Intrapulmonary Paraganglioma

Sir,

Paragangliomas are rare tumors originating from paraganglionic tissue that extends from the skull base to the pelvic diaphragm. Although they can be asymptomatic, they can also cause symptoms by secreting catecholamines or with local expansion. They can also be the component of many hereditary diseases. These tumors are reported in various rare localisations, which include lung parenchyma.[1] The first primary pulmonary paraganglioma (PP) case was reported by Heppleston in 1958.[2] The number of cases diagnosed till date is less than 50 in the English literature.[3]

A 68-year female patient was admitted to chest diseases clinic with nocturnal dyspnea and cough that increased in the last month, but were present for the past 1-2 years. A 16×10 mm nodular lesion in the superior segment of the lower lobe of the right lung was detected in a computed tomography (CT) scan of the thorax of the patient, who was followed up with hypertension on her background. Because of suspicion of metastatic lesion, tru-cut biopsy was performed under CT-guidance from the lesion. Histopathological examination of the mass revealed tumoral lesion composed of large cells with abundant eosinophilic cytoplasm and uniform nuclei, showing mild hyperchromasia without atypical mitosis, embedded in the hyalinised fibrocollagenous stroma (Figure 1a, 1b). The tumor cells stained positive with synaptophysin, NSE, chromogranin, and S-100, and negative with Vimentin, EMA, CK5/6, and CD68 (Figures 1c, 1d). CK7 was focal positive.

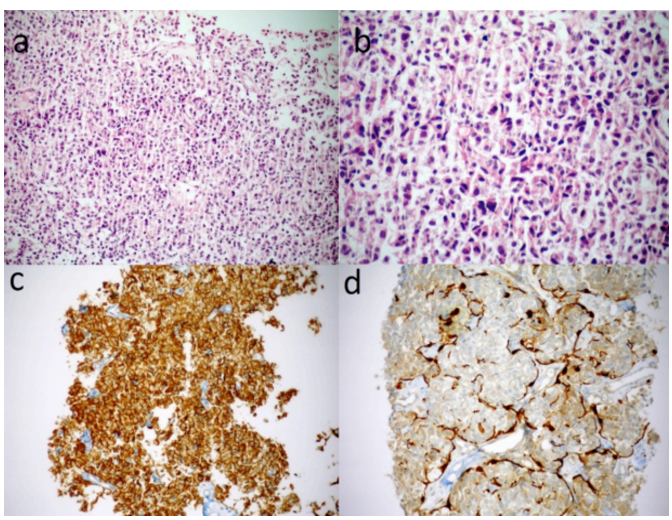

Figure 1: (a) Tumor composed of ovoid cells in nests, surrounded by thin capillary networks (H&E, ×200). (b) Zellballen pattern (H&E, ×400). (c) Synaptophysin expression (×200). (d) S-100 protein expression in sustentacular cells (x200).

Our case did not have clinical symptoms associated with catecholamines; and was endocrinologically "silent". Biochemically, the catecholamine levels were within normal limits. Primary PP was diagnosed with morphological, immunohistochemical, and clinical findings. Considering the age and general condition of the patient, surgery was not considered foreground. The patient has been monitored with annual follow-ups for five years. Radiological examinations revealed no growth of the lesion, and no lymph node/distant metastasis and extra-pulmonary involvement.

In PPs, women show preponderance, and patients are frequently asymptomatic. They are found to be solitary, peripheral, in the form of nonfunctional nodules, located close to pulmonary veins and nerves in the parenchyma of the lung.[3] Metastatic paraganglioma, carcinoid tumor, meningo-endothelial nodule, pulmonary carcinoma, pulmonary tuberculosis, metastatic lung cancer, and organised pneumonia should be considered in the histological differential diagnosis of primary PP; and it should also be noted that these diseases may accompany PP.[4]

The absence of classical carcinoid tumor architecture (trabecular, pseudoglandular, spindle), Zellballen pattern, and cytokeratin negativity help in differentiation from carcinoid tumor. In benign-malignant paraganglioma differentiation, presence of local invasion, lymph node metastasis, distant metastasis, extra-pulmonary involvement, as well as histomorphological and immunohistochemical findings are very important.[5] It has been reported that NSE immunoreactivity decreases in malignant paragangliomas. It is speculated that PPs have a better prognosis than mediastinal paragangliomas.

PATIENT'S CONSENT:
Informed consent was received from the participant.

CONFLICT OF INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
AK: Writing original draft, final approval, analysis and interpretation of data
FY: Literature review and designing.
HAK: Data collection, editing, and critical review.

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