Pulmonary Paraganglioma Manifesting as an Endobronchial Mass

Ki Nam Kim, MD[1]
Ki-Nam Lee, MD[1]
Mee Sook Roh, MD[2]
Pil Jo Choi, MD[3]
Doo Kyung Yang, MD[4]

Thoracic paragangliomas comprise only 1–2% of all paragangliomas, including the adrenal pheochromocytomas, and these tumors are mostly found in the mediastinal compartments (1). To the best of our knowledge, there is only one case report in the pathology literature of endobronchial involvement by a primary pulmonary paraganglioma (2). We report here on the CT and bronchoscopic findings of a case of pathologically proven endobronchial paraganglioma in a 37-year-old woman. In our case, bronchoscopy and CT demonstrated an endobronchial hypervascular mass, which indicated the presence of carcinoid or hypervascular metastasis based on the known incidence of such tumors.

Index terms:
Lung neoplasms, CT
Paraganglioma

DOI:10.3348/kjr.2008.9.1.87

Korean J Radiol 2008;9:87-90
Received October 16, 2006; accepted after revision January 4, 2007.

[1]Department of Radiology, [2]Department of Pathology, [3]Department of Thoracic and Cardiovascular Surgery, and [4]Internal Medicine, College of Medicine, Dong-A University, Pusan 602-103, Korea

Address reprint requests to:
Ki-Nam Lee, MD, Department of Radiology, College of Medicine, Dong-A University, 3-ga, Dongdaesin-dong, Seo-gu, Pusan 602-103, Korea.
Tel. (8251) 240-5367
Fax. (8251) 253-4931
e-mail: gnlee@dau.ac.kr

The term "paraganglioma" is a generic term that's applied to tumors arising from the paraganglia, regardless of their location. This tumor can be found in any part of the body where healthy paraganglia are known to occur. However, the radiologic manifestations of endobronchial paragangliomas have not been well documented on account of the tumor's rarity (3–5). We present here a very rare case of primary endobronchial paraganglioma.

CASE REPORT

A 37-year-old woman presented with a 3-month history of exertional dyspnea, cough and one recent episode of hemoptysis. At the time of admission, she showed no hypertension or systemic symptoms. A chest radiograph showed a left hilar mass with a lobulated margin (Fig. 1A). The axial and coronal CT scans were obtained with using 16-channel multidetector CT after IV administration of contrast media. A mildly enhancing left hilar mass with an endobronchial protrusion and an extension along the left lingular segmental bronchus was noted (Figs. 1B, C). The lung setting image showed localized emphysema distal to the endobronchial lesion (Fig. 1D).

Bronchoscopy revealed a mutilobular, hypervasacular mass obstructing the lumen of the left lingular segmental bronchus (Fig. 1E). A careful biopsy was taken from the peripheral portion because the tumor tended to bleed. This tumor was believed to be either an endobronchial bronchogenic carcinoma or another type of hypervascular tumor. The pathologic diagnosis was chronic inflammation with granulation tissue, which was not in accordance with the radiologic findings.

We subsequently performed a left upper sleeve lobectomy with dissection of the mediastinal lymph nodes. The patient suffered no hypertensive crisis during or after surgery. The gross examination showed an endobronchially growing solid mass along the bronchial lumen, the so-called toothpaste figure, and this mass measured 7 × 3 cm in dimension. The remaining lung parenchyma showed no remarkable change (Fig.

1F). The mass showed a yellowish brown granular appearance with infiltration into the bronchial wall. The microscopic examination revealed that the tumor consisted of nests of epitheloid cells in an organoid or alveolar pattern, and this was surrounded by a delicate, richly vascular reticulin network, producing the classic 'zellballen' or basket pattern (Fig. 1G). The tumor cells showed marked nuclear pleomorphism and up to one or two mitotic figures/10 high power fields. There were associated microscopic necrotic foci. Immunohistochemical staining for chromogranin, which is a marker for neuroendocrine tumors, was strongly positive. There was no metastasis observed in the dissected lymph nodes.

After surgery, a CT evaluation of the neck and abdomen demonstrated no abnormal findings. The biochemical study revealed the following: the urine epinephrine level was 2.3 ㎍/day (0−20 for the normal range), and the norepinephrine level was 37.3 ㎍/day (15−80 for the normal range). The final diagnosis was a primary pulmonary paraganglioma with malignant potential.

DISCUSSION

Paragangliomas are rare neuroendocrine tumors arising from neuroectodermally derived paraganglionic cells that are scattered throughout the body. Paragangliomas have been described in virtually all organs, including the orbits, nasal cavity, thyroid, heart, urinary bladder, gallbladder, liver, biliary system, kidneys, prostate, urethra, spermatic cord, uterus, ovaries, vagina, vulva, cauda equina and lungs. The most common site is the superior paraaortic region between the diaphragm and the lower renal poles (approximately 46% of all cases), and particularly in and around the renal hilus (6). In contrast, primary pulmonary

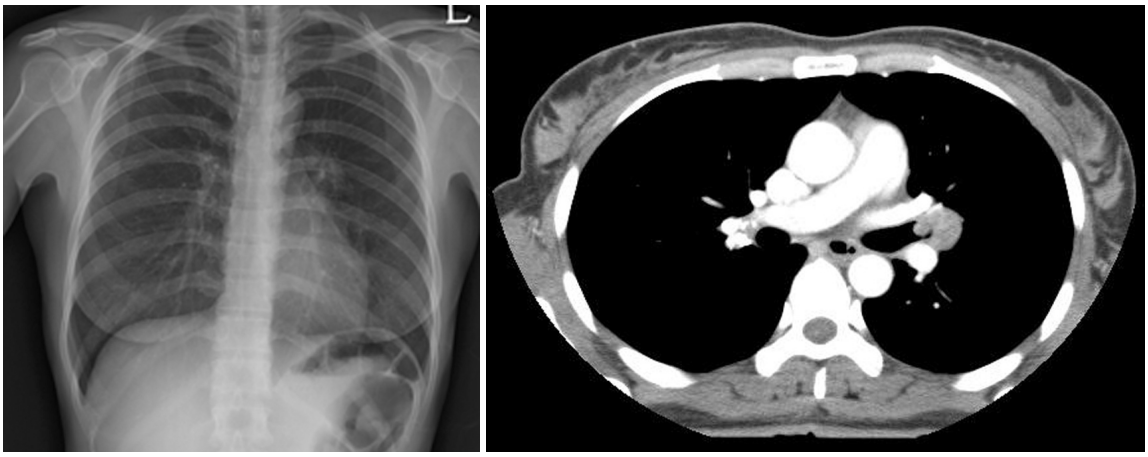
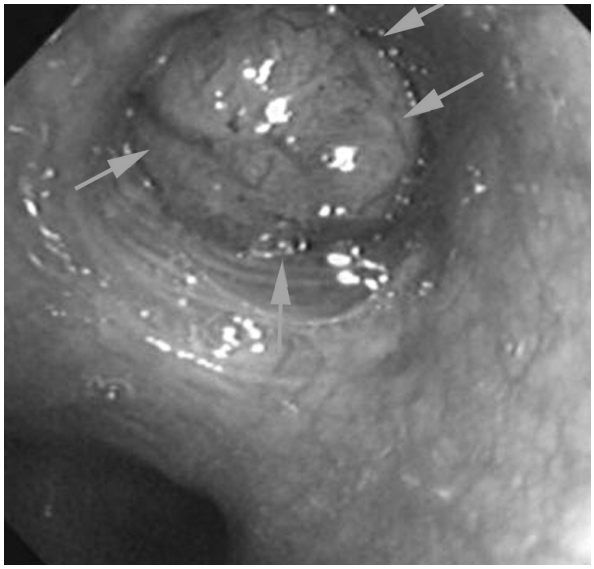
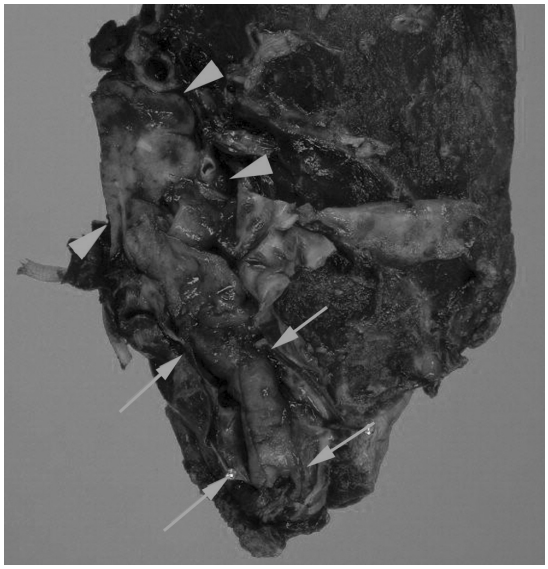


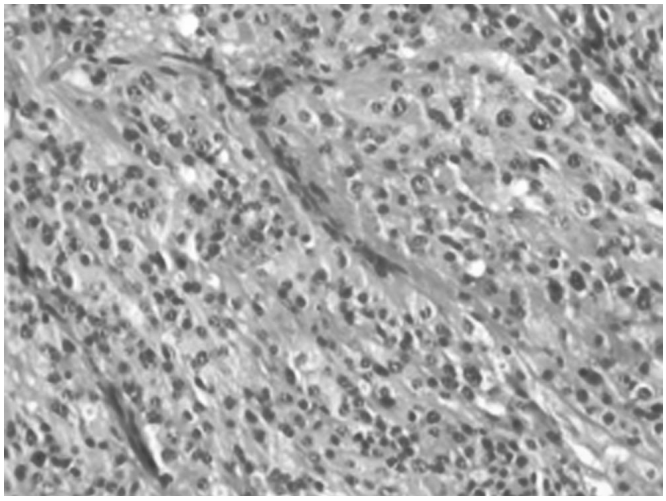
Fig. 1. A 37-year-old woman with a primary pulmonary paraganglioma.
A. The chest radiograph shows a left hilar mass with a lobulated margin.
B, C. The contrast-enhanced CT axial images show a left hilar mass with an endobronchial protrusion, which extends along the bronchial lumen, and the mass shows mild homogeneous enhancement.
D. The lung setting image shows localized emphysema distal to the endobronchial lesion.



E



F



G

Fig. 1. E. Bronchoscopy reveals a hypervascular bulging mass with a lobulated surface (arrows) in the lingular segmental bronchus.
F. Photograph of the left upper lobectomy specimen dissected along the lingular bronchial plane demonstrates a multilobulated, whitish mass (arrowheads) within a dilated left upper lobar bronchus, and this mass extends along the lingular segmental bronchus (arrows).
G. The histopathologic specimen shows that the tumor is composed of a nest of round or polygonal tumor cells separated by prominent fibrovascular septa, producing the typical "zellballen" of a paraganglioma. Also of note are the marked nuclear pleomorphism and up to one or two mitotic figures/10 HPFs, suggesting the tumor's malignant potential (Hematoxylin & Eosin staining, ×200).

tumors are extremely rare (< 1%) (6).

Most paragangliomas are benign, but a small percentage of these tumors produce distant metastases or they invade the nearby structures in the manner of malignant tumors (6). Although some reports have stated that a diagnosis of malignant paraganglioma can only be made after metastasis has occurred, some findings such as an extraadrenal location, macroscopic nodularity and tumor necrosis indicate a paraganglioma's malignant potential (7).

Metastatic paragangliomas are more frequent in the lung than a primary pulmonary paraganglioma. Since primary pulmonary paragangliomas are rare and they present no pathognomic clinical features, they are usually observed as asymptomatic solitary nodules and they can be suspected of being a primary lung malignancy. The reported incidence of metastasis of a pulmonary paragangliomas has varied and the incidence appears to be related to the length of the follow-up period (8).

The thoracic manifestations of paragangliomas include well-enhancing mediastinal masses, metastatic parenchymal nodules, lymphadenopathy from malignant paragangliomas, and pulmonary edema as a complication of epinephrine-producing paragangliomas. The much less common manifestations include a primary mass in the lung, heart, esophagus and/or trachea (5). Primary pulmonary paraganglioma was first reported by Heppleston in 1958 (9), and only 19 cases have been reported since then in the English literature (2). Three of these cases were malignancies with lymph nodes metastases (8). In the remaining cases, the lesions behaved in a benign manner. The patients are usually female and in their middle age. Most patients are free of symptoms and hypertension, and the tumors are often discovered incidentally on routine chest radiographs (8). Two distinct forms of primary pulmonary

paragangliomas have been reported (4, 8). The first and more common form consists of multiple minute tumors, in proximity to the pulmonary veins. The second and less common form consists of large solid tumors. The reported incidence of malignancy for a pulmonary paraganglioma is approximately 18%, which is lower than that for paraganglioma in other locations (20−50%). However, there are reports of metastases after a long time (10). Therefore, a lifetime of follow-up with careful, long-term observation by checking the urinary catecholamines and performing imaging studies is essential.

While pulmonary involvement by an adrenal or extraadrenal pheochromocytoma is uncommon, endobronchial involvement like that seen in our case is very rare. Only one case of hilar and subcarinal lymphadenopathies with endobronchial metastases from recurrent adrenal pheochromocytoma and another case of a 0.9 cm primary endobronchial paraganglioma of the lung have been reported (2, 5).

Functioning extra-adrenal paragangliomas represent more than 10% of all pheochromocytomas (4). Among all the reported cases of pulmonary paraganglioma, two cases developed hypertension and the patients died from a cardiac disorder that was believed to be associated with the functional tumors. There was a report of a case of functioning metastases of a nonfunctioning paraganglioma (10). They assumed that a hypothetical phenotypic heterogeneity of the primary tumor could explain the difference in the biological behavior of the primary tumor and its metastases (10).

The endobronchial paraganglioma in our patient manifested on CT as a hypervascular endobronchial mass, and this manifestation was similar to that of an endobronchial carcinoid or bronchogenic carcinoma. Therefore, we think paraganglioma should be considered when making the differential diagnosis of an enhancing endobronchial mass.

References

1. Aravot DJ, Banner NR, Cantor AM, Theodoropoulos S, Yacoub MH. Location, localization and surgical treatment of cardiac pheochromocytoma. *Am J Cardiol* 1992;69:283-285
2. Aubertine CL, Flieder DB. Primary paraganglioma of the lung. *Ann Diagn Pathol* 2004;8:237-241
3. Rosai J. *Ackerman's surgical pathology*, 8th ed. New York: Mosby, 1996:1015-1058
4. Saeki T, Akiba T, Joh K, Inoue K, Doi N, Kanai M, et al. An extremely large solitary primary paraganglioma of the lung: report of a case. *Surg Today* 1999;29:1195-1200
5. Sandur S, Dasgupta A, Shapiro JL, Arroliga AC, Mehta AC. Thoracic involvement with pheochromocytoma: a review. *Chest* 1999;115:511-521
6. Whalen RK, Althausen AF, Daniels GH. Extra-adrenal pheochromocytoma. *J Urol* 1992;147:1-10
7. Linnoila RI, Keiser HR, Steinberg SM, Lack EE. Histopathology of benign versus malignant sympathoadrenal paragangliomas: clinicopathologic study of 120 cases including unusual histologic features. *Hum Pathol* 1990;21:1168-1180
8. Lemonick DM, Pai PB, Hines GL. Malignant primary pulmonary paragagnlioma with hilar metastasis. *J Thorac Cardiovasc Surg* 1990;99:563-564
9. Hangartner JR, Loosemore TM, Burke M, Pepper JR. Malignant primary pulmonary paraganglioma. *Thorax* 1989;44:154-156
10. Fenandez-Llamazares J, Sabria-Leal M, Armengol-Carrasco M, Garsia-Bonafe M, Salca-Lacombe JA. Functioning metastases of a nonfunctioning paraganglioma. *J Surg Oncol* 1988;37:213-214