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EC PULMONOLOGY AND RESPIRATORY MEDICINE

Case Report

Recurrent Bilateral Secondary Pneumothorax as a Presentation of an Epithelioid Sarcoma in a Young Patient

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Received: November 07, 2020; **Published:** November 26, 2020

Abstract

Epithelioid sarcoma reminds a rare but very aggressive malignant neoplasm of mesenchymal tissues. Mostly developed in young males affecting distal extremities with metastatic presentation in regional lymph nodes and lungs.

We present a 28-year-old male patient operated by uniportal video assisted thoracic surgery (U-VATS) with several episodes of recurrent pneumothorax, secondary to multiple and bilateral cystic metastases of an unknown epithelioid sarcoma on his thigh.

Keywords: *Epithelioid Sarcoma; Cystic Lung Metastasis; Secondary Pneumothorax; U-VATS*

Introduction

Epithelioid sarcomas (ES) are an unusual neoplasm which represents less than 1% of soft tissue sarcomas [1-3]. They are more common in males, with highest incidence between 17 and 60 years [4].

Depending on the location ES are divided in proximal where trunk, head and neck and thigh tissue are involved and distal, those involving the extremities [7].

More than 50% of patients develop metastatic lesions, especially in the lungs and regional lymph nodes. In resected patients, local recurrence is also higher, reaching 35% at 5 years of follow up [5,6]. Male gender, advanced age, and proximal location are considered adverse prognostic factors [2,4,8].

Secondary pneumothorax associated with cystic lung lesion was reported as a rare presentation of metastatic sarcomas [8-12].

Case Report

A 27-year-old male patient was admitted at the emergency room of our hospital with secondary left pneumothorax. He had a previous history of bilateral pneumothorax three months ago with apical thoracoscopic bullectomy performed in another center.

Thoracic CT scan was requested showing a left pneumothorax associated with multiple bilateral aerial cystic images surrounded by ground-glass opacities (Figure 1).

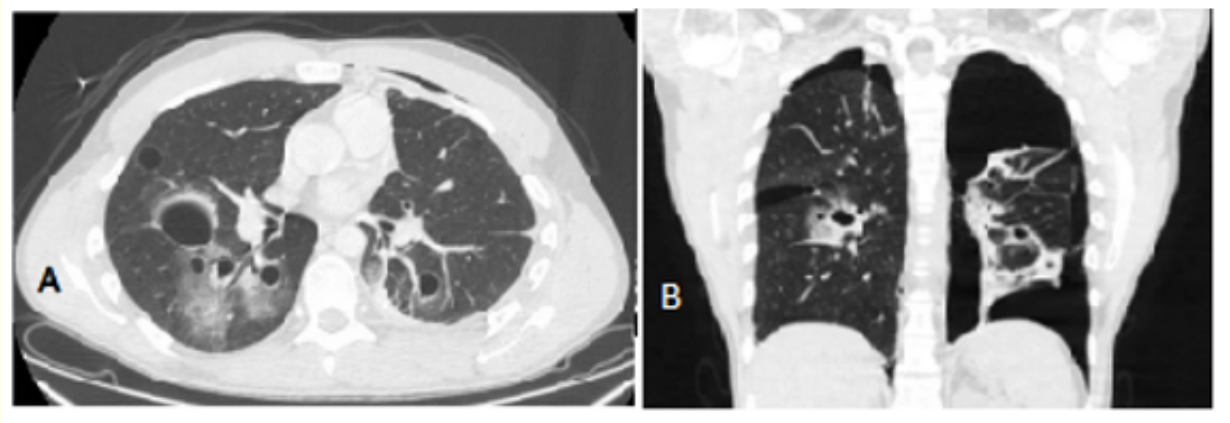

Figure 1: *Thoracic CT scan. A- Showing multiple bilateral aerial cystic images surrounded by ground-glass opacities. B- Coronal section: Left pneumothorax with bilateral cystic images.*

With these findings the patient underwent a left U-VATS, atypical lung segmentectomy and pleural biopsies was performed with talc pleurodesis to prevent pneumothorax recurrences.

The anatomopathological report metastasis from an epithelioid sarcoma. Immunohistochemistry analysis were positivity to cytokeratin, AE1-AE3, CD34, Muscle-specific actin, EMA and vimentin while negative to cytokeratin 7 and 20, TTF-1, CDX 2, S-100, CD117, P 63, CD 30, desmin, factor VIII and CD 31.

PET- CT showed an hypermetabolic image in the anterior aspect of his right thigh, measuring 40 x19 mm and two hypermetabolic right inguinal lymph nodes (Figure 2). With the confirmed diagnosis of "proximal epithelioid sarcoma" (PES)7 chemotherapy was prescribed with doxorubicin and phosphamide. After 2 cycles presented a new episode of right pneumothorax resolved by U-VATS and pleurodesis. Recovered from the last surgery completed 4 more cycles of chemotherapy uneventfully. The patient survived 10 months after the diagnosis dying after a massive hemoptysis.

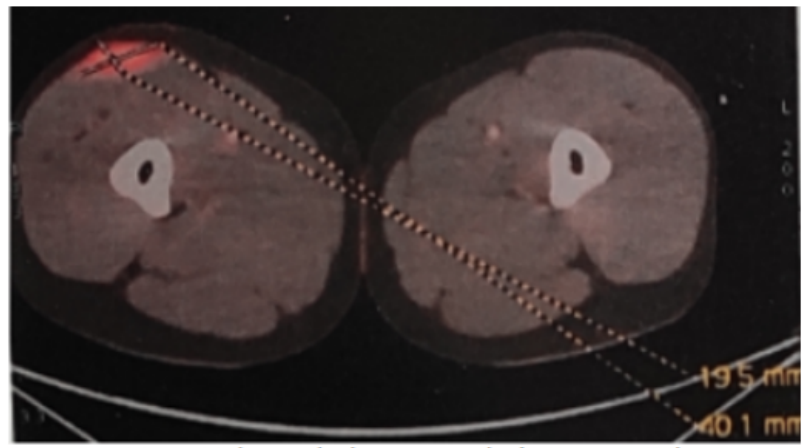

Figure 2: *PET-TC showed a hypermetabolic image at the anterior right thigh, measuring 40 X 19 mm.*

Discussion

Cystic lesions of lung parenchyma are rare being a challenging entity. Differential diagnosis are emphysematous bullae and pneumatocele nevertheless fungal or bacterial infections also take place. Minimally invasive surgery and in our case U-VATS with atypical pulmonary resection and pleurodesis is an accurate and effective method for the biopsy and palliative treatment [10,13].

The epithelioid sarcoma is an infrequent malignant tumor. According to the location it´s important to assess the presence of indurated lesions in subcutaneous space suspecting of those masses recently appeared and fast growing in a short period of time.

Pulmonary metastases forming small and solid tumors are very common for sarcomas however, cystic pulmonary lesions as metastases from ES are extremely unusual [8,12].

The prognoses of sarcomas after pneumothorax is generally poor, with 50% mortality between 4 and 5 months, and 75% with 1-year mortality; furthermore, less than 10% cases survived more than 2 years [14].

Multidisciplinary team (MDT) discussion between oncologist, surgeons, clinics and palliativist is mandatory due to the poor overall survival. In cases as we presented treatment must be palliative.

Conclusion

ES is a rare an unusual soft tissue neoplasm. Most affected population are young males. In cases of recurrent pneumothorax associated with multiple cystic lesions at the CT scan is mandatory an exhaustive pathologist asses of lung parenchyma to discard a metastatic tumor. The presence of indurated lesions in subcutaneous space recently appeared or with fast growth in a short period of time must be taken seriously. The prognoses of sarcomas after pneumothorax is really poor requiring MDT follow up.

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Volume 9 Issue 12 December 2020