Primary Paraganglioma of the Lung: A Case Report

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ABSTRACT

Primary pulmonary paraganglioma, a rare type of tumour, is infrequently observed in clinical practice. The clinical data of a case of primary pulmonary paraganglioma admitted to the Department of Thoracic and Cardiac Surgery of the Hospital were analysed, and the diagnosis and treatment of primary pulmonary paraganglioma are discussed, combined with literature review. The clinical symptoms and imaging findings of primary pulmonary paraganglioma are atypical, and it is easy to misdiagnose. Pathological diagnosis is necessary for the final diagnosis.

Key Words: *Paraganglioma, lung, Diagnosis, Treatment*.

How to cite this article: Huang D, Wu R, Nie J. Primary Paraganglioma of the Lung: A Case Report. *JCPSP Case Rep* 2025; **3**:289-291.

INTRODUCTION

Paraganglioma is a rare neuroendocrine tumour of soft tissue that produces peptide kinins.[1] This tumour originates from neural crest cells. This tumour can be classified into functional and non-functional types based on its ability to secrete catecholamines. Depending on the site of occurrence, paragangliomas are categorised as either intra-adrenal or extra-adrenal.[2] Paragangliomas typically occur in individuals aged between 30 and 40 years, with the majority being benign. The rate of malignant transformation ranges from 2 to 19%. Primary clinical manifestations include local invasion and recurrence, while distant metastasis is uncommon.[3] Pulmonary paragangliomas are rare, and the literature on this condition is limited. Herein, a case of primary pulmonary paraganglioma admitted at the Department of Thoracic and Cardiac Surgery in August 2024 is reported.

CASE REPORT

A 57-year male patient was admitted to the hospital due to a persistent cough and haemoptysis lasting for over one month. The patient had a medical background of hypertension and no previous medical history of other ailments. After admission, the physical examination revealed no positive findings, and the laboratory tests yielded normal results. The contrast-enhanced CT of the chest revealed a well-defined, irregularly-shaped nodular soft tissue density shadow in the middle lobe of the right lung measuring approximately 1.9 cm × 2.6 cm.

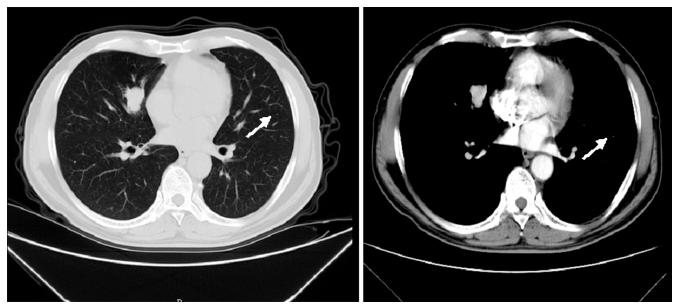

Figure 1: Contrast-enhanced CT of the chest revealed a mass in the right middle lung.

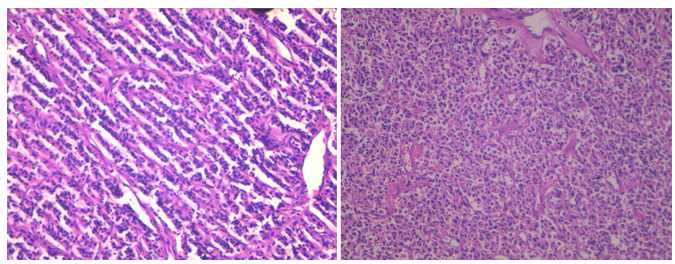

Figure 2: Postoperative pathological picture of the mass.

The density exhibited heterogeneity with a CT value of about 45HU. Following contrast enhancement, the arterial phase showed a CT value of about 113HU, while the venous phase demonstrated a CT value of approximately 81HU (Figure 1). Preoperative examinations, including abdominal ultrasound, cranial CT, and bone scan, showed no abnormality. The thoracoscopic resection was performed following the exclusion of any surgical contraindications. The postoperative pathology findings revealed the presence of a paraganglioma in the right middle lung (Figure 2). The tumour diameter measured approximately 1.5 cm. No evidence of nerve or vascular invasion was detected. There were no signs of involvement in the visceral pleura or bronchial margins. The total number of examined lymph nodes was 13, comprising 4 lymph nodes in groups 2-4, 2 lymph nodes in group 7, and 7 lymph nodes in groups 10-12, all of which showed a result of reactive hyperplasia. The immunohistochemical results suggest that the tumour cells were nega-

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Received: November 30, 2024; Revised: January 20, 2025; Accepted: February 06, 2025
DOI: https://doi.org/10.29271/jcpspcr.2025.289

tive for TTF-1, 1, Napsin A, CK7, and p40. The support cells show positive staining for S-100 protein. CD31 is negative while CD56, CgA, Syn, and Ki-67 are positive with a low proliferation index (<1%). The patient made a successful recovery and was discharged from the hospital.

DISCUSSION

The paraganglioma is a neuroendocrine tumour originating from neural crest cells and falls under the category of peptide hormone-producing tumours,[1] exhibiting a low incidence rate. The phenomenon of paragangliomatosis was initially documented by Aleza's and Peyron in 1908, while Pick introduced the term paraganglioma for extra-adrenal pheochromocytoma in 1912. The occurrence of paraganglioma is closely associated with the parasympathetic nervous system. It is commonly observed in anatomical regions, such as the neck, aortic body, vagus nerve, and retroperitoneal areas, where paragangliomas tend to accumulate. The occurrence of primary pulmonary paraganglioma is infrequent in clinical practice, constituting approximately 1% of reported cases of paraganglioma.[4] There is a paucity of literature on primary pulmonary paraganglioma, with the existing reports being limited to case studies and lacking epidemiological investigations.

The primary pulmonary paraganglioma is a non-functional hormone-producing tumour that does not secrete catecholamines.[5] The clinical symptoms are atypical. In the event of tumour invasion into the trachea and bronchus, it may result in obstructive pneumonia, and the primary clinical manifestations include cough and sputum production. The initial clinical manifestations of the patient in this case report were also cough with sputum, which was similar to literature reports. Meanwhile, the CT findings of pulmonary paraganglioma reported in the literature were also nonconforming to typical patterns, where the CT scan revealed circular nodules characterised by uniform density, well-defined margins, and mild homogeneous enhancement. The chest CT of this case showed a circular, uniform-density nodule. The differentiation of pulmonary paraganglioma from benign and malignant lung tumours using CT imaging poses challenges, necessitating a definitive pathological diagnosis for accurate determination.[6,7]

Most paragangliomas are benign tumours; however, the diagnosis of malignant paragangliomas is based on tumour invasion into surrounding tissues and organs, as well as recurrence and metastasis. The prevailing view among scholars is that paraganglioma exhibits a slow growth pattern and possesses the potential for malignancy or low-grade tumour formation. The majority of primary pulmonary paragangliomas are benign tumours, with malignant tumours being infrequent.[8]

The primary treatment of choice for primary pulmonary paraganglioma is surgical intervention, with thoracoscopic surgery being the recommended approach.[9,10] In this case, the clinical diagnosis of primary pulmonary paraganglioma was a space-occupying lesion found by chest CT, which was a suspected diagnosis of lung cancer. After excluding any surgical contraindications, thoracoscopic surgery was performed in order to obtain a definitive diagnosis of pulmonary paraganglioma. The literature has reported that paragangliomas exhibit a higher sensitivity to radiotherapy with reduced relative risk and complications.[11,12] The role of chemotherapy in paraganglioma lacks evidence-based medical support. The option of radiotherapy is available for patients with primary pulmonary paraganglioma who are unable to undergo surgical treatment.

The incidence of primary pulmonary paraganglioma is relatively rare, and the clinical symptoms as well as imaging manifestations present atypical features. Therefore, a definitive diagnosis can only be established through pathological examination. The limited number of literature reports hinders clinicians from gaining a comprehensive understanding of the disease, consequently leading to potential misdiagnosis. Although misdiagnosis is a common occurrence, the authors assert that it will not significantly impact patient treatment, as primary pulmonary paraganglioma patients can undergo surgical intervention prior to receiving a pathological diagnosis. However, the clinicians still need to acquire a comprehensive understanding of the disease in order to accurately differentiate it from other condition and minimise the occurrence of misdiagnosis.

PATIENT'S CONSENT:
Consent was obtained from the patient to publish the data concerning this case.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
DH: Designed the research, implemented the teaching, and prepared the manuscript.
DH, RW: Processed the imaging data and cases, designed the tests and questionnaires, and performed the statistical analysis.
JN: Revised the manuscript.
All authors approved the final version of the manuscript to be published.

REFERENCES

1. Bravo EL, Kalmadi SR, Gill I. Clinical utility of temozolomide in the treatment of malignant paraganglioma: A preliminary report. *Horm Metab Res* 2009; **41(9)**:703-6. doi: 10.1055/s-0029-1224135.
2. Joynt KE, Moslehi JJ, Baughman KL. Paragangliomas: Etiology, presentation, and management. *Cardiol Rev* 2009; **17(4)**:159-64. doi: 10.1097/CRD.0b013e3181a6de40.
3. Harari A, Inabnet WB 3rd. Malignant pheochromocytoma: A review. *Am J Surg* 2011; **201(5)**:700-8. doi: 10.1016/j.amjsurg.2010.04.012.

4. Kim MK, Park SH, Cho HD, Cho SJ, Kim A, Kim HK, *et al*. Fine needle aspiration cytology of primary pulmonary paraganglioma. A case report. *Acta Cytol* 2001; **45(3)**:459-64. doi: 10.1159/000327650.
5. Warren BC, Bond T, Hessler R, Boldt J. Primary paraganglioma of the lung in an asymptomatic patient. *Cureus* 2022; **14(6)**:e25562. doi: 10.7759/cureus.25562.
6. Ghigna MR, Dorfmuller P, Crutu A, Fadel E, de Montpreville VT. Bronchial paraganglioma with SDHB deficiency. *Endocr Pathol* 2016; **27(4)**:332-7. doi: 10.1007/s12022-016-9422-y.
7. Huang X, Liang QL, Jiang L, Liu QL, Ou WT, Li DH, *et al*. Primary pulmonary paraganglioma: A case report and review of literature. *Medicine* 2015; **94(31)**:e1271. doi: 10.1097/MD.0000000000001271.
8. Fiorentino G, Annunziata A, De Rosa N. Primary paraganglioma of the lung: A case report. *J Med Case Rep* 2015; **9**:166. doi: 10.1186/s13256-015-0639-z.
9. Zhang JJ, Liu T, Peng F. Primary paraganglioma of the lung: A case report and literature review. *J Int Med Res* 2012; **40(4)**:1617-26. doi: 10.1177/147323001204000442.
10. Yuksel C, Kocaman G, Yenigun BM, Ozakinci H, Sak SD, Enon S, *et al*. Primary pulmonary paraganglioma: Two cases. *Turk Gogus Kalp Damar Cerrahisi Derg* 2020; **28(2)**:394-8. doi: 10.5606/tgkdc.dergisi.2020.18844.
11. Elshaikh MA, Mahmoud-Ahmed AS, Kinney SE, Wood BG, Lee JH, Barnett GH, *et al*. Recurrent head-and-neck chemodectomas: A comparison of surgical and radio therapeutic results. *Int J Radiat Oncol Biol Phys* 2002; **52(4)**:953-6. doi: 10.1016/s0360-3016(01)02751-1.
12. Gilbo P, Morris CG, Amdur RJ, Werning JW, Dziegielewski PT, Kirwan J, *et al*. Radiotherapy for benign head and neck para-gangliomas: A 45-year experience. *Cancer* 2014; **120(23)**:3738-43. doi: 10.1002/cncr.28923.