

## Case Report

# Curious case of extraskeletal myxoid chondrosarcoma

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## ABSTRACT

Extraskeletal myxoid chondrosarcomas (EMC) are a rare entity of soft tissue tumors that have slow growth with metastatic potential. We discuss here a case of EMC presenting with right upper extremity pain and hemoptysis. Computed tomography scans chest showed diffuse metastatic numerous lung nodules bilaterally. Biopsy confirmed the diagnosis of the tumor. Chemotherapy was a bigger challenge for our patient due to sparse research and data in the literature about the disease.

**KEY WORDS:** Lung metastasis, soft tissue tumor, vorinostat

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## INTRODUCTION

EMC has been known for its well distinguished histological, immunohistochemistry and cytogenetic features. The tumor has a slow growth curve but it does carry a high risk of metastasis. The tumor is more common in males as compared to females with a ratio of 2:1, and it generally occurs in the 50-60 years of age group population.

## CASE REPORT

A 50-year-old Hispanic male presented with complaints of right upper extremity pain while lifting heavy objects at work, from several months. He also complained of cough and hemoptysis for the past several weeks. On examination, the patient was afebrile, hemodynamically stable, and maintaining oxygen saturation on room air. There was no significant past medical, travel, family, or surgical history. Routine investigations were noncontributory. Magnetic resonance imaging (MRI) of the right upper extremity demonstrated a soft tissue mass with central necrosis measuring 2.1 cm within the axilla. Core biopsy of the lesion confirmed the diagnosis of extraskeletal myxoid chondrosarcomas (EMC).

Initial computed tomography (CT) and positron emission tomography/CT scans demonstrated innumerable well circumscribed bilateral pulmonary nodules of varying sizes most consistent with metastases, without evidence of metastatic disease outside of the chest [Figures 1-3]. The patient underwent preoperative radiotherapy followed by gross surgical resection of the primary lesion in 2008. Thereafter, the treatment of the pulmonary metastases was initiated, and the patient took part in several research trials as the standard cytotoxic chemotherapeutic agents have historically shown no benefit in EMC. Several novel therapies including PF-02341066 (a CDK inhibitor), Brivanib, and PTC299 (antiangiogenic agent) were not effective as the pulmonary nodules continued to progress. The patient also continued to have several episodes of nonmassive hemoptysis which did not require intervention. For the last several years, the patient has been maintained on vorinostat 300 mg QOD with improvement in the symptoms and has relatively halted the growth of pulmonary nodules. The patient continues to be followed via serial CT scans of the chest 3 times a year.

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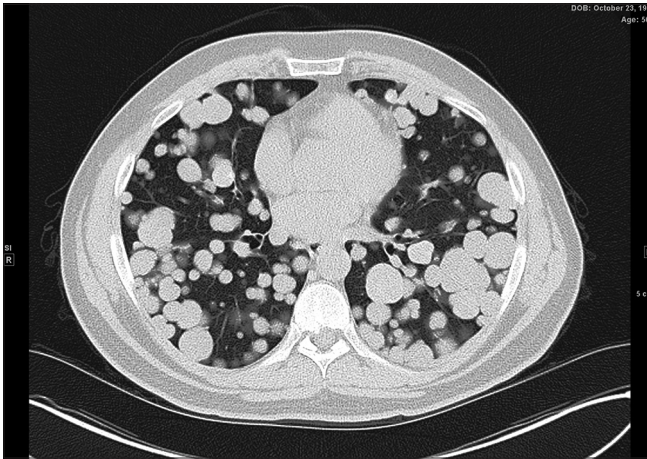
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**Figure 1:** Scout image demonstrates innumerable well circumscribed bilateral pulmonary nodules



**Figure 2:** Axial view of noncontrast enhanced computed tomography images of the chest demonstrate innumerable well-circumscribed bilateral pulmonary nodules



**Figure 3:** Coronal noncontrast enhanced computed tomography images of the chest demonstrate innumerable well-circumscribed bilateral pulmonary nodules, most numerous, and largest within the lower lobes typical of metastatic disease

## DISCUSSION

EMC tumor was first described by Stout and Verner in 1953.<sup>[1]</sup> Enzinger and Shiraki in 1972 first reported EMC as a rare soft tissue sarcoma with a separate histologically identity.<sup>[2]</sup> The revised version of the World Health Organization classification of tumors of soft tissue and bone in 2002 classified EMC under tumors of uncertain differentiation.<sup>[3]</sup> Soft tissue sarcomas are usually rare with mesenchymal origin, contributing only 1% of all adult malignancies; and EMC is accountable for even <2% of all soft tissue sarcomas.<sup>[4,5]</sup> Usually, EMC occurs in the lower extremities; but here, we report an unusual occurrence of EMC in axilla with extensive metastasis to the lungs. EMC is a very rare low-grade soft tissue sarcoma of chondroblastic origin. It comprises strands or cords of oval and spindle cells embedded in abundant myxoid stroma. Pulmonary metastases are common and cavitation of the nodules can occur, but EMC also metastasizes to the abdomen, bone, and peritoneum.<sup>[6]</sup> The differential diagnosis of EMC includes mucus liposarcomas and soft tissue myxomas. There is a characteristic chromosomal reciprocal translocation  $t(9;22)(q22;q12.2)$  resulting in fusion of EWSRI to NR4A3 (formerly known as CHN, TEC, or NOR 1) responsible for carcinogenesis in approximately 70% of cases, whereas another fusion gene, RBP56/TEC associated with translocation of  $t(9;17)(q22;q11.2)$  comprises 15% EMC cases.<sup>[7]</sup> Hence, it is believed that the remaining 15% of cases could be due to different translocation or gene anomalies still under research such as TCF12/TEC and TGF/TEC. The tumor is hypodense with no internal calcification on CT scan whereas on the MRI, it has low signal intensity on T1 and high signal intensity on T2-weighted scans.<sup>[8]</sup> In terms of treatment, wide local excision with or without radiation is the preferred modality of treatment due to its efficacy.<sup>[9]</sup> Radiotherapy alone has a more individualized effect, causing minimal reduction in tumor size; but larger comparative studies have failed to document reasonable outcomes with radiotherapy alone.<sup>[9]</sup> EMC has limited response to chemotherapy; single or combined chemotherapeutic agents did not show many significant data in treating EMC, hence warrants more research in identifying novel target therapies.<sup>[9]</sup> Vorinostat is a histone deacetylase inhibitor and inhibits the enzymatic activity of histone deacetylases that are overexpressed in some cancer cells, resulting in cell cycle arrest induction or apoptosis. Large tumor size, older patient age, tumor location in the proximal extremity, and metastasis play an important role in determining the prognosis and overall survival of the patient.<sup>[10]</sup> Tumor of >10 cm and metastatic disease are presumed to have a worse prognosis.<sup>[6]</sup> EMC is an indolent tumor and many patients despite metastatic disease survive for longer periods; the 5 years, 10 years, and 15 years overall survival rates were 82%, 65%, and 58%, respectively.<sup>[9]</sup>

## CONCLUSION

Despite the extensive disease burden within the lungs, patients can commonly survive for years with surprisingly little respiratory compromise, as in this case. The goal of presenting this case is to emphasize the importance of recognizing this rare location of the tumor and to provide appropriate therapy early in the hospital course. We would also like to stress on the role of vorinostat in curbing the growth of the tumor. However, further studies are warranted to find an appropriate chemotherapy regimen for better outcomes and palliation. A thorough workup should be done to rule out other metastatic foci, including imaging of chest given the frequency of lung metastasis. Although EMC is a rare tumor, it should be included in the differential for extra skeletal soft tissue masses. It is a slow-growing tumor, but simultaneously carries a high local recurrence rate and metastases, warranting long-term follow-up with quarterly CT chest surveillance. The most gratifying aspect is that the patient is still able to perform his activities of daily living.

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### Conflicts of interest

There are no conflicts of interest.

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