

Primary Paraganglioma of the Lung Causing Bronchial Obstruction

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Abstract

Introduction: Paragangliomas are very rare extra-adrenal tumors that are often associated with sympathetic and parasympathetic nerve, but are rarely found in the lung.

Case Report: A 66-year-old man presents with shortness of breath and is found to have right lower lobe endobronchial lesion causing atelectasis and an associated pleural effusion. Endobronchial lesion biopsy showed unclassified epithelioid neoplasm of uncertain malignant potential. The patient underwent a right lower lobectomy and bronchoplasty of the airway. His final pathology was primary paraganglioma of the lung.

Conclusion: Here we present a very rare case of primary paraganglioma of the lung presenting with bronchial obstruction and pleural effusion. It is a rare entity that needs to be a part of the differential for possible cause of bronchial obstruction.

Keywords: Lung cancer; Paraganglioma; Pleural effusion; Endobronchial lesion.

Introduction

Paragangliomas are rare extra-adrenal tumors of the paraganglia that are often found in association with sympathetic and parasympathetic nerves. They have been documented in a variety of locations including the orbit, nose, ear, carotid, vagus nerve, larynx, mediastinum, and

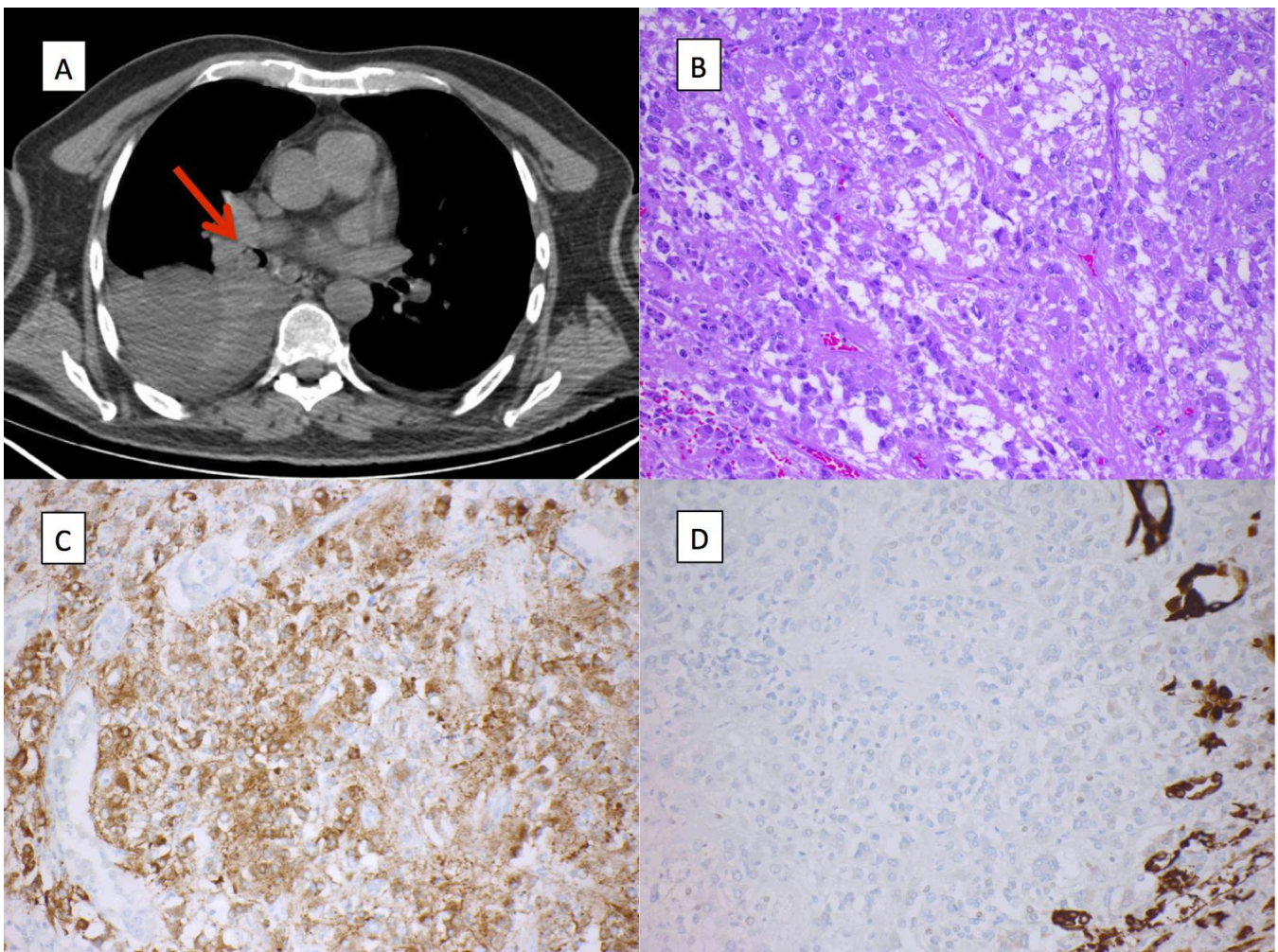
retro peritoneum, organ of Zuckerkandl, gallbladder, cauda equina, duodenum, prostate, cheek, thyroid, and lung [1]. Primary paraganglioma of the lung is quite rare with only a few cases reported since Heppleston's first report in 1958 [2]. Here we present a case of a patient who presents with primary paraganglioma of the lung causing bronchial obstruction.

Case Presentation

A 66-year-old man from Venezuela initially presented to an outside hospital with shortness of breath. A chest X-ray showed a large, right-sided pleural effusion. At that time he underwent a thoracentesis and was told he had stage IV primary lung adenocarcinoma with tumor in the right lower lobe. He presented to our institution for a second opinion. A PET/CT was performed which showed a 1.3 cm FDG-avid obstructing mass in the right lower lobe bronchus with SUV of 3.5, as well as some focal uptake along the thickened medial right lung pleura with SUV 4.3. Since the patient was symptomatic from the obstructing lesion, he was evaluated by pulmonary medicine for palliative resection of the lesion. On bronchoscopy, the patient was found to have mass in the right bronchus intermedius with tumor extending from the right lower lobe. He underwent endobronchial resection which showed unclassified epithelioid neoplasm of uncertain malignant potential. The patient underwent a repeat thoracentesis at our institution that showed no malignant cells. After partial endobronchial resection and thoracentesis, a CT was performed which showed a right lower lobe posterior hilar mass measuring 3.1 x 1.8 x 4.2 cm (Figure 1A). Considering the significant discrepancies

between pathology in Venezuela and our institution, the patient was recommended to undergo a right VATS exploration and if there were no pleural disease to undergo right lower lobectomy with mediastinal lymph node dissection.

Right VATS exploration showed no malignancy in the pleural space. Instead, the patient had a very inflamed right lower lobe with enlarged hilar lymph nodes. Due to dense adhesions and enlarged hilar lymph nodes, we converted to a thoracotomy by extending the utility incision. A right lower lobectomy was completed, however there was positive malignancy identified at the bronchial margin. Additional bronchial margin was resected and final margin was negative. We performed a bronchoplasty to reconstruct the airway without narrowing the bronchus to the right middle lobe. The patient tolerated the procedure well and was discharged home on postoperative day 4. His final pathology was stage IB paraganglioma of the lung. The tumor was composed of epithelioid cells on H&E (Figure 1B). Immunohistochemistry showed that the lesion was diffusely positive for chromogranin (Figure 1C), synaptophysin, CD56, and calretinin but negative for keratin (Figure 1D), EMA, and MOC-31.



(A) Computed tomography of the patient with obstructing lesion in the right lower lobe (arrow) with right lower lobe atelectasis and pleural effusion. (B) Image of H&E staining of the tumor composed of epithelioid cells with eosinophilic cytoplasm arranged in a nested pattern with moderate pleomorphism and nuclear atypia (100x). (C) Chromogranin immunohistochemical staining was diffusely positive (200x). (D) Keratin immunohistochemical staining was negative (200x).

Discussion

Pulmonary paragangliomas are nearly impossible to differentiate from other malignant tumors based on imaging alone, therefore histopathological evaluation is necessary for a definitive diagnosis. Pulmonary paraganglioma share many similarities with carcinoid tumors, including architectural and cytological features with a nested growth pattern and an overlapping immunohistochemical profile with chromogranin, synaptophysin, and CD56 staining [3]. In the case of paraganglioma, the positive staining of vimentin and smooth muscle differentiation as well as lack of NE, TTF-1 and

cytokeratin staining will lead to a correct diagnosis [3]. The tumors are potentially low-grade malignant, however invasive behavior has been reported such as adjacent lymph node metastasis [4]. Recommended treatment of primary paraganglioma of the lung is surgical resection [5]. Due to the rarity of this disease, the prognosis of patients with primary paraganglioma of the lung is unclear.

Conclusion

Primary paraganglioma of the lung presenting with bronchial obstruction and pleural effusion is very rare entity. It needs to be a part of the differential for possible cause of bronchial obstruction.

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