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CASE REPORT

Pulmonary Epithelioid Haemangioendothelioma

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ABSTRACT

This report is of a patient with pulmonary epithelioid haemangioendothelioma, a rare neoplasm. This report highlights the potential multicentric nature of this disease, which can be easily mistaken for widespread metastasis of unknown primary or granulomatous conditions such as sarcoidosis.

Key Words: *Hemangioendothelioma, epithelioid; Lung; Pulmonary medicine; Tomography, X-ray computed*

INTRODUCTION

Pulmonary epithelioid haemangioendothelioma is a rare neoplasm. Most patients are asymptomatic and the condition is usually detected by chest radiograph during a health examination.[1] This report is of a patient with a multicentric form of pulmonary epithelioid haemangioendothelioma. The imaging findings, histopathology, prognostic factors, and treatment options of pulmonary epithelioid haemangioendothelioma are discussed.

CASE REPORT

A 47-year-old woman with mild mental retardation presented in 2006 after an abnormal chest radiograph was taken during a pre-employment health examination (Figure 1). She had no symptoms except for an occasional cough. Sputum samples for cytology and acid fast bacilli had negative results. Bronchoscopy was unremarkable, but bronchial aspirate showed atypical cells.

Subsequent multidetector computed tomography (MDCT) scans and chest X-ray showed bilateral non-calcified non-cavitating pulmonary nodules. There was no pleural effusion (Figure 2). The differential diagnoses included pulmonary granulomatous disease or lung metastases.

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Diffuse thickening of the wall of the duodenum was seen (Figure 3). A small hypoenhancing area in the spleen was also noted (Figure 4). Osteolytic lesions were seen in the L2 vertebra and left iliac wing (Figure 5).

Serial chest radiographs showed no progression. Video-assisted thoracoscopy with wedge biopsy of the lingular lobe showed epithelioid haemangioendothelioma.

The patient received 2 cycles of carboplatin and etoposide with no radiological improvement. Chemotherapy was stopped and she was monitored clinically and radiologically. The patient has since been repeatedly

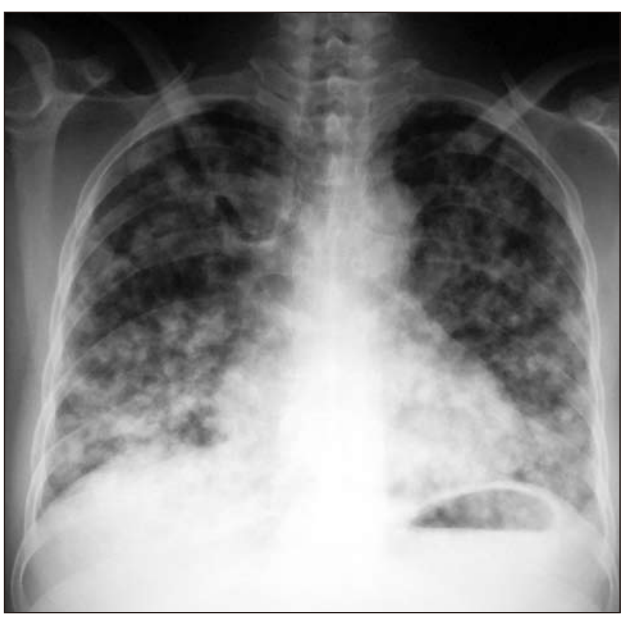

Figure 1. Chest radiograph showing bilateral small roundish nodules scattered in both lungs.

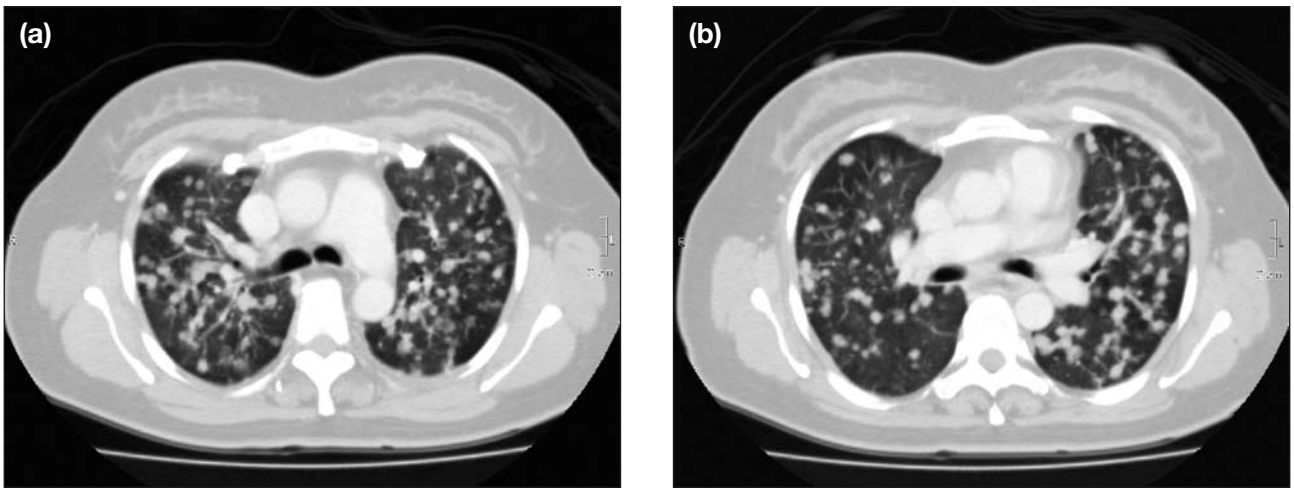

Figure 2. (a and b) Axial contrast-enhanced multidetector computed tomography showing bilateral non-calcified non-cavitating pulmonary soft tissue nodules. No pleural effusion or enlarged mediastinal lymphadenopathy was detected.

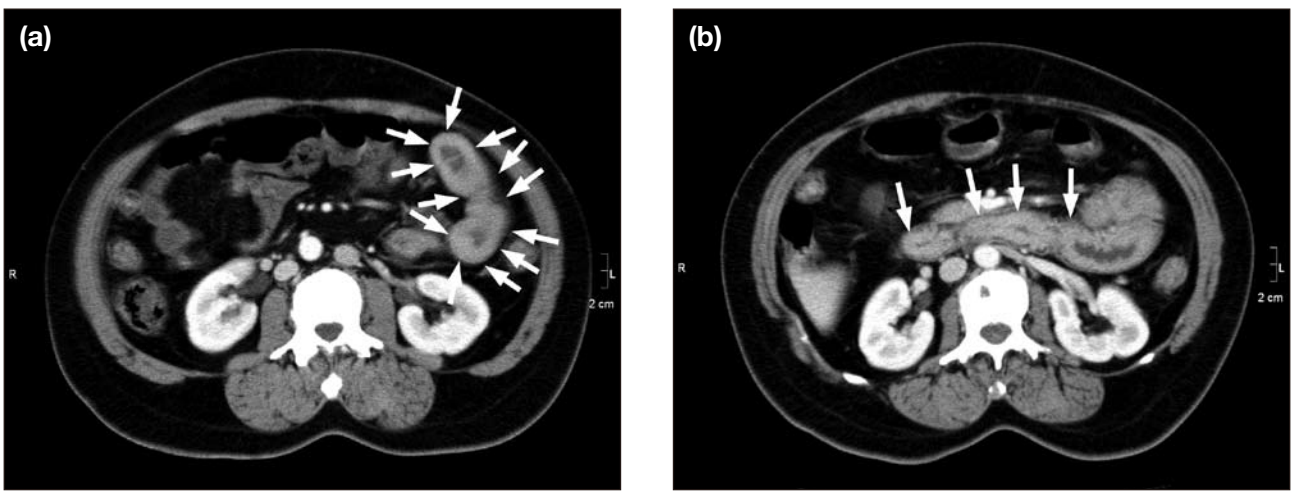

Figure 3. Axial contrast-enhanced multidetector computed tomography showing diffuse bowel wall thickening involving the (a) distal duodenum (arrows) and (b) proximal jejunum (arrows).

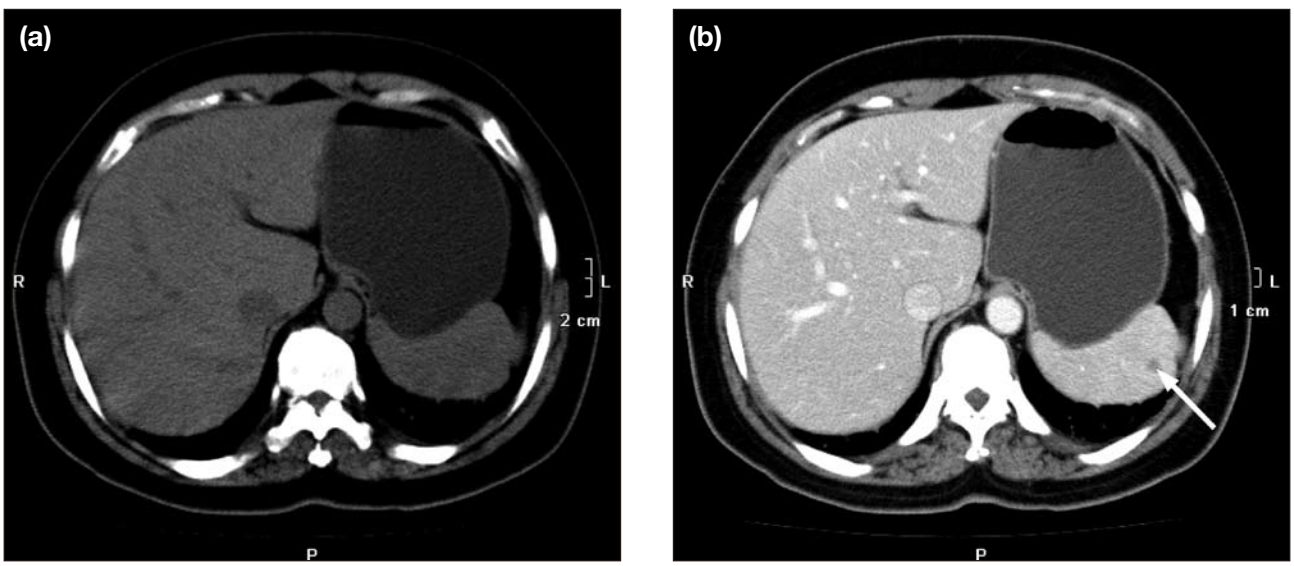

Figure 4. Axial (a) non-contrast–enhanced and (b) contrast-enhanced multidetector computed tomography showing a small hypoenhancing lesion in the spleen (arrow).

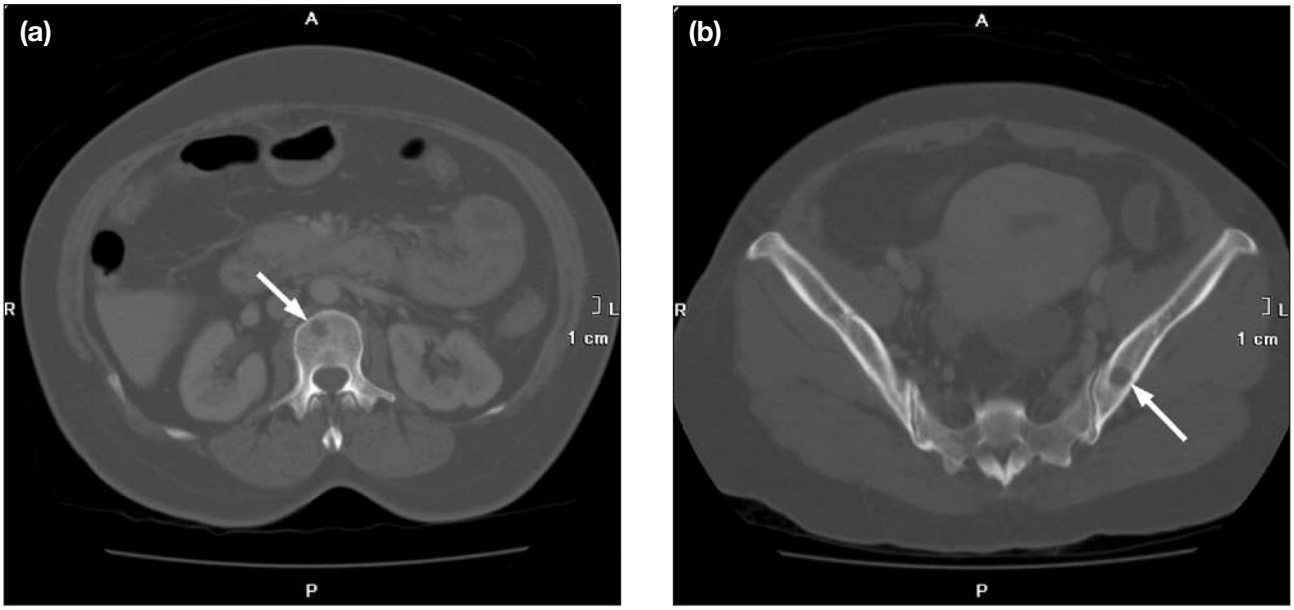

Figure 5. Axial contrast-enhanced multidetector computed tomography showing osteolytic lesions in the (a) L2 vertebra and (b) left iliac wing (arrow).

admitted to hospital for shortness of breath. Serial chest radiographs showed development of a small left pleural effusion. The patient died 1 year and 3 months after the initial diagnosis of pulmonary epithelioid haemangioendothelioma was made and 2 months after she was noted to have a left pleural effusion.

DISCUSSION

Epithelioid haemangioendothelioma is a rare tumour originating from the vascular endothelial cells. The pulmonary form is more common among younger women, and is usually asymptomatic. The pleural form is more common in older men, and symptoms include chest pain, dyspnoea, shortness of breath, cough, and weight loss.[2] The tumour can arise from many organ systems, including the liver, bone, bowel, and soft tissue, either simultaneously or sequentially.[3] This presentation is often mistaken for metastatic carcinoma, although minimal or no growth is seen on serial chest radiographs or MDCT scans.

This patient was a middle-aged woman with pulmonary epithelioid haemangioendothelioma. She was asymptomatic initially. Chest radiograph and MDCT showed multiple non-calcified non-cavitating pulmonary nodules scattered in both lungs with no pleural effusion. MDCT revealed bilateral lung nodules, and small bowel, splenic, and skeletal involvement. There is no documented relationship of this disease with mental retardation, so this was probably an incidental finding for this patient.

Most patients with pulmonary epithelioid haemangioendothelioma reported in the literature were diagnosed using open-lung or thoracoscopic biopsy specimens.[1,3] Diagnosis of other patients was based on the result of transbronchial lung biopsy or autopsy. The diagnosis for this patient was established using video-assisted thoracoscopy with wedge biopsy of the lingular lobe.

At gross examination, epithelioid haemangioendotheliomas of the lung are discrete well-circumscribed firm gray-white nodules that can resemble amyloid or cartilage. Histological analysis of these nodules demonstrates a hyalinised centre and a cellular advancing edge that extends into the alveolar spaces, bronchioles, and blood and lymph vessels. Some lesions can demonstrate moderate atypia, necrosis, and mitoses. Indeed, these tumours are considered to be low-grade sclerosing angiosarcomas.[4]

The clinical course varies. A poor prognosis has been associated with severe symptoms and with airway, vascular, or pleural involvement.[5,6] Kitaichi et al investigated 21 Asian patients with pulmonary epithelioid haemangioendothelioma and found that 2 of the 3 patients with pleural effusion died within 1 year, while 16 patients with no effusion were alive after more than 1 year, and 2 patients with fibrinous pleuritis and extrapleural proliferation of tumour cells died within 2 years compared with only 1 of 14 patients without these manifestations.[1] However, these authors also noted partial spontaneous regression in 3 asymptomatic patients 5, 13, and 15 years after diagnosis.[1]

Several chemotherapy protocols, including a variety of cytostatic agents (mitomycin C, 5-fluorouracil, cyclophosphamide, vincristine, tegafur, cisplatin, carboplatin, etoposide, and vinorelbine) have been tried for patients with pulmonary epithelioid haemangioendothelioma with varying results, ranging from complete remission to no beneficial effect.[1] Radiation is not effective for this disease.[4] Resection seems to be the treatment of choice whenever the lesion is solitary or the number of lesions is limited.[1]

REFERENCES

1. Kitaichi M, Nagai S, Nishimura K, et al. Pulmonary epithelioid haemangioendothelioma in 21 patients, including three with partial spontaneous regression. Eur Respir J. 1998;12:89-96.
2. Gurney JW, Winer-Muram HT, Stern EJ. Diagnostic imaging: chest. Salt Lake City: Amirsys Publisher: 2006. p 48-51.
3. Cronin P, Arenberg D. Pulmonary epithelioid hemangioendothelioma: an unusual case and a review of the literature. Chest. 2004;125: 789-92.
4. Giménez A, Franquet T, Prats R, Estrada P, Villalba J, Bagué S. Unusual primary lung tumors: a radiologic-pathologic overview. Radiographics. 2002;22:601-19.
5. Carter EJ, Bradburne RM, Jhung JW, Ettensohn DB. Alveolar haemorrhage with epithelioid haemangioendothelioma. Am Rev Respir Dis. 1990;142:700-1.
6. Case Records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 6-2000. Hemoptysis in a 20-year-old man with multiple pulmonary nodules. N Engl J Med. 2000; 342:572-8.