**Mediastinal PEComa with Associated Tuberos Sclerosis Complex**

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Perivascular Epithelioid Cell Tumors (PEComas) are rare benign mesenchymal tumors associated with tuberous sclerosis complex. Of these tumors, mediastinal PEComa’s are an infrequent subtype. Due to the sparsity of these tumors, mediastinal PEComa’s represent a diagnostic challenge.

A 27 year old Indian male presented to the hospital with necrotizing pneumonia, hypochromic microcytic anemia, and hypokalemia. Over the past month, patient has reported fever, left sided pleuritic chest pain, dyspnea, and productive cough. Past medical history is notable for tuberous sclerosis complex. Patient has no family history of lung disease or malignancy. Patient is a lifetime nonsmoker.

At the hospital, numerous imaging modalities are used. Chest X-Ray revealed extensive pneumonic infiltrate in the left lower lobe, emphysematous changes bilaterally, and with perihilar interstitial markings in the right upper lobe. CT revealed severe bullous emphysema and necrotizing pneumonia in both lung bases. CT also detected a large, subcarinal heterogeneous mass, measured at 6.7 x 5.5 cm. Subsequent video assisted thoracoscopy identified an encapsulated mass within a well defined envelope. Mass was later biopsied. H&E staining showed spindle cell neoplasm, consisting of cells arranged in long intersecting fascicles showing moderate pleomorphic with ovoid nuclei finely stippled chromatin, small nucleoli, and abundant fibrillary eosinophilic cytoplasm with large cytoplasmic vacuoles. Biopsy samples were immunoreactive and positive for actin, desmin, and HMB-45.

Diagnosis of PEComa was made due to immunoreactivity. Patient was started on regiment of sirolimus. After three months, tumor size had regressed to 5.0 x 4.0 cm.

This case illustrates an example of a neoplasm associated with tuberous sclerosis complex. Although this type of tumor is very rare, it is a possible complication of tuberous sclerosis complex. Furthermore, this case reveals the potential for sirolimus to be used to treat tuberous sclerosis associated neoplasms.