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CASE REPORT

Pulmonary Cystic Metastasis from Epithelioid Sarcoma Leading to Secondary Spontaneous Pneumothorax- A Case Report and Review of Literature

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Abstract

Well defined solid nodules are a recognized entity in pulmonary metastasis due to sarcoma, but cystic lung metastasis is seldom reported. A rare case of a 33 year old male presenting with left spontaneous pneumothorax secondary to underlying cystic metastasis from epithelioid sarcoma of left forearm is discussed. The Patient had undergone wide resection of epithelioid sarcoma in the left forearm 5 years back and had received chemotherapy for 2 years. His imaging study 2 years back did not reveal any evidence of metastasis. He presented to emergency department with acute onset left sided chest pain and breathlessness. A chest radiograph revealed spontaneous pneumothorax left side. A chest tube insertion and subsequent pleurodesis was done for the patient. He is on follow up and was restarted with chemotherapy. Oncologists need to be cognizant of this unusual sarcomatous metastasis and maintain high vigilance during the follow up of epithelioid sarcoma.

Keywords: Epithelioid sarcoma- cystic metastasis- pneumothorax

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Introduction

The incidence of soft tissue tumor is extremely rare. Epithelioid sarcoma is an uncommon malignancy and typically arises in the skin or subcutaneous portion of the distal limbs in young men [1]. It has an enormous predilection for local recurrence (77%) and secondary metastasis (30-58%) [2]. Pulmonary metastasis from sarcomas commonly occurs in the form of discrete solid nodules. Cystic pulmonary metastasis of sarcomas is an unusual manifestation [2]. Also, malignancy associated secondary spontaneous pneumothorax (MSSP) is a rare entity. We report a case of young male who had cystic lung metastasis of epithelioid sarcoma and presented with pneumothorax.

Case report

A 33 year old male presented to the emergency department with complaints of sudden onset chest pain and breathlessness. The patient had undergone wide resection of epithelioid sarcoma in the left forearm and subsequently received chemotherapy for 2 years. After that he was on follow up. During his last follow up, which was 2 years back a CT chest was done and was essentially normal. On arrival, he had tachypnea (respiratory rate was 30/min) and tachycardia (pulse rate was 108/min). His blood pressure was 108/70 mm Hg. His saturation was 90% at ambient air. His chest auscultation revealed absent breath sounds in the left hemithorax. A chest radiograph revealed pneumothorax of left side (Figure 1).

He underwent intercostal drainage(ICD) tube insertion. Post lung expansion a successful pleurodesis was done after 2 days. Consequently, a computerized tomography (CT) chest was done which showed variable sized nodules, few solid and few cystic with thin walls in anterior segment of bilateral upper lobes, apico-posterior segment of left upper lobe and lateral basal segment of left lower lobe (Figure 2a and 2b). Largest cyst was measuring 5 mm. He underwent bronchoscopy and no endobronchial lesion was identified. Furthermore, lavage samples were

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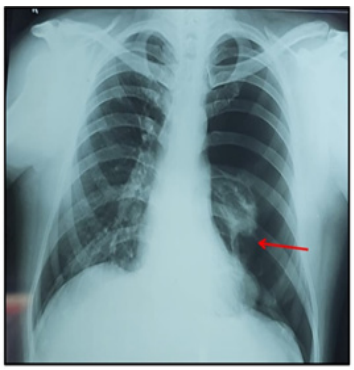

Figure 1. Chest Radiograph Depicting Pneumothorax Left Side (red arrow- collapsed lung).

negative for bacteria or fungus on stain and cultures. His broncho-alveolar lavage sample was also negative for cartridge based nucleic acid test for mycobacterium tuberculosis. A CT guided aspiration was performed from the nodules which depicted malignant cells (Figure 2c). He was restarted on chemotherapy. He is under follow up since 1 year and his lung lesions are static radiologically.

Discussion

Epithelioid sarcoma is an uncommon malignant tumor with a reported incidence of 0.6 – 1% among sarcomas. Sarcomas have a high tendency for metastasis which has been described in as high as 40-50% of patients. Metastasis commonly occurs to the lungs and local lymph nodes. Cystic lung metastasis from epithelioid sarcoma is a rare entity and has been described previously in case reports [3]. These cystic lesions are commonly not discernible on chest radiographs and warrant CT scan for diagnosis.The cysts are thin and smooth walled resembling a pneumatocele or emphysematous bullae. Therefore, it is imperative that the imaging differentials in such cases include cystic metastasis in addition to primary lung cancer or benign bullae [3]. When multiple nodules are present on imaging it is comparatively easier for radiologist to recognize metastasis, however the presence of few or cystic lesions should also demand heightened suspicion and encompass all differentials rather than considering it as a primary benign cystic pulmonary disease. A histopathological evaluation is the gold standard for confirmation of diagnosis [3].

MSSP is a rare occurrence and in a study of 2532 patients with secondary spontaneous pneumothorax, the incidence was meagre 0.045% [4]. Hoeg et al in their retrospective study on 126 patients of pneumothorax secondary to sarcomatous metastasis found osteogenic sarcoma as the commonest etiology seen in 31.4% and MSSP due to epithelioid sarcoma was seen in 1.3% of cases [3]. Two previous review of cystic metastasis from epithelioid sarcoma found 7 and 6 reported cases in literature and ,most had bilateral involvement with pneumothorax incidence in 85.7% of cases [2, 3]. The most common modality of treatment used was ICD placement (73.2%), followed by thoracic surgery (36.1%). Pleurodesis was done in 21.6% of cases [3]. Thoracic surgery is a feasible modality only if the lung metastasis is locally resectable. Otherwise chest tube placement and subsequent pleurodesis is performed for pneumothorax. We did a literature search and identified 10 previous reported cases (Table 1).

Rupture of subpleural cysts across the transpleural surface leads to pneumothorax. Three mechanisms have been implicated in the pathogenesis of these cystic lesions (i) excavation of nodular growth via expulsion of the necrotic debris inside the lesion. (ii) Permeation of tumor cells into the walls of already existing benign pulmonary bullae, (iii) Ball valve effect of the tumor due to enlargement of alveoli and small airways by incomplete bronchial occlusion [5]

A protocol for screening with CT scan should be followed by treating physicians to ensue timely identification of metastasis occurring simply as air filled lung cysts. These cysts are not obvious on radiography and are likely to be misread on imaging if not done in the correct clinical context. Primarily cystic metastasis have a smooth thin wall in comparison to solid lesions which undergo cavitation and have a thick irregular wall [2]. In the report described by Chan DP et al these metastasis were initially considered as benign and, differentials diagnosis of pneumatocele ,lymphocytic interstitial pneumonitis and lymphangioleiomyomatosis were deliberated upon [6]. In another case report these metastasis were the first presentation and primary tumor of the calf was identified much later [2]. In a review by Chan DP et al on 06 cases, 2 patients primarily had only cysts and 4 had cysts in conjunction with solid nodules. Commonly cysts measure from 10 to 40 mm in size and enlarge slowly [6].

In conclusion,though cystic metastasis from epithelioid sarcoma is a rare phenomenon it warrants a high

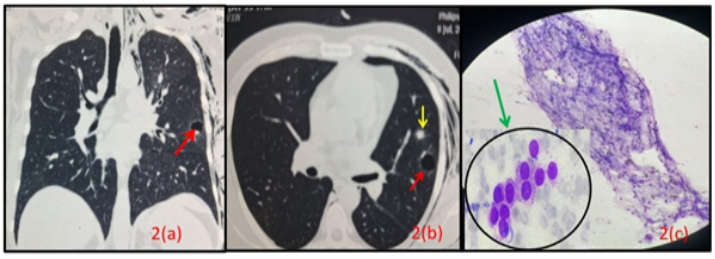

Figure 2. (a and b) CT Chest depicting cystic metastasis of epithelioid sarcoma (red arrow- cystic metastasis, yellow arrow- solid lesion). Fig 2(C) - CT guided fine needle aspirate Romanowsky stained shows polygonal cells with eccentric nuclei, prominent nucleoli and moderate eosinophilic cytoplasm against a background of RBCs(1000x) (green arrow)

Table 1. Characteristics and Outcomes of Previously Reported Cases of Cystic Pulmonary Metastasis from Epitheloid Sarcoma (M-male,F-female,N/A-not available)

Authors	Journal	Age/ Sex	Primary disease	Pneumothorax	Features of cyst	Treatment	Prognosis
Hasegawa et al. [7]	CHEST, 1999	20/F	Right Forearm	Bilateral	Cysts	Op	Alive at 7 years
Chan et al. [6]	Ann Thorac Surg, 2003	42/M	Thigh	Bilateral	Cysts	Op Chemo	Alive
Kikuchi et al [8]	Respirology 2006	39/F	Right hand	Bilateral	Cysts and nodules	Op chemotherapy (Carboplatin+paclitaxel and ifosfamide+doxorubicin)	Died 38 months after pneumothorax
Choi et al [5]	2008	35/M	Scalp	Bilateral	Cysts	N/A	N/A
Barnoud et al [9]	Ann Pathol, 2010	24/M	Right Foot	Bilateral	Cysts	Op chemotherapy (Adriamycin+ifosfamide)	Died 38 months after pneumothorax
Liu et al [10]	EJD, 2011	30/M	Right forefinger	Right	N/A	Untreated	Not given
Chong and Casserly [11]	Br J Hosp Med	31/M	Forearm	Bilateral	N/A	N/A	Dead
So- Yean Joen et al [1]	KJIM, 2016	42/M	Perianal area	Right	Cysts and nodules	Opchemotherapy (ifosfamide_doxorubicin+dacrbazine)	Died 3 months after pneumpthorax
Miyazawa T et al [2]	Ann Clin case reports, 2019	42/F	Left foot	left	cysts	Op Chemotherapy (doxorubicin and docetaxel+ gemcitabine and Ecteinascidin)	Died 21 months after Pneumothorax
Alesandrini A et al [12]	EC pulmonology, 2020	28/M	Right Thigh	Bilateral	Cysts	Chemotherapy (doxorubacin +phosphamide)	Died 10 months after diagnosis
Present study	-	33/M	Left forearm	left	Nodules+ cyst	Op+Chemotherapy (adriamycin+ifosfamide)	Alive at 1 year

level of awareness by the treating physician. Also, a multidisciplinary approach between the oncologist, radiologist and pulmonologist can help in timely identification of this cystic metastasis and prove beneficial in management of the patient.

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Statement of Transparency and Principals

- Author declares no conflict of interest
- Study was approved by Research Ethic Committee of author affiliated Institute.
- Study's data is available upon a reasonable request.
- All authors have contributed to implementation of this research.

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