PRIMARY PULMONARY PARAGANGLIOMA- A RARE CASE REPORT

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Extra-adrenal paragangliomas are rare with head and neck being the most common sites.^[1] Primary paraganglioma of lung is even rarer.^[2] Only a few cases of primary pulmonary paraganglioma have been reported in literature and was first reported by A. G. Heppleston in 1958.^[3]

A 57-year-old male, non-smoker presented with fever and persistent cough. The results of his clinical laboratory examinations as well as his blood pressure were normal. Arterial blood gas analysis showed normal oxygen saturation.

PATHOLOGICAL DISCUSSION

A chest x-ray and a contrast enhanced computed tomography (CECT) were performed which revealed a rounded, well-circumscribed, high density solid mass with a diameter of 1 cm located in the right middle lobe. No enlarged lymph nodes were observed at the pulmonary hilum or mediastinum (Figure 1, 2).

A CT-guided core biopsy was done followed by video-assisted thoracoscopic surgery (VATS)- wedge excision biopsy of the lesion.

The resected specimen on grossing showed a grey-white firm lesion of 0.8 cm diameter which is situated 0.5 cm beneath the pleural surface. The resected margins appeared free.

Histopathological evaluation showed a neoplasm composed of cells arranged in nesting (Zellballen) pattern separated by thin fibrovascular septae. The tumour cells were round to polyhedral with moderate amount of eosinophilic granular cytoplasm. The nucleus appeared pleomorphic, mildly hyperchromatic with some showing inconspicuous nucleoli [Figure 3, 4]. With immunohistochemical staining the tumour cells showed strong diffuse positivity for neuron-specific enolase (NSE), chromogranin, and synaptophysin (Figure 5, 6, 7) and were negative for cytokeratin, TTF-1 & LCA. The spindle cells in the periphery of tumour nests (Sustentacular cells) showed strong positivity for S-100 (Figure 8).

Paragangliomas are tumours that arise from neural crest-derived endocrine cells or organs, known as paraganglia. These paraganglia can be divided into two groups-

- Paraganglia in the head and neck region (Including aorticopulmonary paraganglia), which are in close alignment with the parasympathetic nervous system,
- Paraganglia of the sympathoadrenal neuroendocrine system which are usually functional.^[4] The terminology of paragangliomas is based on the anatomic site of origin rather than the chromaffin or nonchromaffin status of the tumour. The histopathological features of the lesion is similar regardless of the location.

Paragangliomas have been described in virtually all organs but primary pulmonary tumours are extremely rare (<1%).^[5] Erickson et al., in 2001 described 28 cases of thoracic paragangliomas in a study conducted from 1978-1998, out of which 19 cases were pulmonary paragangliomas.^[1] They are slowly growing tumours, presenting as painless masses, and have a culture doubling time of approximately 42 years.^[6]

Metastatic paragangliomas are more frequent in the lung than a primary paraganglioma. Most patients with a pulmonary paraganglioma are usually free of symptoms and hypertension, and the tumours are often discovered incidentally as a space occupying lung lesion on routine chest X-rays.^[7] Some patients may present with cough or dyspnoea. The primary pulmonary paraganglioma can have two distinct type of presentation. The most common type is in the form of multiple small tumours in close proximity to the pulmonary vein. The second less common type is usually a solitary large solid nodule.^[8]

Pulmonary paragangliomas has to be differentiated from bronchial carcinomas and metastatic tumours. Imaging studies along with histopathology and immunohistochemical evaluation are needed for a definite diagnosis.

According to the current World Health Organization classification, malignancy of pheochromocytomas and paragangliomas is defined by the presence of metastases and not local invasion.^[9] The incidence of malignancy in a primary paraganglioma of lung (18%) is reported to be lower than that in paraganglioma of other location (20-50%). However, metastasis of lung paraganglioma to other sites has been reported after a long time.^[10] Currently there is no histopathological or biochemical markers to predict the malignant tendency of a pulmonary paraganglioma.^[9]

Complete resection is the treatment of choice for solitary primary pulmonary paraganglioma. Thoracoscopic surgery (VATS) is considered as the standard method which allows excellent exposure of the operating field of vision and causes less per-operative and postoperative complications than thoracotomy.^[11]

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Figure 1, 2. Chest X-Ray (1) and CECT (2) Showing a Rounded High Density Lesion in Right Lung

Figure 3, 4. Diffuse Nested (Zellballen) Pattern of Tumour Cells

Figure 5. IHC for Neuron Specific Enolase

Figure 6. Diffuse Strong Positivity for Synaptophysin

Figure 7. Diffuse Strong Positivity for Chromogranin

Figure 8. S-100 Positivity in Sustentacular Cells

DISCUSSION OF MANAGEMENT

The patient was treated with surgical excision alone. He is on regular follow-up and is symptomatically better.

FINAL DIAGNOSIS

Pulmonary Paraganglioma

The tumour in our case was diagnosed as pulmonary paraganglioma due to the diffuse nesting (Zellballen) pattern, cellular features and the characteristic immunohistochemical findings. A follow-up PET scan confirmed the absence of a paraganglioma elsewhere. Hence a final diagnosis of primary pulmonary paraganglioma was made. The biological behaviour of this rare neoplasm in lung is favourable when compared to any other site. However, no histopathological features can predict their malignant potential.

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