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## Case Report

# A rare benign lesion in the lung: Clear Cell “Sugar” tumor (CCST)

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## ABSTRACT

Clear cell “Sugar” tumor of the lung (CCSTL) is an atypical and mostly benign peripheral primary pulmonary tumor of Perivascular Epithelioid Cell Tumors (PEComas) family. We presented a case of CCSTL of a 59-year-old woman with a well-defined, lobulated, solitary pulmonary nodule of soft tissue density located in the right lower lobe which is persistently enhancing to 120 Hounsfield Unit (HU) in the venous phase. The patient underwent a right lower lobectomy for both the curative and the diagnostic purpose. The pathological examinations and immunohistochemical studies proved the case as a benign clear cell tumor. With only a limited number of cases reported, clinicopathological aspects and radiological characteristics of this tumor are not proposed systematically. In contrast enhancement CT imaging, the persistently up sloping enhancement characteristic of nodule like in this case might be helpful to add in separating CCSTL from other lung neoplasms.

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## Introduction

It's initially described by Liebow and Castelman in 1963 [1]. Clear cell tumor of the lung (CCTL) comprises of clear cells with large amounts of intracytoplasmic periodic-acid-Schiff positive glycogen so also known as “sugar tumor”. The tumor cells show immunoreactivity for HMB-45 and S-100 protein but not for CK or EMA, and these can collectively establish the definitive diagnosis [2, 3]. In the present case, immunohistochemistry shows positive for HMB-45 and negative for CK and EMA and persistent intense post-contrast enhancement in venous phase.

## Case Report

We report here in a case of 59-year-old woman, asymptomatic with no any significant past medical and surgical history. During a general physical examination, a solitary lobulated nodule measuring 3.4cm×2.1cm in size in the right lower lobe was found accidentally on a plain chest CT (Fig. 1, A, B) with a CT value of 29 HU and no enlarged hilar or mediastinal lymph node and no calcification, necrosis, or cavitation within the lesion. She denied of smoking. Her physical examination was found to be normal. The initial basic routine blood test, CT head and abdominal ultrasonography was also normal. On contrast enhanced examination of CT chest, the nodule showed intense heterogeneous enhancement in the arterial phase with a CT value of 78

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Hounsfield units (HU) and a persistently increasing homogeneous enhancing nature in the venous phase with a CT value of 120HU (Fig. 1, C, D, respectively). Being more suspicious for malignancy, the patient has gone through right lower lobectomy for the treatment purpose and to obtain a definitive diagnosis. The intraoperative frozen section diagnosis was epithelioid tumor without malignancy. The gross appearance of the tumor was gray red, irregular, soft tissue intrapulmonary mass on cut surface and measured 3cm×2cm×1cm in size. The tumor was noncapsulated.

Histologic examination with Hematoxylin and Eosin (H&E) staining showed diffusely distributed oval shaped tumor cell with abundant clear cytoplasm (Fig.2) which was glycogen rich as shown by positive staining for Periodic-acid-Schiff (PAS). There was no necrosis or hemorrhage or mitosis. Lymphovascular invasion was absent. Immunohistochemical analysis showed the tumor had a positive reaction to Human Melanoma Black (HMB)-45, Vimentin, CD34, CD99, Ki-67(3%) and Bcl-2 but no reactivity to SMA, S-100, Melan-A, cytokeratin (CK), epithelial membrane antigen (EMA), CD56, CD10, ALK, Syn, CgA, PLAP or CD117. Based on these findings, diagnosis of benign CCTL on this patient was strongly confirmed. There was no evidence of tumor recurrence or metastasis in repeat chest CT scan in 13-months follow-up.

## Discussion

Being an extremely sporadic entity, CCSTL is still a puzzle in the field of diagnosis in pre-operative settings. It is acquiring perivascular epithelioid cells and considered as a benign PEComa in the lung, as opposed to malignant PEComa in 2015 WHO Classification of Lung Tumors [4]. No more than 60 cases of sugar cell tumors have been found in the English literature [5]. Incidence is much higher in 4th and 5th decades (range 8-73) of life and most of them are asymptomatic except for a few cases with symptoms of chest pain, hemoptysis or thrombocytosis [6]. Since it is difficult to establish specific risk factors as there is other types of small cell lung cancers tend to be very aggressive, the need for diagnosis is paramount.

CCTL radiographically presents as a solitary, round, peripheral parenchymal nodule with no evidence of cavitation or calcification and on CECT scans, they demonstrate heterogeneous intense postcontrast enhancement in arterial phase and persistent homogeneous enhancement in venous phase and our case exhibited the similar findings [7, 8]. For the first time, we are going to mention this case as having the nodule near the hilum with highly intense homogenous enhancement (120 HU) in venous phase as a benign entity. It may imply the tumor has an ample blood supply made up of large and thin-walled blood vessels indicating the tumor as being primary instead of metastasis. A recent study indicates that tumors  $\geq 2.2$ cm in diameter are more likely to have lobulated appearance and more aggressive symptoms [5]. Similar to this, our case was also suggestive of lobulation with size more than 2.2cm but didn't show any aggressive nature.

Macroscopically, CCTL appears as a pink nodule of about 2 cm (range, 1-6.5 cm) that is well demarcated from the lung parenchyma, unencapsulated, without necrosis or bleeding. Microscopic study shows typical large cells with clear abundant cytoplasm, without atypia or

mitoses [9]. CCTL is characterized immunohistochemically by immunoreactivity for HMB-45 and S-100 protein and no reactivity for cytokeratin or epithelial membrane antigen (EMA) [2, 3]. Our present case showed the similar results: positive for HMB-45 and negative for cytokeratin.

Often these tumors can be easily confused with metastases of renal clear cell tumor. These metastases also show abundant glycogen, like sugar tumors, but will show reactivity to cytokeratin as well as epithelial membrane antigen [2]. Another leading differential is a clear cell pulmonary carcinoma [2, 3]. This type will have abundant mitosis, necrosis, and reactivity to cytokeratin.

According to WHO guidelines, excision is the only treatment of choice and no adjuvant therapy is recommended [9].

## Conclusions

In brief, CCTL is very rare type of benign nodule. Immunohistochemistry is the only sure method of separating this benign tumor from other neoplasms. Surgical removal of the lesion is both diagnostic and curative. Sometimes, it could also, be a highly enhancing (120 HU) mass in venous phase should be kept in mind.

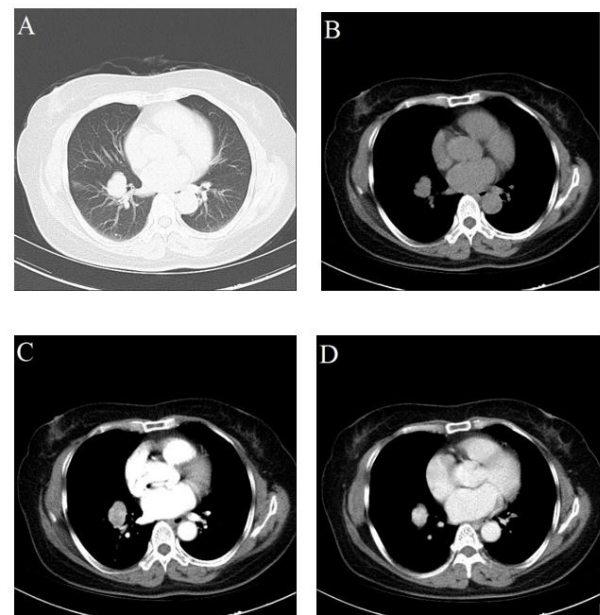


Figure 1: Chest computed tomography (CT) images of a 59-year-old woman. Showing a solitary, lobulated nodule (of 29HU) of 3.4cm×2.1cm size located in basilar segment of the right lower lobe near by the hilum in lung window (A) and mediastinal window (B). In contrast-enhanced CT imaging, revealing heterogeneous enhancement measuring 78HU in the arterial phase(C), and a markedly increased and homogeneous enhancement measuring 120HU in the venous phase(D).

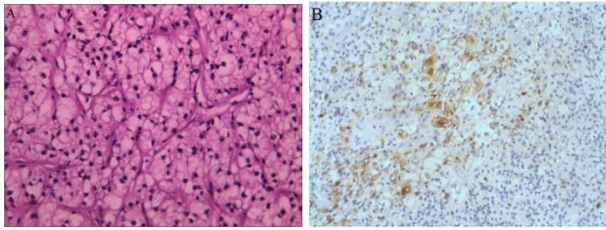


Figure 2: A: Histological examinations showed an accumulation of oval cells with diffuse distribution and abundant clear cytoplasm at high magnification(x400) B: Cytoplasmic Staining for HMB-45

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