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Pulmonary metastatic angiosarcoma from scalp with fatal complication: A case report

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ABSTRACT

INTRODUCTION: Angiosarcoma is a rare malignant neoplasm with poor prognosis. Angiosarcoma of the scalp is frequently recurs locally, and metastasizes early despite various treatments. The common sites of metastatic are lung, liver, and lymph nodes. Pulmonary metastasis with hemoptysis and pneumothorax is rare but threatening.

PRESENTATION OF CASE: A 77-year-old male had recurrent angiosarcoma of the scalp even with post operation radiotherapy. At the same time, recurrent pneumothorax was noted, thus he underwent wedge resection of the right upper lobe of the lung plus pleural biopsy. The final pathologic report of cystic lesions showed metastatic Angiosarcoma. He received intravenous paclitaxel and the lung lesions dramatically diminished subsequently.

DISCUSSION: Pulmonary metastasis from soft tissue sarcoma had fatal complications and poor prognosis. Metastases of AS to the lung have a well-described morphology on CT scan, but appear to be hypometabolic on PET scan and are easily misinterpreted as benign cysts.

CONCLUSION: Angiosarcoma is a rare but highly vascular invasive endothelial tumor that generally metastasizes to the lung. It could cause repeated hemoptysis pneumothorax and pleural effusion. Preoperative chest CT may be recommended routinely. Aggressive treatment resulted in not only symptoms control but also good prognosis.

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1. Introduction

Angiosarcoma (AS) most commonly arises from the skin and soft tissues. AS of the face and scalp in the elderly is a distinct subgroup of AS that tends to recur locally and to metastasize early despite various aggressive treatments, resulting in a poor prognosis. Complete surgical resection and postoperative radiation are recommended for the treatment of AS. The most predominant metastatic sites of AS is the lung and pleura. The bone and liver are secondary common sites [1]. Management of metastatic lesions of the lung is important because pulmonary metastasis of AS frequently induces severe complications. Here we report the successful management of pulmonary complications of scalp AS with paclitaxel administration. The present work has been reported in accordance with the SCARE criteria [2].

Abbreviation: AS, Angiosarcoma.

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2. Case presentation

A 77-year-old Taiwanese male heavy smoker had chronic obstructive pulmonary disease for 40 years. Itchy spots over his left frontoparietal scalp were noted 1 year before admission and seborrheic keratosis was initially suspected. He underwent topical treatment in the clinic. He presented at our hospital for a second opinion 3 months ago because of disease progression with ulceration and bleeding in response to touch. Cutaneous AS (Fig. 1) of the scalp was confirmed by biopsy. Wide excision with free radial forearm flap coverage was performed. After surgery, the patient underwent adjuvant radiotherapy (60 Gy in 30 fractions). Unfortunately, multiple protruding lesions developed on the contralateral side of the scalp 1 month after surgery, and local recurrence was suspected. The patient was admitted and underwent surgical treatment with wide excision plus a free anterolateral thigh split-thickness skin flap. The endotracheal tube was removed 2 days after surgery. The patient developed a sudden onset of hemoptysis and shortness of breath 3 days after surgery. Chest radiography revealed bilateral pneumothorax (Fig. 2A). The patient underwent bilateral tube thoracotomy. A computed tomography (CT) scan of the chest showed multiple cystic lesions with mild surrounding ground-



Fig. 1. Cutaneous angiosarcoma of scalp measuring 6 × 4.5 cm in size was confirmed by tissue biopsy.

glass opacities in both lungs and minimal right pleural effusion (Fig. 2B). Chronic obstructive pulmonary disease with secondary pneumothorax complicated with hemorrhage because of anticoagulant medication was suspected. One week later, after removal of the chest tube, the patient sustained a recurrent pneumothorax, and was sent immediately to our hospital where a chest radiograph revealed right side pneumothorax. A tube thoracotomy was performed. In this hospitalization, video-assisted thoracoscopic surgery with wedge resection of the right upper lobe of the lung plus pleural biopsy was performed. The results of pathological examination for cystic lesions were compatible with metastatic AS (Fig. 3). In addition, the cytology of the pleural fluid was positive for

malignant cells. Mechanical pleurodesis was performed to reduce the recurrent pneumothorax. The patient underwent intravenous paclitaxel administration at a fixed dose of 90 mg weekly over 2 weeks. The linear and nodular infiltration of both lungs dramatically diminished over sequential chest radiographs.

3. Discussion

AS is a very rare vasoformative malignancy arising from either blood or lymphatic vessels. Cutaneous lesions on the scalp or the face are the most common sites of origin. Head and neck AS tends to affect older Caucasian men. The most common sites of metastasis are lung, liver, and lymph nodes. The clinical manifestations of patients with pulmonary metastasis are variable, but the most common symptom is hemoptysis [3]. In the case presented here, the patient had hemoptysis together with hemopneumothorax and pleural infiltration. A small number of cases have been reported to present with thin-walled pulmonary cysts that developed and enlarged within a few months [4]. The role of positron emission tomography (PET)-CT in evaluation of distal metastasis of AS is not clear. Metastases of AS to the lung have a well-described morphology on CT scan, but appear to be hypometabolic on PET scan and are easily misinterpreted as benign cysts [5]. Usage of F-18 fluorodeoxyglucose PET/CT as an imaging tool for staging and restaging of cutaneous AS of the scalp has also been reported [6]. In the present case, there was no uptake by the metastatic lung lesion observed on PET-CT scan before surgery. As AS of the scalp is rapidly progressive, the potential of metastatic lesions should be considered, and a preoperative chest CT may be recommended.

Recurrent spontaneous hemopneumothorax associated with metastatic tumors is rare. Hemopneumothorax is thought to be associated with the rupture of cysts that are involved with a neoplasm. Tateishi et al. [7] proposed four pathophysiological mechanisms of the formation of cystic metastatic lung lesions: (1) excavation of a solid nodular lesion, (2) infiltration of tumor cells



Fig. 2. (A) Chest X-ray showing right side spontaneous pneumothorax. (B) Axial view of high-resolution computed tomography of the thorax demonstrating multiple cystic lesions with mild surrounding ground-glass opacities in both lungs.

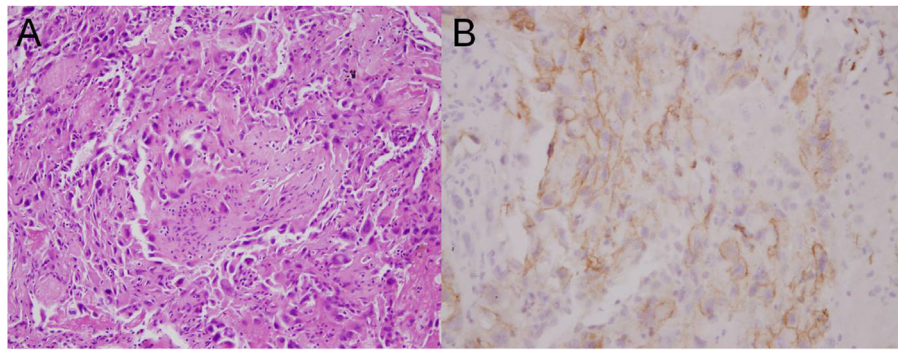


Fig. 3. (A) Median power view presents some pleomorphic tumor cells infiltrating in the lung parenchyma (H&E, 200 $\times$). (B) Immunohistochemical stains of CD34 reveal positive expression of tumor cells in lung tissues (CD34, 400 $\times$).

into preexisting bullous lung tissue, (3) infiltration of malignant cells into the walls of sacs and cysts, strengthening the ball-valve effect, and (4) the characteristic of AS to undergo cell proliferation to form blood-filled cystic spaces. According to these theories, tumor cells invading the pleural space may seed to form a pleural effusion. In our patient, minimal pleural effusion was noted on chest CT, a pathology consistent with CD31-positive AS. The cytology of pleural effusion is considered as inconclusive in diagnosis of metastases for AS patients. In the present case, examination of pleural effusion resulted in positive results for pleural metastasis. Cytology of pleural effusion maybe first considered for such patients due to lesser invasive procedure. However, it was difficult in such kind of cystic lesion. The radiologist hesitated in performing CT-guided biopsy due to high risk of bleeding. In the present case, examination of pleural effusion resulted in positive results for pleural metastasis. Cytology of pleural effusion maybe first considered for such patients due to less invasive procedure.

There is limited evidence upon which is the base treatment for AS of the head and neck. Complete surgical resection with free margins is suitable for local and locoregional disease. Reconstruction can be performed following excision even with sizable deficits. Because of insidious local infiltration, resection should be combined with per protocol radiation therapy (RT). Some reports have argued the benefits of RT [8]. However, our patient developed local recurrence within 2 months after surgery even with adjuvant RT, although the excisional margin was tumor-free in the first operation. The rapid relapse of this tumor may be due to tumor aggressiveness. The role of adjuvant chemotherapy in treatment of AS has not been assessed in any large randomized trials. The main drug groups used for treatment of AS are anthracyclines, ifosfamide, and taxanes [9]. Management of pulmonary metastases and their associated complications are discussed in the literature rarely. Successful treatment of pulmonary metastasis of AS with docetaxel has been reported [10]. In the present case, the pulmonary symptoms subsided after two courses of chemotherapy with paclitaxel, and the linear and nodular infiltration of both lungs diminished dramatically. The patient has been completely free of lung metastasis for 6 months to date.

4. Conclusion

In conclusion, AS is a rare but highly vascular invasive endothelial tumor that generally metastasizes to the lung. It can manifest as a rapidly enlarging pulmonary cyst with hemopneumothorax and pleural effusion. Preoperative evaluation should include a recent CT. Aggressive treatment resulted in not only symptoms control but also good prognosis.

Conflict of interest

The authors have no conflicts of interest to declare.

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Ethical approval

Does not apply.

Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Yung-Sheng Cheng: study concept and design.

Wen-Chiuan Tsai, Tim-Mo Chen: acquisition of data.

Yung-Sheng Cheng, Tsai-Wang Huang: drafting of the manuscript.

Yung-Sheng Cheng, Tsai-Wang Huang: critical revision of the manuscript for important intellectual content.

Guarantor

Tsai-Wang Huang.

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