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TITLE PAGE

CASE REPORT:

A 65-Year-Old Female with Endobronchial Mass Lesion

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65-Year-Old Female with an Endobronchial Mass Lesion

Abstract

Glomus tumor is a rare predominantly benign soft tissue tumor. Lower respiratory tract is an uncommon site of origin of glomus tumor and hence endobronchial glomus tumor is extremely rare. This is mostly benign and identified incidentally on imaging. Diagnosis is confirmed by immunohistochemical staining and resection is the treatment of choice.

We report a middle age female with endobronchial glomus tumor which is 23rd case of reported pulmonary glomus tumor to the best of our knowledge.

Key Words

Benign, Glomus tumor, Endobronchial tumor, Soft tissue tumor, Electrocautery.

Introduction

Endobronchial mass lesion may be an incidental finding of chest imaging or it may present with symptoms of dyspnea, cough or hemoptysis. We present a case of an endobronchial mass lesion in a patient of renal cell carcinoma and CLL, which was incidentally detected on chest imaging. We will discuss the differential diagnosis of endobronchial lesions in this case review.

Case Summary

A 65-year-old female presented to clinic with a complaint of non-resolving dry cough of two months duration. There was no history of fevers, chills, rigors, chest pain, dyspnea or hemoptysis. Her past medical history was significant for chronic lymphocytic leukemia (CLL), renal cell carcinoma (RCC), hypertension, schizophrenia and depression. She had a history of 30 pack years of smoking, but no alcohol or drug use. The rest of the review of system was normal.

On examination, she was alert, awake, and oriented. Pulse 70 beats per minute, blood pressure 120/80 mm of Hg, saturation 98% on room air, weight 205 pounds. General physical examination was non-contributory with no thyromegaly, JVD or pedal edema. Cardiac examination revealed soft S1 and loud S2 with no murmur. Chest examination showed decreased air entry at the bases. Abdominal and neurological examination was normal.

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Routine laboratory tests were within normal limits except mild anemia and hypercalcemia. Hypercalcemia resolved after resection of renal cell carcinoma. EKG and pulmonary function tests were within normal limits.

Chest x-ray did not reveal any pulmonary pathology. CT scan of chest showed a right lower lobe endobronchial mass lesion of 8 mm in size **[Image 1].** No other pulmonary nodules, masses, or areas of consolidation were identified. CT scan of abdomen and pelvis revealed 6.8 cm solid left upper pole renal neoplasm and liver hemangiomas.

Bronchoscopy confirmed a large, vascular endobronchial lesion emanating from the posterior wall of right lower lobe (RLL) bronchus which obstructed it almost completely **[Image 2]**. Olympus BF-UC160F-OL8 curvilinear endobronchial ultrasound (US) probe was used to evaluate the vascularity of the tumor so that it could be resected safely. Endobronchial ultrasound revealed a highly vascular lesion. Electrocautery (up to 20 Watts) via snare loop1 was used to resect the mass. Most of the mass was resected but partial remnants were still present in the bronchus. At the end of procedure, the bronchoscope was able to pass through the lower bronchus to visualize the basilar segments, which were clear and normal in appearance.

The resected lesion was 1.2x0.4x0.5 cm aggregate of red soft tan tissue. Microscopically, it was found to be composed of small rounded cells with prominent eosinophilic cytoplasm centering on and clustering around and in between gaping vascular spaces. No mitotic activity, cellular atypia or necrosis was noted **[Image 3].** Immunohistochemical stains revealed positive staining of neoplastic cells for SMA and H-caldesmon, and negative staining for TTF1, chromogranin and synaptophysin. Immunohistochemical stain for Ki67 (MIB-1) revealed proliferation index of less than 1%. It was finally diagnosed as a benign solid glomus tumor.

DISCUSSION

Glomus tumors are rare neoplasms derived from glomus bodies of dermis and subcutaneous tissues. They account for 1.6% of all soft tissue tumors and can be present in almost all parts of body. Nail bed is the most common site for glomus tumor and extremities account for 70 % of all cases[2]. Glomus tumors are extremely rare in lower respiratory tract, accounting for around 22 cases in literature. Intrapulmonary glomus tumors are mostly benign and only 4 cases of pulmonary malignant glomus tumors have been described in literature[3]. Generally, glomus

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tumors affect young adults of 20-30 years of age with no sex predilection. However, glomus tumors specific to the lungs affect men twice as often as women with an average age of around 48 years[3].

Pulmonary glomus tumors are mostly asymptomatic and are usually detected accidentally on chest imaging. The size of most of the nodules in reported cases varies from 1 to 5 cm[3]. Chest x-ray (CXR) may not detect very small parenchymal or endobronchial lesion and appears normal in those cases. CXR and CT scan findings may vary from solitary pulmonary nodules to a partial or complete collapse of lung segments due to endobronchial lesion. On CT scan, a solitary pulmonary nodule appears as a well-delineated round mass which can distinguish it from irregular lung cancer lesions. Dynamic contrast-enhanced MRI shows strong homogenous enhancement of glomus tumor due to its vascularity. MRI can distinguish these lesions from hamartomas, which show lesser degree of contrast enhancement[4]. Glomus tumors should be kept as a differential diagnosis in case of round masses showing strong enhancement on pulmonary MRI. Despite all the advances, imaging studies cannot distinguish pulmonary glomus tumor from carcinoid tumor, hemangiomas or metastatic tumor.

Benign glomus tumors can be divided pathologically into three subtypes: glomus tumor proper, glomangioma and glomangiomyoma. Glomus tumor proper has a predominance of round glomus cells as seen in our case, while glomangioma and glomangiomyoma have a predominance of blood vessels and spindle cells respectively. Glomus tumor proper is the most common of the three followed by glomangioma and then glomangiomyoma, the rarest of all with a frequency of about 8%[2]. Malignant and atypical glomus tumors of the lung are very rare. These can be classified as: malignant glomus tumor (glomangiosarcoma), glomus tumor with nuclear atypia only (symplastic glomus tumor), glomus tumor of uncertain malignant potential, and glomangiomatosis (histologically benign glomus tumor with diffuse growth)[5].

Glomus tumors have a wide range of differential diagnoses including carcinoid tumor, smooth muscle neoplasm, paraganglioma, primitive neuroectodermal tumor (PNET), metastatic tumor and hemangiopericytoma. A uniform cell with central nuclei and clear to eosinophilic cytoplasm on histology suggests the differential diagnosis of glomus tumor, carcinoid tumor, paraganglimona and, occasionally, PNET. Diagnosis can only be confirmed on immunohistochemical staining **[Table 1]**.

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Glomus tumors are most often confused with carcinoid tumor due to simliar histology. Immunohistology is important for differentiating glomus tumor from carcinoid tumor. Glomus tumors are positive for smooth muscle actin (SMA) and type IV collagen, and are negative for neuroendocrine markers of carcinoid tumor, such as synaptophysin, chromogranin A, and cytokeratin[6]. Immunostaining in our case was strongly positive for SMA and H-caldesmon (markers of smooth muscle neoplasm) and negative for cytokeratin and neuroendocrine markers.

Hemangiopericytoma (HPC) shows branching vessels (staghorn-type vessels) in close proximity to tumor cells and is similar to glomus tumor. Microscopically, HPC does not have epithelioid cells with round central nuclei like glomus tumor but it is composed of spindle cells with elongated nuclei. HPC are negative for SMA and positive for CD 34 & 57[7].

Smooth muscle neoplasm should also be considered in the differential diagnosis of glomus tumor, where an immunohistochemical profile is less important in diagnosis than the histological appearance. Leiomyoma of the respiratory tract is an unusual tumor that involves either bronchus or alveolar parenchyma. It expresses smooth muscle actin (SMA), desmin and shows fascicles of mature, spindled, smooth muscle cells unlikely to be confused with the round or polygonal cells of glomus tumor[6].

Glomus tumors of lungs are mostly benign and complete surgical resection is the treatment of choice of these benign lesions. Glomus tumor confined to lumen of bronchus, as in our case, can be resected by bronchoscopic techniques like Nd-YAG laser, electrocoagulation, forceps extraction and cryotherapy[8]. If the tumor extends through the tracheobronchial wall or if a small tumor involves the parenchyma, limited surgical resection (sleeve and wedge parenchyma-sparing bronchial resections) would be the treatment of choice. For malignant and large parenchymal glomus tumors, sublobar resection and lobectomy are the treatment of choice[9,10]. Even with metastases, patients have an excellent prognosis.

Conclusion

Glomus tumors are rare and limited cases of endobronchial glomus tumors have been described in literature. To the best of our knowledge, this is only the 23rd case of lower respiratory tract glomus tumor to be reported. Awareness about glomus tumors as differential diagnoses of endobronchial vascular lesion is important to clinicians for effective management of these cases.

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Table 1: Immunohistochemistry for differentiating tumors similar to glomus tumor

Diagnosis	SMA	Desmin	CD34	CK	NSE	Chromogranin	S100	CD99	Collagen IV
Glomus Tumor	+	+/-	+/-	-	-	-	-	-	+
HPC	-	-	+	-	-	-	-	-	+/-
Carcinoid	-	-	-	+	+	+	-	-	-
Paraganglioma	-	-	-	-	+	+	+ (SC)	-	-
PNET	-	-	-	-	+	+/-	+/-	+	-

SMA: Smooth Muscle Actin, CK: Cytokeratin, NSE: Neuron Specific Enolase, HPC: Hemangiopericytoma, SC: Sustentacular Cells, PNET: Premitive Neuroectoderml Tumor. (**Gaertner EM et al [10]).**

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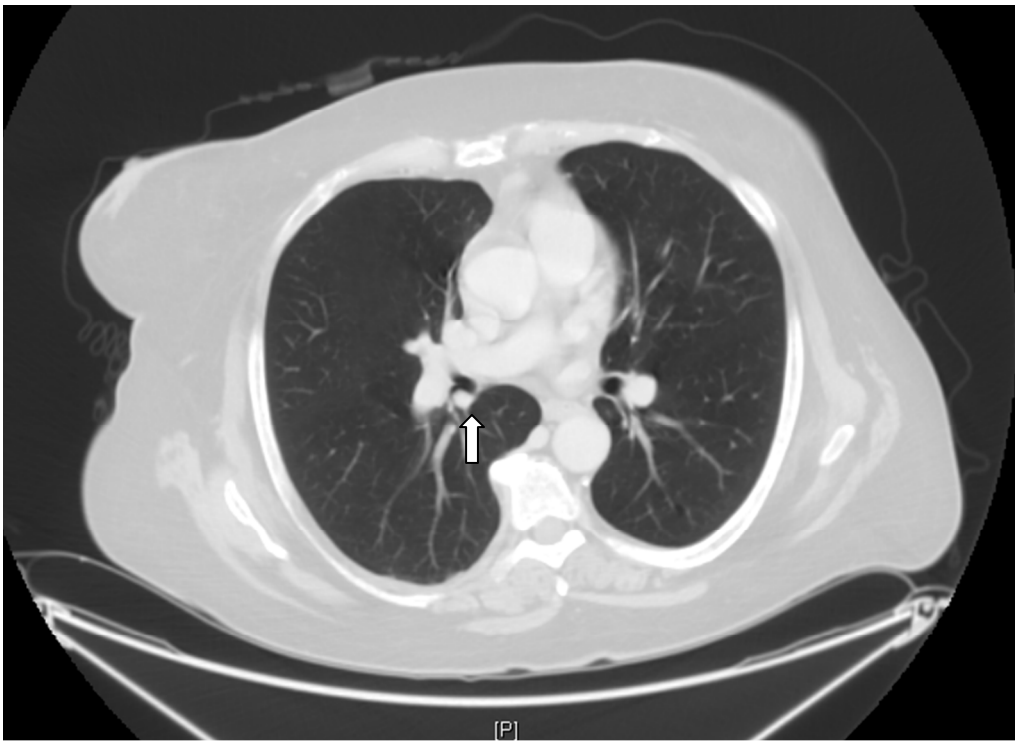
IMAGE 1



CECT chest: Showing right lower endobronchial mass lesion of 8 mm

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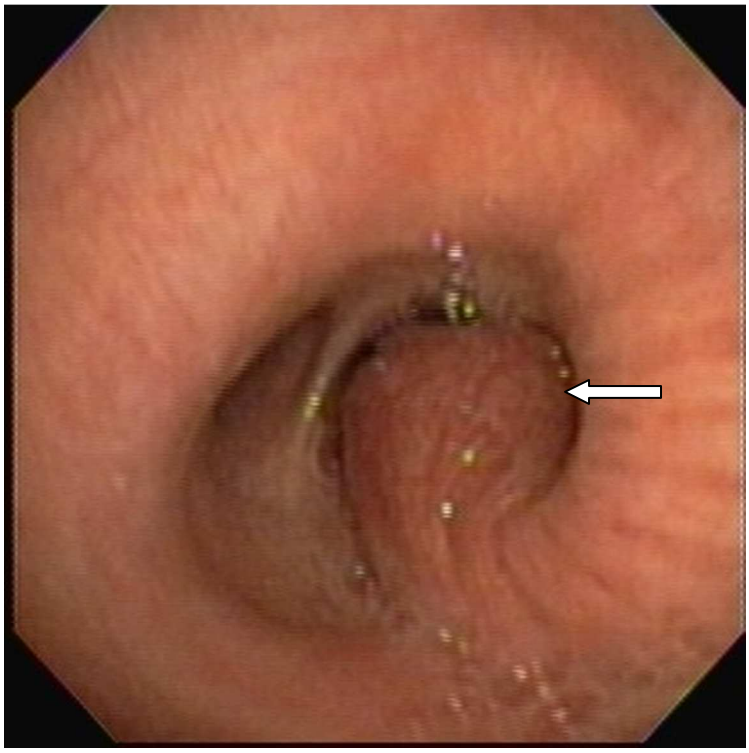
IMAGE 2



Endobronchial mass lesion- obstructing right lower bronchus

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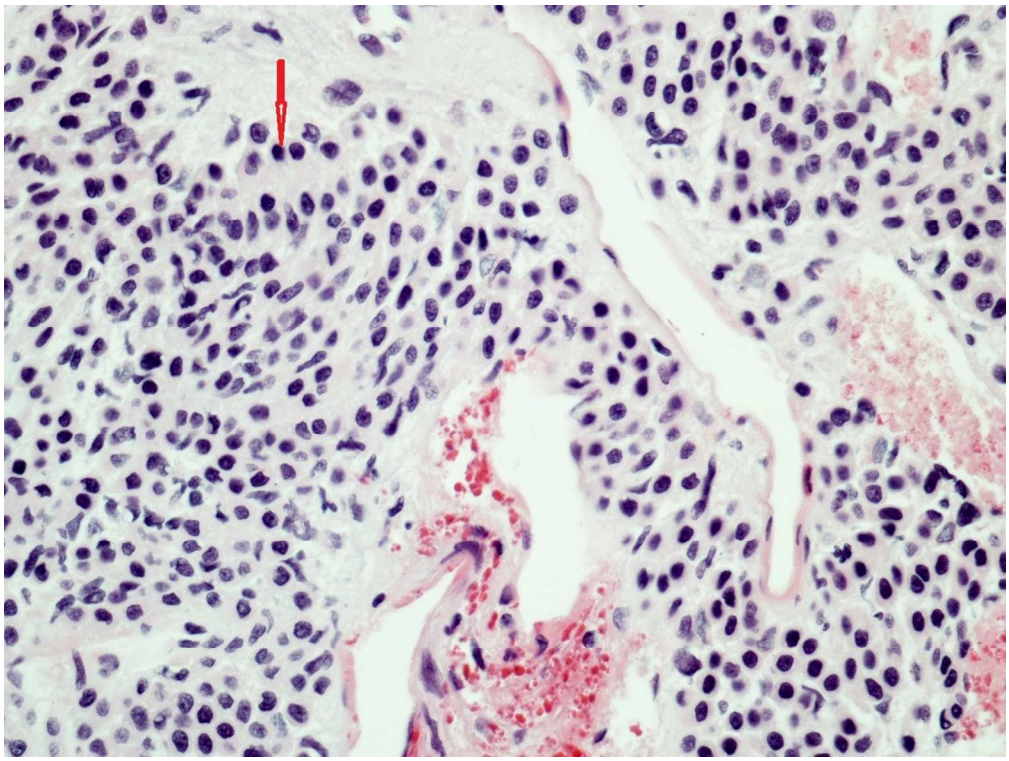

Histology: Small rounded cells with prominent eosinophilic cytoplasm centering on and clustering around and between gaping vascular spaces. No mitotic activity, cellular atypia or necrosis.
108x81mm (300 x 300 DPI)