Clear cell carcinoma of the lung revisited

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Clear cell carcinoma (CCC) of the lung is uncommon, and thus far only 51 cases have been reported in the literature. Presentation as an endobronchial lesion is even more unusual.¹ We present this case to illustrate clinical and pathologic aspects of this rare lung cancer.

Clinical Summary

A 37-year-old male patient was seen with persistent cough and intermittent hemoptysis for 3 months. Chest radiography revealed right upper lobe atelectasis. Chest computed tomography demonstrated a 4.7-cm right hilar mass closely abutting the superior vena cava, encasing the right main bronchus, and protruding endoluminally into the right main bronchus (Figure 1). There was no evidence of either nodal or distant metastasis. The patient underwent rigid bronchoscopy, which confirmed

a polypoid mass originating from the right upper lobe bronchus protruding into and

causing complete obstruction of the right main bronchus. The endobronchial mass was cored out and confirmed to be CCC. The patient underwent a few days of aggressive chest physiotherapy and then underwent right upper lobe sleeve lobectomy.

Grossly, the right upper bronchus was filled with a whitish, solid tumor 1.2 cm in diameter and 3.0 cm long, with slight extension to the right main stem bronchus. Histologically, the tumor was characterized by sheets of distinctive ovoid clear cells with ill-defined and infiltrative margins (Figure 2). The tumor had grown through the bronchus into the peribronchial cartilage and infiltrated many hilar nerves but spared the hilar lymph nodes. The bronchial epithelium adjacent to the tumor also exhibited clear cytoplasmic changes. The tumor cell ultrastructures showed primitive intercellular junctions, abundant cytoplasmic glycogen with few to many mitochondria, and a poorly developed endoplasmic reticulum. The tumor cells shared with the normal bronchial mucosa a similar immunoreactivity to cytokeratins CK7 and AE1/AE3 and to the transcriptional protein p63, whereas they were negative for the marker of renal cell carcinoma, the thyroid transcription factor-1 expressed by alveolar pneumocytes, and the neuroendocrine markers chromogranin and synaptophysin.

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Figure 1. Chest computed tomographic scan showing tumor encasing right upper lobe bronchus, attached to superior vena cava, and protruding through right main bronchus (arrow).

Figure 2. Photomicrograph of CCC showing infiltrative pattern, with clear cytoplasm and moderate pleomorphism (arrow). (Hematoxylin and eosin, ×120.)

Discussion

As a pathologic entity this particular subtype may not constitute a distinct tumor from the other types of clear cell tumors, as suggested in the early 1980s.^{2,4} However, further clinicopathologic, immunohistochemical, and ultrastructural studies of subsequent case reports support CCC as a distinct subtype of large cell carcinoma with purely clear cell features or classified into squamous or adenomatous variants, depending on the type of cellular differentiation.³ In our case, no squamous or adenomatous differentiation was seen, and the tumor grew into and completely occluded the right main bronchus. This biologic behavior appears to be more benign than that seen in other types of non-small cell lung cancer.

Conclusion

CCC has rarely been reported in the literature, and cases of endobronchial compromise are even rarer. Histopathologically, this tumor

must be differentiated from metastatic renal cell carcinoma, benign sugar tumor of the lung, predominant squamous mucoepidermoid carcinoma, and myoepithelial tumor. Immunohistochemical staining of tumor cells facilitates the differential diagnosis of these entities. Because of the low metastatic potential of these lesions and their rarity of recurrence, the role of chemotherapy for these lesions is dubious.

References

1. Iyoda A, Hiroshima K, Toyozaki T, Tsukamoto Y, Haga Y, Fujisawa T, et al. Clear cell adenocarcinoma with endobronchial polypoid growth. *Pathol Int*. 2000;50:979-83.
2. Gaffey MJ, Mills SE, Ritter JH. Clear cell tumors of the lower respiratory tract. *Semin Diagn Pathol*. 1997;14:222-32.
3. Hsu AA, Yeo CT, Ang HK, Ng HS, Ong YY. Clear cell carcinoma of the lung—a case report. *Ann Acad Med Singapore*. 1992;2:827-9.
4. Edwards C, Carlile A. Clear cell carcinoma of the lung. *J Clin Pathol*. 1985;38:880-5.

Successful enucleation of a giant cardiac hemangioendothelioma showing an unusual proliferation pattern

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Primary cardiac hemangioendothelioma is extremely uncommon, and just a few cases treated with surgical intervention are reported in the English literature.¹⁻³ We report a case involving the peculiar appearance of a huge hemangioendothelioma that arose from the right ventricle and was successfully enucleated.

Clinical Summary

A previously healthy 36-year-old woman presented at our cardiac clinic with a 4-month history of cough and increasing edema in both legs. Cardiac examination revealed a Levine grade 2/6 systolic murmur and a sinus rhythm of 76 beats/min. Blood examination was unremarkable, including tumor markers, except for slight anemia (hemoglobin of 9.2 g/dL).

Chest radiography revealed cardiomegaly. On transthoracic echocardiography, it appeared as if the right ventricle was filled by a huge mass encircled with calcification and moderate tricuspid regurgitation. Computed tomography and magnetic resonance imaging of her chest revealed pericardial effusion and a mass lesion along the inferior border of the right ventricle, with calcification of its verge (Figure 1, A). The inner space of the mass matched to a hematoma on both images. Coronary angiography from the right coronary artery (RCA) revealed that the distal RCA was extended and occluded (Figure 1, B). Coronary angiography from the left coronary artery demonstrated late filling of the occluded distal RCA by reversed flow from the left coronary artery.

We diagnosed a cardiac tumor and operated on April 30, 2002. Through a median sternotomy, the pericardium was opened. The heart was surrounded by fibrin tissue, and there was a giant cardiac tumor that pushed up the whole heart in a cephalad manner.

Cardiopulmonary bypass was established with ascending aortic, superior vena caval, and right femoral venous cannulation. After cardiac standstill, the right atrium was opened. The right atrial intima and the tricuspid valve appeared intact. The right ventricular wall, including the RCA, was thin and extended by the cardiac tumor (Figure 1, C). We then cut the right ventricular wall and removed the tumor, which could be enucleated from the right ventricular mural wall without connection to the intraventricular cavity, except for small pin holes. We repaired these small pin holes and the right ventricular wall after trimming.

Histopathologic examination showed the excised tumor was an oval monolocular cyst of 12 × 11 cm in diameter and clearly encapsulated by fibrous connective tissue. There was no extracapsular

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