is poor. Sale et al[11] reported a case in which a patient with lung clear cell carcinoma died from hepatic and peritoneal metastases of the tumor. Another 2 cases reported rapid tumor growth in a short period of time.[12,13] In only 1 report we can find clear cell carcinoma with trachea involvement suggesting a good prognosis of the tumor after surgical resection, with no evidence of metastasis or recurrence of the tumor during the 6-year follow-up period.[4]

As there is no definite conclusive evidence on the biological behavior of the tumor, some controversies still exist. Surgical removal of the tumor should be taken into consideration in the first place if possible. Should chemotherapy be followed? To date, the role of chemotherapy following surgery is still inconclusive with conflicting reports in the literature. In the cases by Kung,[4] Wang,[5] and Santana,[9] who believed that the primary clear cell in lung had low potential of metastasis and recurrence, the patients did not receive any adjuvant chemotherapy after surgical treatment. In Duan's report,[10] the patient received complete resection of the tumor and the postoperative chemotherapy of Navelbine and cisplatin (NP). The patient showed marked recovery in the following 16 months of follow-up period, without any evidence of metastasis or recurrence. Shomura et al reported an operative case of primary clear cell carcinoma of the lung, in which the patient received adjuvant chemotherapy with cisplatin and vinorelbine ditartrate.[14]

The patient presented with no recurrence for 3 years after surgery. In our case, the tumor grew into the trachea without invasive growth, squamous or adenomatous differentiation was not found in the histopathological examination, and the biological behavior appears to be more benign than other types of nonsmall cell lung cancer. After the complete removal of the tumor by surgery, the patient did not receive chemotherapy and is still on follow-up. As the role of chemotherapy of clear cell carcinoma in lung is dubious, more evidence-based researches are crucially desired to answer this question in the future.

In conclusion, tracheal clear cell carcinoma is rare and the clinical, imaging, and bronchoscopy findings are nonspecific and mimic other neoplasms, and the rare diagnosis of clear cell carcinoma is often established only during histopathological diagnosis. Bronchoscopy is an essential tool to obtain the pathological specimen. Surgical removal should be performed if possible once the diagnosis of tracheal clear cell carcinoma is made. The role of chemotherapy for the disease remains dubious.

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