

Primary paraganglioma of the lung: A case report

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Background Primary pulmonary paraganglioma is an uncommon neuroendocrine tumor of the lung, mostly discovered accidentally.

Case presentation We present the case of an 18-year-old female patient with multiple pulmonary nodules discovered accidentally during investigations of another presentation.

Conclusion This case report describes a low-grade, malignant primary pulmonary paraganglioma that presented specific characteristics upon computed tomography imaging. *Egypt J Broncho* 2014 8:64–65
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Case presentation

An 18-year-old female patient, with no special habits, presented with left neck swelling along with low-grade fever and generalized fatigue. Otherwise her clinical examination was unremarkable.

All laboratory investigations were normal and her tuberculin test was negative. Neck ultrasonography was performed before biopsy of the neck swelling to exclude any relation to nearby vascular structures. A right lobe thyroid nodule was indicated, and therefore a thyroid profile was performed, which was normal.

A routine chest roentgenogram was performed, which revealed multiple pulmonary nodules (Fig. 1). Computed tomography (CT) findings confirmed the presence of multiple nodules close to the pulmonary vasculature (Fig. 2).

Fig. 1



Chest radiograph shows multiple pulmonary nodules.

The patient had three lesions, the most accessible one being in the neck. A true cut biopsy was performed, which showed a scattered muscle and collagen fibers in a bloody background. CT-guided fine needle aspiration from the pulmonary nodule was attempted, which failed.

Thyroid isotope scanning was performed, revealing a cold nodule (Fig. 3). Hemithyroidectomy was carried out as it is known that cold nodules have greater risk of malignancy. The pathology showed goiter with no activity or malignancy.

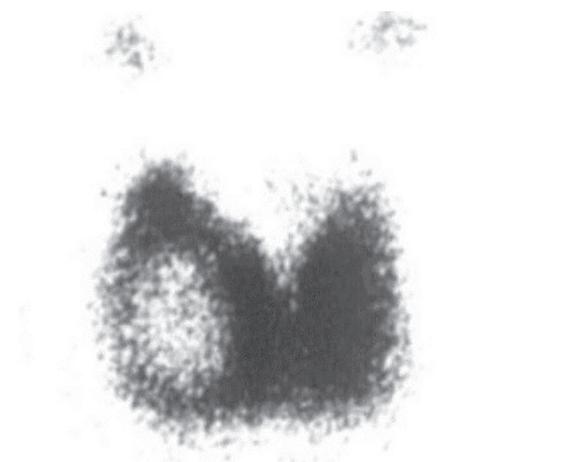
The patient was referred to a multidisciplinary thoracic tumor clinic, where an open lung biopsy was performed to resolve the case. Open lung biopsy and histopathological examination of the biopsied specimen revealed primary pulmonary paraganglioma (PPP). The oncologist started cyclophosphamide chemotherapy for follow-up.

Fig. 2



Computed tomography showed close relation of nodules to the pulmonary vasculature.

Fig. 3



Thyroid isotope scanning, revealing a right lobe nodule.

Comment

Paragangliomas are rare neuroendocrine tumors that arise from the extra-adrenal autonomic paraganglia, small organs consisting mainly of neuroendocrine cells that are derived from the embryonic neural crest and have the ability to secrete catecholamine [1]. These low-grade tumors frequently occur in the superior and inferior para-aortic region, but they have also been reported in a variety of uncommon locations, including the lung parenchyma [2].

The normal paraganglia consists of nests of chief cells (type 1) and sustentacular cells (type 2). Two types of paraganglioma have been reported: the first consists of multiple miliary tumors, as in our case; the latter consists of parenchymal or subpleural solid tumors [1]. According to the WHO classification, they are tumors belonging to the group of parasympathetic (nonchromaffin) branchiomeric paragangliomas, known as chemodectomas. Pulmonary paraganglia are identified proximal to blood vessels and nerves and are frequently located at the branching pulmonary artery [3].

PPP usually presents as asymptomatic nodules, coin lesions, or as a nonfunctioning solitary mass. Some of these patients were asymptomatic with a mass being found incidentally on routine chest roentgenogram. Symptoms are usually related to catecholamine excess or local expansion of the mass [4]. In a few cases, mild or transient elevation of blood pressure was observed, but etiology remains unclear [5].

Table 1 WHO classification of pulmonary neuroendocrine tumors

Origin	Subtype
Epithelial	Typical carcinoid
	Atypical carcinoid
	Small cell neuroendocrine carcinoma
Neural	Paraganglioma

Table 2 Immunohistochemical markers in neuroendocrine neoplasm

	Paragan- glioma	Typical carcinoid	Atypical carcinoid	Small cell neuroendocrine carcinoma
Chromogranin	+	+	+	+
Synaptophysin	+	+	+	+
Neuron-specific enolase	+	+	+	+
Calcitonin	-	+	+	+
Cytokeratin	-	+	+	+
Carcinoembryonic antigen	-	+	+	+

Differential diagnoses of this case include neuroendocrine tumors (Table 1) with immunohistochemical differentiation (Table 2).

Conclusion

This case report describes a low-grade, malignant PPP that presented specific characteristics upon CT imaging. This case is helpful in understanding the defining morphological features of PPP evident on CT imaging that may help in the differential diagnosis of lung tumors.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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