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CASE REPORT

Prolonged Survival with Multiple Pulmonary Metastases from Adenoid Cystic Carcinoma after Conservative Management

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ABSTRACT

This report is of a 38-year-old woman with adenoid cystic carcinoma and extensive lung metastases. The patient had prolonged survival of almost 2 decades after conservative treatment. The radiological features of metastatic adenoid cystic carcinoma have rarely been discussed. This report illustrates the finding of multiple lung lesions in an asymptomatic patient and discusses the differential diagnosis.

Key Words: *Carcinoma, adenoid cystic; Lung neoplasms; Neoplasm metastasis*

INTRODUCTION

Adenoid cystic carcinoma (ACC) is a rare slow-growing tumour arising mainly from the salivary glands. The lung is the commonest site of metastases, followed by bone, brain, and liver. Metastatic lesions of ACC may show radiological features indistinct from other pathologies. The correct diagnosis requires correlation of the clinical history, clinical progress, radiological features, and histological findings.

CASE REPORT

An asymptomatic 38-year-old woman with multiple lung lesions detected on chest radiograph was referred to the Princess Margaret Hospital (PMH), Hong Kong, in 2005 for a routine check-up (Figure 1). She had a history of left submandibular gland excision performed in China in 1990; the pathology was unknown. She had had a recurrence of the neck mass in 2000, with the excision performed in China. Multiple lung shadows were detected on chest radiograph and computed tomography (CT) of the thorax was performed in China in 2002. She did not receive any active treatment for the lung lesions.

At the time of presentation to PMH, she had no chest symptoms and no neck mass on examination. Blood test results were unremarkable. Sputum was negative for cytology and acid-fast bacilli. Chest radiograph showed multiple bilateral lung shadows of up to a few cm in size. CT of the thorax showed multiple lesions ranging in size from 5 mm to 4 cm distributed diffusely in both lungs (Figure 2a). Mild contrast enhancement and slightly ill-defined borders were noted. One of the lesions contained coarse calcification. No mediastinal lymphadenopathy or pleural effusion was detected. The lung lesions showed no significant interval enlargement

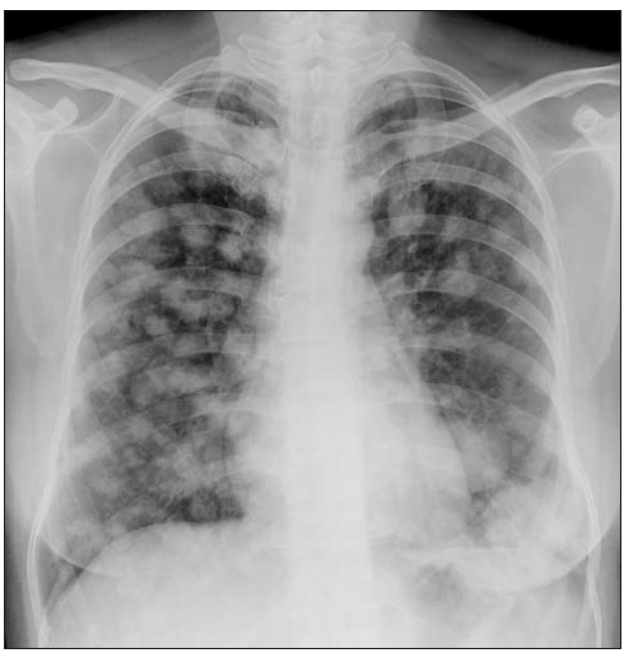

Figure 1. Chest radiograph at presentation showing multiple bilateral lung nodules.

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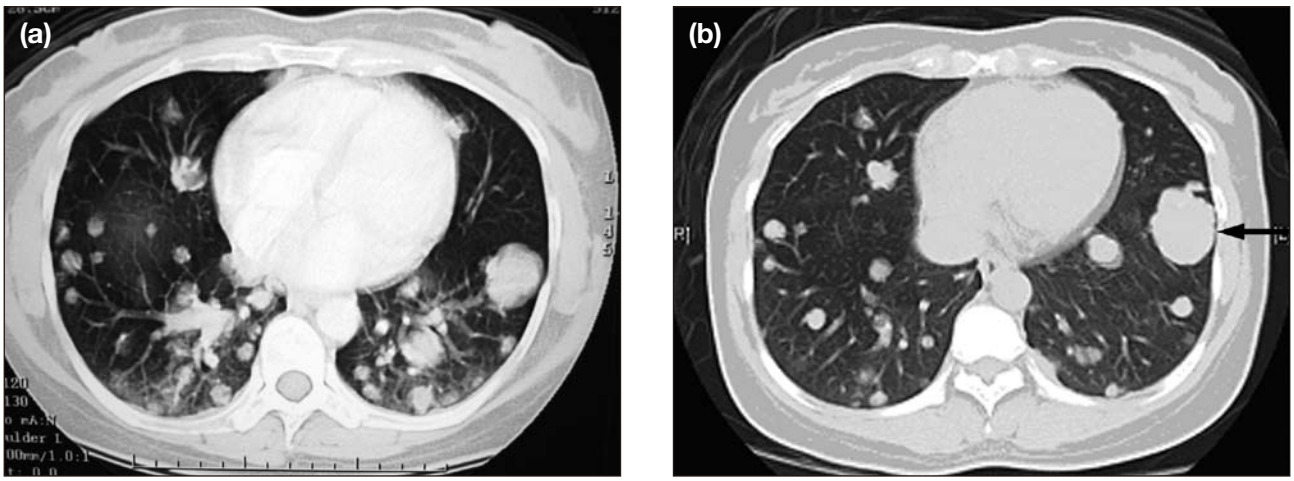

Figure 2. Computed tomography of the thorax performed in (a) 2006 showing multiple lung lesions of variable sizes; and (b) 2007 showing a mild increase in the size of one of the lung lesions (arrow). The apparent reduction in the number of lesions in 2007 and the difference in image quality are due to motion artifacts and imaging on a new computed tomography scanner in 2007.

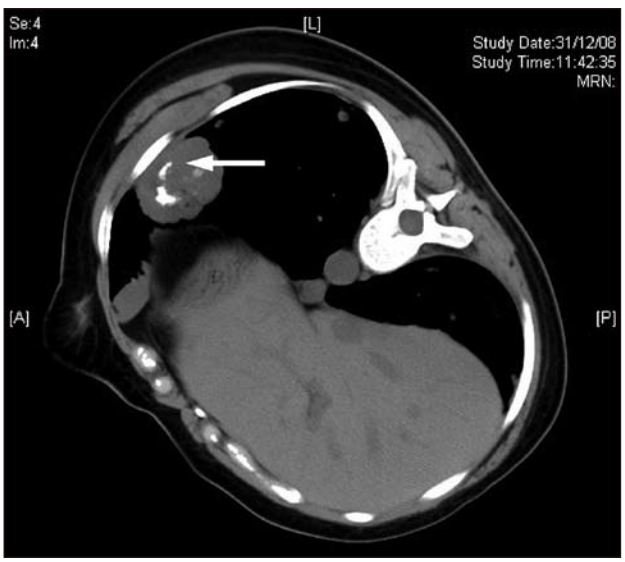

Figure 3. Computed tomography image taken during fine needle aspiration biopsy in 2008. Coarse calcification (arrow) is clearly demonstrated on the largest lung lesion, which showed no significant interval increase in size.

compared with the CT performed in China in 2002. Repeat CT of the thorax in 2007 showed only a mild increase in size of one of the lung lesions (Figure 2b).

CT-guided fine-needle aspiration (FNA) of the lung lesion was performed in 2008 (Figure 3). The largest lung lesion showed no significant interval enlargement compared with the CT of 2007. Histological analysis showed ACC of cribriform pattern with an abundant amount of intervening hyalin myxoid stroma without a solid or high-grade component.

In view of her past medical history of submandibular mass excision, the clinical picture was consistent with primary salivary gland ACC with multiple lung metastases. The lung lesions remained similar in size and number at the subsequent regular follow-up chest radiographs. The patient refused active treatment, so was managed conservatively and remained asymptomatic at follow-up in 2009. No palpable neck mass suggestive of a local recurrence has been documented.

DISCUSSION

ACC is a rare malignant tumour, which accounts for approximately 10% of all neoplasms of the salivary glands.[1] Approximately 60% of ACCs arise in the minor salivary gland.[1] ACC also affects other exocrine glands, including the lacrimal glands, ceruminous glands and, occasionally, excretory glands in the female genital tract.[2] There is a strong correlation between the site of origin and prognosis. The more favourable outcome for major salivary gland ACC compared with minor salivary gland ACC is attributed to the earlier discovery of the more accessible lesions.[1] It has been reported that ACC of the nasal cavity and paranasal sinuses has a worse prognosis than that of any other area of the head and neck region.[1]

ACC can be categorised into 3 histological growth patterns: cribriform, tubular, and solid. The solid growth pattern is associated with a worse prognosis, more advanced stage, and development of distant metastases.[1] No solid or high-grade component was seen in the lung metastasis of this patient.

Tumour stage is considered the most reliable indicator of overall outcome.[1] Major perineural invasion, positive margins, and solid histological features are associated

with treatment failure. Nodal metastases, major nerve invasion, solid histological features, and multiple presenting symptoms are associated with shorter survival.[2]

ACC tumour growth is slow with a protracted natural history, and prolonged survival with the disease is common.[2] Perineural extension and delayed onset of distant metastases are also common. Despite aggressive local therapy, approximately 60% of patients will develop recurrence.[1] Approximately 50% of recurrences are detectable within 2 years of surgery and radiotherapy.[1] It has been reported that distant metastases are seen in 43% of patients, of whom 70% have pulmonary metastases.[3] Large tumour size at presentation and locoregional treatment failure are the 2 factors most predictive of distant metastases.[1]

Late onset of pulmonary metastases from ACC is recognised, and the metastatic lesions can remain relatively stable for more than 10 years,[1] as in this patient. Ohara et al reported on a patient with ACC with multiple pulmonary metastases.[4] Ohara et al's patient also presented with an incidental finding of multiple lung nodules on chest radiograph.[4] These lesions were detected 14 years after excision of the primary ACC of the submandibular gland; the patient died 4 years after discovery of the lung metastases. Kömürcü et al reported a patient with pulmonary metastasis from ACC.[5] Kömürcü et al's patient had ACC of the external ear canal, and had had surgery done 20 years before he presented with multiple lung nodules on chest radiograph. This patient was treated with chemotherapy, but died of myocardial infarction 1 year later.

One of the lung lesions of this patient contained coarse calcification. The presence of calcification in lung ACC has been reported by Yoshikawa et al.[6] This finding is not commonly found in ACC, and whether or not the presence of calcification has any implication on the prognosis is unclear.

Surgical resection for isolated pulmonary metastasis may be worthwhile if the tumour histology is low grade and the patient has years of disease-free interval between treatment of the primary lesion and detection of the metastasis.[1] However, bony metastasis has a rapidly fatal course. Median survival times after the appearance of distant metastases among patients with isolated lung metastases and those with bone metastases with or without lung involvement were 54 and 21 months, respectively.[1] Prolonged follow-up is essential for early detection and management of delayed complications or metastases.

This patient presented with an incidental finding of multiple lung lesions on chest radiograph. Another patient with a similar history was reported by Ko et al.[7] Their patient had an incidental finding of multiple lung lesions 11 years after excision of a submandibular pleomorphic adenoma; the lung lesions were metastases of pleomorphic adenoma. Uterine leiomyoma is another example of a benign condition that can give rise to multiple lung metastases.[8] Slow-growing lung metastases from malignant tumours have also been reported in patients with thymoma[9] and renal cell carcinoma.[10] Uncommon causes of multiple chronic lung lesions include nodular pulmonary parenchymal amyloidosis[11] and epithelioid haemangioendothelioma (EH).[12]

EH was one of the differential diagnoses for this patient because she had the demographic, radiological, and clinical features commonly seen in association with EH. EH is a rare vascular tumour of low-grade to borderline malignancy.[12] This condition is more commonly seen in women than in men, with a mean age of onset of 35 years.[12] Patients are usually asymptomatic and the disease is often detected incidentally. EH may affect the lungs. Patients with EH may have a long survival, and survival of up to 20 years has been reported.[12] Radiological features of pulmonary EH include perivascular nodules up to 2 cm in size, with little or no growth on serial chest radiographs or CT. Ground glass opacities, interlobular septal thickening, hilar lymph node enlargement, and pleural effusions may also be present.[12]

ACC is a rare slow-growing tumour with a protracted natural history. The lung is the commonest site of metastasis. Lung metastases from ACC may present as an incidental finding in an asymptomatic patient. Diagnosing incidental lung lesions requires correlation with the clinical history, clinical progress, and radiological and histological findings.

REFERENCES

1. Bradley PJ. Adenoid cystic carcinoma of the head and neck: a review. Curr Opin Otolaringol Head Neck Surg. 2004;12:127-32.
2. Fordice J, Kershaw C, El-Naggar A, Goepfert H. Adenoid cystic carcinoma of the head and neck: predictors of morbidity and mortality. Arch Ololaryngol Head Neck Surg. 1999;125:149-52.
3. Yu T, Gao QH, Wong XY, et al. Malignant sublingual gland tumors: a retro-spective clinicopathologic study of 28 cases. Oncology. 2007;72:39-44.
4. Ohara G, Satoh H, Ohtsuka M. Late pulmonary metastasis of submandibular adenoid cystic carcinoma. Arch Oncol. 2007;15:48.

5. Kömürcü S, Özet A, Safali M et al. Delayed pulmonary metastasis of adenoid cystic carcinoma originated from external earway: case report. Turkish J Cancer. 2005;35:93-5.
6. Yoshikawa J, Takashima T, Miyata S, Kitagawa M. CT demonstration of calcification in an adenoid cystic carcinoma of the lung. AJR Am J Roentgenol. 1990;154:419.
7. Ko YS, Chui WH, Wang E, Chiu SW. Multiple pulmonary metastases from benign pleomorphic adenoma. Asian Cardiovasc Thorac Ann. 2008;16:62-4.
8. Londero AP, Perego P, Mangioni C, Lellé RJ, Londero F, Marchesoni D. Locally relapsed and metastatic uterine leiomyoma: a case report. J Med Case Reports. 2008;2:308.
9. Nomori H, Watanabe K, Ohtsuka T, et al. Pulmonary metastasis 12 years after resection of thymoma with microscopic capsule invasion. Jpn J Clin Oncol. 2004;34:630-3.
10. Tatsuta M, Shiozaki K, Masutani S, et al. Splenic and pulmonary metastases from renal cell carcinoma: report of a case. Surg Today. 2001;31:463-5.
11. Utz JP, Swensen SJ, Gertz MA. Pulmonary amyloidosis. The Mayo clinic experience from 1980 to 1993. Ann Intern Med. 1996;124: 407-13.
12. Cronin P, Arenberg D. Pulmonary epithelioid hemangioendothelioma: an unusual case and a review of the literature. Chest. 2004; 125:789-93.