



Metastatic Epithelioid Sarcoma ES with Pneumothorax, Hemoptysis and Thoracic Radiographic Bilateral Cavity Lesions in Young Male Patient – Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Epithelioid sarcoma is a rare soft tissue sarcoma in young adults (20-39 year olds) involving the upper extremities 60% of the time. The incidence of upper extremity epithelioid sarcoma was 0.1 cases per million per year [1]. This tumor is more common in males (1.8:1) and affects the young adult population [2]. It is a slow growing tumor with a high rate of recurrence and metastasis. In a long term study, 45% of patients with epithelioid sarcoma developed metastatic disease, with the lung (51%), lymph nodes (34%), scalp (22%) being the most common sites [3]. Close, long-term follow up is necessary because recurrence and metastases may occur long after definitive treatment.

This case report documents young male patient with chest pain and right sided pneumothorax, for which he underwent decortication. After 6 months he presented with recurrent hemoptysis and

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bilateral cavity lesions. Bronchoscopy was done and showed active bleeding coming from right lower lobe. Surgical resection done to right middle and lower lobe. The histopathology tissue report came as epithelioid sarcoma.

Keywords: Hemoptysis; pneumothorax; lung cavity; epithelioid sarcoma.

1. INTRODUCTION

1.1 Presenting Concerns

The subject of this report is a 20-year-old non married, non-drinking, non-smoking male, Bangladeshi student in Islamic University who had amputation for his left fifth digit eight year back in his country (malignant fibrous histiocytoma), presented to emergency room in September of 2014 with chest pain found to have right sided pneumothorax (Fig. 1). Chest tube was inserted in the right side (Fig. 2).

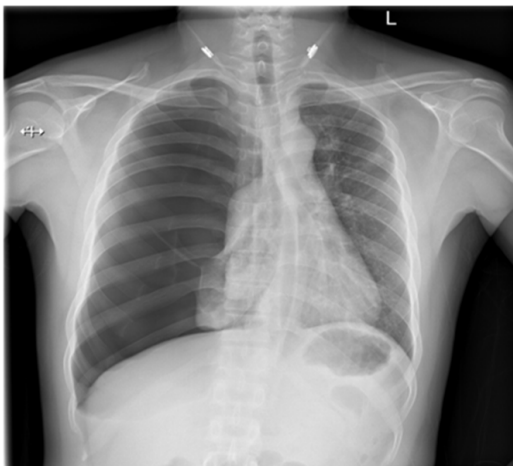


Fig. 1. Right sided pneumothorax

The patient had Video-assisted thoracoscopic surgery (VATS). There was complete collapse of the right lung with thick peel of fibrous tissue. He underwent decortication and developed bronchopleural fistula and started to have hemoptysis. Three days later he developed left sided pneumothorax for which the chest tube inserted. The patient was improved, and discharged from the hospital.

1.2 Clinical Findings

Patient was suffering from hemoptysis and asked medical advice at 17th Feb. 2015. On examination, he was fully conscious, oriented, pale, and uncomfortable at rest. His vital

signs were as follows: temperature, 37.1C; pulse, 102 beats/min; BP, 110/70 mm Hg; respiratory rate, 22 breaths/min; and oxygen saturation, 92% while on room air Chest: diminished Vesicular Breath Sound with bilateral basal crackles. X-ray done for the patient showed bilateral cavity lesions (Fig. 3). Computed tomography to chest showed Bilateral multifocal cavity lesion with large left hilar caseating lymph node. Consolidation collapse with airbronchogram in the superior and anterior segments of right lower lobe (Fig. 4a, 4b, 4c).



Fig. 2. Chest tube was inserted in the right side

The patient admitted in isolation room and started anti Tuberculosis therapy in the form of (rifampicin, isoniazid, pyrazinamide, ethambutol, moxifloxacin, and pyridoxine) and investigation done for tuberculosis. The laboratory result was remarkable for: low hemoglobin 6.6 g/dL and high erythrocyte sedimentation rate (ESR) 66 mm/h.

1.3 Diagnostic Focus and Assessment

The differential diagnosis for recurrent hemoptysis and bilateral cavity lesions are: Infections caused by commonly encountered gram-positive or gram-negative bacteria, Mycobacterium tuberculosis, fungal infection as Aspergillosis, autoimmune diseases as Wegener's granulomatosis, and lung cancer. Tuberculin skin test (TST) was negative. Sputa

for acid fast bacilli six times were negative. Serology for HIV and Hepatitis were negative. Anti-neutrophil cytoplasmic antibodies (ANCA) (P, C) were also negative. The patient underwent bronchoscopy. Active bleeding was coming from right lower lobe; bronchoalveolar lavage (BAL) for acid fast bacilli (AFB) and fungus two times were negative. The histopathology of transbronchial biopsy showed no evidence of TB but revealed a high-grade sarcoma with epithelioid differentiation. The tumor cells co-expressed vimentin and keratin and they were negative for TTF-1, CK5/6, S-100, SMA and CD31. The morphology and the immunohistochemical findings, along with the history of amputated finger for sarcoma, were consistent with metastatic epithelioid sarcoma (Fig. 5a, 5b, 5c, 5d).

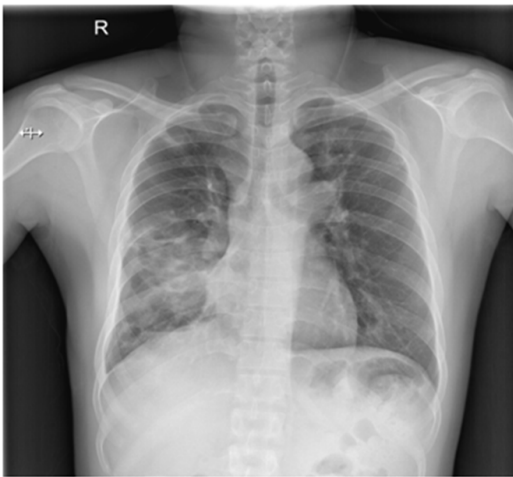


Fig. 3. Bilateral cavitory lesions

1.4 Therapeutic Focus and Assessment

The patient continue to have hemoptysis and started to deteriorate and his hemoglobin continue to drop requiring multiple transfusion .He was referred to thoracic surgery for resection the bleeding site. They decided to try embolization first and it was done, but he continued to bleed. Finally resection of the right middle and lower lobe was done, and was transferred to the intensive care unit and intubated for severe respiratory distress He continue to have hemoptysis, there was air leak at the of site right chest tube. He developed left sided pneumothorax.

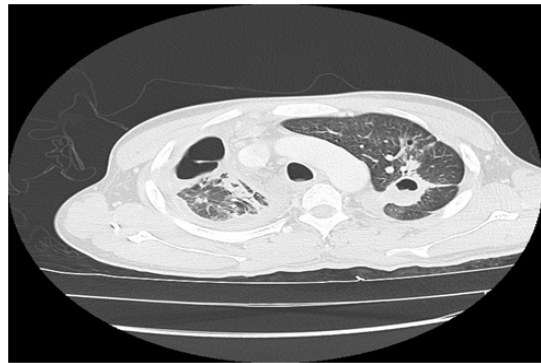


Fig. 4a. Bilateral multifocal cavity lesions

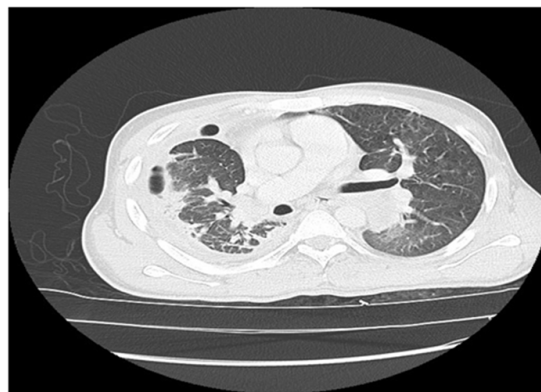


Fig. 4b. Consolidation collapse with airbronchogram in the superior and anterior segments of right lower lobe

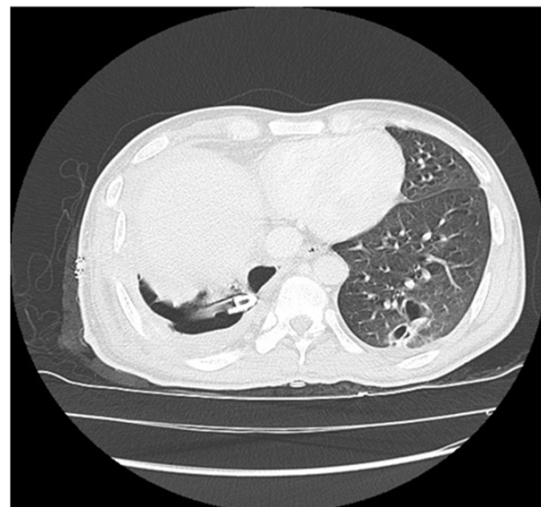


Fig. 4c. Multifocal cavity lesions in posterior segment of the left lower lobe

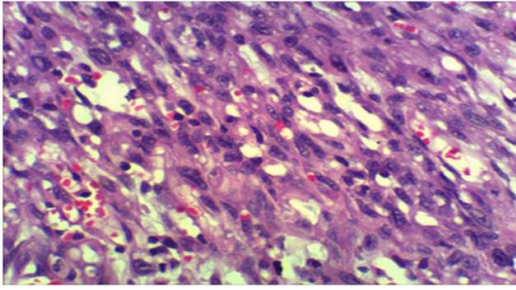


Fig. 5a. Section shows elongated spindled tumor cells with hyperchromatic pleomorphic nuclei (H&E stain, x400)

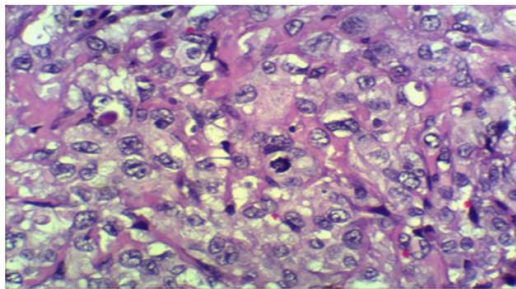


Fig. 5b. Section shows sheets of epithelioid cells with pale eosinophilic cytoplasm and pleomorphic vesicular nuclei (H&E stain, x400)

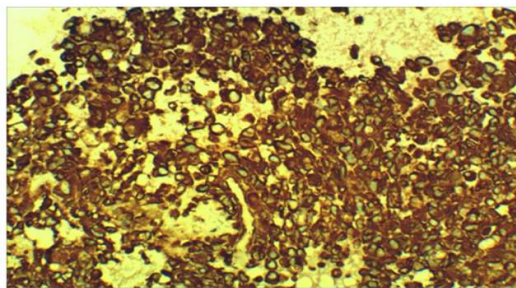


Fig. 5c. Section shows vimentin positivity in tumor cells (H&E stain, x200)

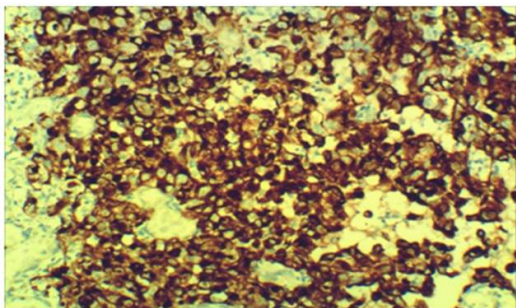


Fig. 5d. Section shows pankeratin positivity in tumor cells (H&E stain, x200)

The patient was referred to oncology service for further evaluation and management, but the general condition was bad they decided to continue conservative management only. He died on 7th of July 2015.

2. DISCUSSION

Our patient presented initially with pneumothorax. Pneumothorax occurs most frequently the result of tumor necrosis. It has been described in sarcoma's tumors, and in tumors with an aggressive and necrotic nature [4,5]. Necrosis of subpleural metastases is thought to produce a bronchopleural fistula that results in a pneumothorax.

In the general population only 10 of 1,143 cases with a spontaneous pneumothorax have been attributed to a malignancy [6]. A spontaneous pneumothorax in a patient with a sarcoma should raise the possibility of occult pulmonary metastases [5]. In such instances, CT may help detect occult pulmonary metastatic nodules.

Cavitation in primary lung cancer is not rare. Cavitation detected on plain chest radiographs has been reported in 2% to 16 % of primary lung cancers [5,6], and it is detected with computed tomography (CT) in 22% of primary lung cancers [7].

Metastatic disease from other primary sites may also cavitate, but this occurs less frequently than in primary lung cancers.

ES is slow-growing tumor with a predilection for the hands, fingers, and forearms [8].

Dissemination via the subdermal lymphatic vessels or the blood stream is a common feature of ES. The tumor can involve one or more lymph nodes when distant metastasis occurs [8]. The lungs are the most common site of distant organ metastasis [3,9].

The course of ES is often unpredictable, and it is common for patients to present with extensive disease, lymph node metastases, or distant metastases.

Local recurrence often occurs within 1 to 2 years of treatment, and these patients often proceed to develop distant metastases [8,9].

The median post metastatic survival was reported to be eight months [8]. Chase and Enzinger reported that the width of the tumor was

directly proportional to the rate of metastasis and this directly correlated with a lower 10-year survival [10].

Anatomic location of ES appears to play a role in prognostication. The overall survival and metastases-free survival are worse in lesions proximal to the elbow or knee [10,11]. Conversely, location on the distal extremities predicts a more favorable outcome [12]. In one study the 5-year and 10-year disease-free survival rates for all patients were 34% and 17%, respectively; for the 16 patients who received curative treatment. Tumor size >5 cm ($P < .0026$) at diagnosis and local recurrence ($P < .0008$) were significant predictors of survival [13].

Radical tumor excision is the primary treatment for patients with ES. Clean surgical margins must be obtained. Therapeutic lymph node dissection is indicated when lymph node metastases are present [8]. Halling et al. [9] suggest that, in the case of in-transit metastases, surgical resection of single or a small number of metastatic lesions may result in increased long-term survival. Radiation can also be considered for patients with marginal primary resection, local regional recurrence, or palliative treatment. Adjuvant chemotherapy is based on anthracyclines and ifosfamide appears to be indicated in the case of metastatic disease, but this is less clear in no metastatic ES [14].

Retrospective analysis of the clinicopathological features of 44 patients with ES. Among these patients, 26 were diagnosed histologically as having classic-type ES, whereas the remaining 18 had proximal-type ES. Thirty-three of the patients, all without distant metastases, underwent curative surgery, and the remaining 11 with distant metastases (M1) received palliative treatment. The overall survival rate at 5 years for the 44 patients was 45%. The proximal subtype was associated with unfavorable local recurrence-free survival and overall survival, although not to a statistically significant degree [15].

3. CONCLUSION

Epithelioid sarcoma is a rare, high-grade, soft tissue tumor that has a known propensity for local recurrence, regional lymph node involvement, and distant metastases. This malignancy can easily be mistaken for a benign process due to its often innocuous presentation. Misdiagnosis of this tumor can lead to delayed

and improper treatment, adversely affecting patient survival.

In this case report, the patient's recurrent hemoptysis and pneumothorax related to lung metastases. The patient most likely had ES in his amputated fifth digit. Early detection could have improved the outcome.

CONSENT

It is not applicable.

ETHICAL APPROVAL

As per international standard ethical approval has been taken from local ethical committee by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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