Clear cell sugar tumor of the lung masquerading as tuberculosis in a pediatric patient

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We report the case of a 10-year-old girl with a clear cell sugar tumor (CCST) of the lung and highlight the clinical presentation and diagnostic difficulties associated with this rare tumor.

CLINICAL SUMMARY

A 10-year-old Central American girl with a history of positive purified protein derivative tests presented with decreased energy, anorexia, and pleuritic chest pain. She denied fever, shortness of breath, and cough. Social history was notable for visitors from El Salvador. Family history was strongly positive for cancer, including gastric, hepatocellular, colonic, and lymphoma. Lung examination showed no abnormalities, and no lymphadenopathy was appreciated. Laboratory studies showed a normal white blood cell count of 9.6.

Treatment with antibiotics for presumed pneumonia was initiated on the basis of an opacity in the left lung on chest x-ray film. She had no improvement in symptoms and the lesion persisted on a repeat chest x-ray film (Figure 1, A). Chest computed tomography (CT) showed a 2.7 × 2.7 × 2-cm solid nodular opacity in the left upper lobe (Figure 1, B). An ectatic bronchus passed through the superior aspect of the lesion and multiple small vessels coursed through the periphery of the lesion. Tuberculosis was considered and a purified protein derivative test was strongly positive. A 4-drug treatment regimen for tuberculosis was initiated. Examination with a flexible bronchoscope showed no abnormalities. All cultures remained negative. Repeat imaging showed an enlarging mass and operative intervention was undertaken.

A left muscle-sparing thoracotomy was performed. There was a 4-cm firm mass in the left upper lobe extending toward the hilum. A left upper lobectomy was performed. The patient had an uneventful recovery and was discharged on postoperative day 4.

Pathologic evaluation showed a firm, well-circumscribed 2.8-cm mass. Biopsy results for all surgical margins and lymph nodes were negative. Histologically, the tumor had a diffuse growth pattern of polygonal cells with abundant clear cytoplasm and distinct cell borders. The cells were strongly positive for periodic acid–Schiff and sensitive to diastase, indicative of abundant glycogen content (Figure 2). No significant mitotic activity was seen. The cells were supported by many thin-walled sinusoidal vessels. On immunohistochemistry, the tumor cells were positive for beta-hydroxy beta-methylbutyric acid (HMB)-45 and melan A and negative for cytokeratin, S-100, and smooth muscle actin (Figure 2). This histologic and immunohistochemical pattern was consistent with a benign CCST.

At her 6-month follow-up visit, the patient was doing well and there was no evidence of recurrent tumor on CT.

DISCUSSION

Originally described by Liebow and Castleman¹ in 1963, CCST of the lung is an extremely uncommon benign neoplasm. This often incidentally discovered tumor is referred to as a “sugar tumor” because of the large amount of intracytoplasmic glycogen. The tumor most commonly occurs in adults, and there are only two published reports describing the tumor in the pediatric population.^{2,3}

CCST belongs to a family of related mesenchymal lesions having perivascular epithelioid cell differentiation (PEComas).⁴ CCSTs are usually well circumscribed and solitary. They comprise round or oval cells with distinct cell borders and clear or eosinophilic cytoplasm that contains abundant glycogen. There is mild variation in nuclear size with prominent nucleoli, but mitoses and necrosis are usually absent. Thin-walled sinusoidal vessels are characteristic. There is strong diastase-sensitive periodic acid–Schiff positivity owing to the glycogen-rich cytoplasm. Common to nearly all PEComas is their distinct immunoreactivity for melanocytic (HMB-45/melan-A) markers and lack of immunoreactivity for cytokeratin. Smooth muscle (actin and/or desmin) markers are positive in 75% of CCSTs. Despite considerable advances in our recognition of these lesions, the origins of the PEC remain elusive.⁴

CCST of the lung most commonly occurs in middle-aged to elderly patients with a slight female predominance.²⁻⁴ Most patients are asymptomatic and the tumor is classically located under the visceral pleura in the periphery of the lung. CT usually demonstrates a round, peripheral, parenchymal nodule

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FIGURE 1. Radiologic imaging. A solid nodular opacity was seen in the left upper lobe on both radiography (A) and computed tomography of the chest (B).

FIGURE 2. Histology. The tumor consisted of epithelioid round to oval cells with eosinophilic cytoplasm (A) showing periodic acid–Schiff positivity that is lost after diastase treatment. By immunohistochemistry, the tumor cells are positive for HMB-45 (B).

without calcification or cavitation.⁵ The tumor is benign and surgical resection without adjuvant therapy is curative.^{2,5}

The differential diagnosis of a mass in the lung of a pediatric patient must include chronic infectious, inflammatory, and neoplastic diseases. Although there are a number of primary lung tumors, these neoplasms are all extremely uncommon in the pediatric age group, with bronchial adenoma and carcinoid being the most frequent. Metastatic lesions, especially related to Wilms tumor, osteogenic sarcoma, and hepatoblastoma, must be considered.

In summary, CCST of the lung is an extremely uncommon benign neoplasm of the lung. We report the exceptionally unusual case of a pediatric patient with this rare tumor and highlight some of the diagnostic dilemmas. An aware-

ness of this entity and recognition of its cytomorphologic and immunohistochemical features are important in making the correct diagnosis.

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