Paragangliomas are uncommon tumors arising from paraganglionic tissue dispersed from the base of skull to the pelvic diaphragm. These tumors produce symptoms by secreting catecholamines or by local tumor expansion. They can also be part of several hereditary disorders. These tumors have been reported in a variety of uncommon locations, including the lung parenchyma. Here, we report a case of primary pulmonary paraganglioma in a 46-year-old man. We also highlight the thorough evaluation for other occult tumors with emphasis on new methods of topographic diagnosis of paraganglioma.

**Case Report**

A 46-year-old man complained of cough and yellowish sputum for 2 months. A chest radiograph and computed tomography (CT) of the chest at a community hospital disclosed a mass, measuring 4.5 × 4.5 cm³, in the superior segment of the right lower lobe (RLL) of the lung near the hilum (Fig. 1). Bronchoscopic examination disclosed a polypoid mass obstructing the superior segmental bronchus of RLL. Endobronchial biopsy revealed poorly differentiated adenocarcinoma.

The patient was transferred to our chest clinic for further evaluation in June 1998. His past medical and family history were unremarkable. He was an engineer without smoking habit. On examination, his vital signs were stable. His chest expanded symmetrically with clear breathing sound. A hemogram and blood biochemistry values were all within normal limits. A whole-body bone scan revealed no abnormal focal area of increased uptake of radionuclide activity. There was no evidence of recurrence or metastasis found during the follow-up period of 3 years. Primary pulmonary paragangliomas are very uncommon tumors. Literature relevant to this disease entity is discussed.

**Key Words**

metaiodobenzylguanidine scintigraphy; paraganglioma; pulmonary neoplasm

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Correspondence to: Li-Han Hsu, MD, Division of Pulmonary and Critical Care Medicine, Sun Yat-Sen Cancer Center Hospital, 125 Lih-Der Road, Taipei 112, Taiwan. Tel: +886-2-2897-0011 ext. 1700; Fax: +886-2-2897-2233; E-mail: lhhsu@mail.kfcc.org.tw
radical lymph node dissection on July 3, 1998.

A well-circumscribed soft brownish tumor, measuring $5.5 \times 3.5 \times 3.5$ cm$^3$, with 1 cm from the bronchial resection margin was found. Grossly, the tumor was friable and sharply demarcated from surrounding lung parenchyma without invasion of the visceral pleura. There were several enlarged lymph nodes, soft and black to gray-colored, over the right interlobar, hilar, subcarinal, and paratracheal regions. On pathological examination, the tumor was composed of ovoid cells with characteristic granular amphophilic to basophilic cytoplasm (Fig. 2). The nuclei were round to ovoid with salt and pepper-type chromatin pattern.

There were giant tumor cells with intranuclear inclusions. The tumor cells were arranged in characteristic nests separated by delicate fibrovascular septa. Characteristic neuroendocrine patterns with the apical granular cytoplasm towards the capillaries rather than into the glandular lumens were also noted. Some tumor cells showed brownish pigments suggestive of lipochrome pigments or neuromelanin pigments. All resected lymph nodes were free of tumor. Immunohistochemistry demonstrated intense cytoplasmic staining reaction for synaptophysin and S-100 protein-reactive sustentacular cells. A staining pattern for neuron-specific enolase was also present. Stainings for mucicarmine, carcinoembryonic antigen (CEA) and cytokeratin were negative. The findings were consistent with paraganglioma.

In the following evaluation, there was no visible mass lesion in the whole-body CT scan imaging. The 24-hr urine vanillylmandelic acid was 6.54 mg (normal range: 1.0-7.5 mg). The patient had an uneventful recovery. There has been no recurrence or metastasis during the subsequent 3 years. In the interim, metaiodobenzylguanidine (MIBG) scintigraphy was done on May 14, 1999, which did not find any $^{131}$I-MIBG avid lesion in the whole-body survey (Fig. 3).

Fig. 1. Chest CT scan showing a well-circumscribed mass with inhomogeneous density and relatively low vascularity.

Fig. 2. Nesting ovoid cells separated by delicate fibrovascular septa are characteristic for paraganglioma. (H & E; original magnification, $\times$ 200).

Fig. 3. The $^{131}$I-MIBG scintigraphy at 48 hr after injection of 1 mCi of agent revealed no abnormal focal area of uptake in the whole-body survey.
Discussion

Intrathoracic paragangliomas are in frequent tumors. Most of them are located in the mediastinum, originating from aorticopulmonary paraganglia or paravertebral sympathetic chains. In the lung, the so-called “multiple minute chemodectomas” are more frequent than primary pulmonary paragangliomas. The former are found in an area of 3% of autopsies and occur frequently in association with chronic lung diseases. They are believed to originate from either muscle cells or cells strongly resembling meningioendothelial cells and are stimulated by ischemia. Primary pulmonary paragangliomas are very rare. The majority of them originate from the glomera in relation to the pulmonary vessels and nerves. Most are closely associated with the pulmonarv arterioles. Some authors have used this as criteria for diagnosis. According to the WHO classification, they are tumors being long to the group of parasympathetic (non-chromaffin) branchiomeriic paragangliomas, for merely also known as chemodectomas.

Since the first case of primary pulmonary paraganglioma described by Heppleston in 1958, only 23 cases have been described. It is female-predominated and usually presents as an asymptomatic, subpleural, pulmonary nodule. Cytologically, it is difficult to distinguish from bronchial carcinoid by microscopic examination, polygonal neoplastic cells or groups of cells arranged into cords, nests, and sheets with occasional acinar configuration. The former has been found to be accurate in the localization of functioning paragangliomas, especially at extra-adrenal sites. Consideration was even given to the therapeutic potential of 113-MIBG-targeted radiotherapy in neuroendocrine tumors. The latter has been found to be more accurate in the localization of functioning paragangliomas, especially at extra-adrenal sites. Consideration was even given to the therapeutic potential of 113-MIBG-targeted radiotherapy in neuroendocrine tumors.

Recently, the introduction of MIBG scintigraphy and magnetic resonance imaging (MRI) imaging has provided new insights into paragangliomas and has tremendously changed the topographic diagnosis of paragangliomas. The former has been found to be more accurate in the localization of functioning paragangliomas, especially at extra-adrenal sites. Consideration was even given to the therapeutic potential of 113-MIBG-targeted radiotherapy in neuroendocrine tumors. The latter has been found to be more accurate in the localization of functioning paragangliomas, especially at extra-adrenal sites.

Primary pulmonary paraganglioma usually presents as a solitary, peripheral asymptomatic nonfunctioning mass in the lung parenchyma. Our patient had a much more uncommon disease process comprising parenchymal and airway components.
The endobronchial lesion with obstruction accounts for his initial manifestation of cough with purulent sputum. The prognosis was determined by the tendency of neighboring lymph node invasion. Complete resection predicted a favorable outcome.

References