Pulmonary epithelioid haemangioendothelioma mimicking lung cancer

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DESCRIPTION

A 63-year-old asymptomatic woman with abnormal chest shadow was referred to our hospital. The patient was a never smoker, and had a history of bronchial asthma and hypertension. Chest CT revealed a 4 cm diameter tumour in the right upper lobe, two nodules accompanied spicula and pleural indentation in the right S2 and S6, and right hilar and mediastinal lymphadenopathies (figure 1A,B). Each lesion possessed high fluorodeoxyglucose (FDG) accumulation according to FDG-positron emission tomography (PET)/CT (maximum standardised uptake value: 9.03) (figure 1C,D). Hence, we suspected locally advanced lung cancer. CT-guided lung biopsy revealed epithelioid cells with occasional vacuoles and hyalinised stroma. Tumour cells were positive for CD31 and CD34, and negative for cytokeratin AE1/AE3. These findings led to the diagnosis of a rare low-grade malignant vascular neoplasm called epithelioid haemangioendothelioma (EHE), which makes up <1% of all vascular tumours. The lesions were completely resected via right upper lobectomy and partial resection of right lower lobe with reactive changes in lymphadenopathies. No recurrence has been observed for more than 6 years.

Most patients with EHE (64%) had only one affected organ, with the liver as the most common organ (34%) followed by the bone (21%) and lungs (19%); EHE of the lungs is called pulmonary EHE (PEH).¹ The typical radiographical findings of PEH

Learning points

- ▶ Although typical radiographical findings of pulmonary epithelioid haemangioendothelioma (PEH) are reported to be multiple small nodules in both lungs, the images of the present case, including fluorodeoxyglucose-positron emission tomography/CT, revealed a mass and lymphadenopathies, which suggested lung cancer.
- ▶ The prognosis of completely resected PEH is good.

are multiple small nodules in both lungs;² therefore, radiographical images resembling primary lung cancer are rare. Generally, PEH development is slow, but it becomes rapid when there is high accumulation of FDG in the lesions.³ As a result, FDG-PET/CT results might be a useful indicator to determine whether PEH resection is needed.⁴ Forty per cent of patients survive less than 5 years;⁵ however, there have been cases of long-time survival and spontaneous regression. Symptomatic patients, pleural effusion, and lymph node and liver metastases are associated with poor prognosis.³⁻⁵ Nevertheless, the prognosis of completely resected PEH is good. Therefore, this case highlights the successful treatment of PEH mimicking locally advanced lung cancer.

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Figure 1 (A,B) Chest CT showing a 4 cm diameter tumour in the right upper lobe, two nodules accompanied spicula and pleural indentation in the right S2 and S6, and right hilar and mediastinal lymphadenopathies. (C,D) FDG-positron emission tomography/CT showing high accumulation of FDG in each lesion (maximum standardised uptake value: 9.03). FDG, fluorodeoxyglucose.

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