

Young Adult with Canon Ball Lung Metastasis and Unknown Primary: A Case of Primary Pulmonary Myxoid Sarcoma

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Abstract— Extra skeletal Myxoid Chondrosarcoma (EMC) is a rare soft tissue sarcoma, which primarily occurs deep in extremities, especially in the skeletal muscle or tendon. Unusual locations include tongue, retroperitoneum, spine, intracranium, testis, inguinal region, synovium, mammary gland, labium and pleura, however no case of has been described the aggressive involvement of lung with multiple canon ball metastatic atypical chondromyxoid neoplasm with unknown primary.

We hereby present a 38 year old Asian male patient initially presented for cough and occasional blood stained sputum with chest pain since few days, found to have multiple canon ball lung lesions which histopathological suggestive of atypical chondromyxoid sarcoma and primary source remained to be unknown.

Index Terms— chondrosarcoma, extra skeletal, myxoid.

I. INTRODUCTION

Extra skeletal myxoid chondrosarcoma (EMC) is a rare soft tissue sarcoma that was recognized as a distinct pathologic entity by Stout and Verner in 1953 [1], which primarily occurs deep in extremities, especially in the skeletal muscle or tendon. Unusual locations include tongue, retro peritoneum, spine, intracranium, testis, inguinal region, synovium, mammary gland, labium and pleura. However, it was not until 1972 that Enzinger and Shiraki [2] defined the clinic pathologic features of EMC, showing a relatively protracted clinical course and a better prognosis than that with conventional bone chondrosarcomas.

In 1992, Saleh et al. [3] reassessed the clinical features of EMC in a series of 10 patients and showed the "indolent but resilient" nature of this neoplasm; 7 of the patients died of tumor up to 17 years after the initial diagnosis.

Primary pulmonary malignancies are most commonly of epithelial origin. Tumours of other histologic differentiation have been described in association with the airway and bronchi; however primary tumours with cartilaginous or osteoid features originating in the lung are extremely rare and typically indicate metastatic disease. While cases of extra skeletal myxoid chondrosarcoma of the extremities are extremely infrequent, descriptions of this tumour histology arising from the lung is one of the rare and isolated to case

reports in the literature [4].

Primary lung cancers resembling EMC have been rarely reported as "primary pulmonary myxoid sarcoma (PPMS)". Primary lung tumours resembling EMC include PPMS [4,5], pulmonary angiomatoid fibrous histiocytoma (AFH) [6], and pulmonary microcystic fibromyxoma [7]. PPMS is a rare tumour of uncertain differentiation, characterized by EMC-like histology and EWSR1-CREB1 fusion [4,5], and this entity would encompass "low-grade pulmonary myxoid sarcoma" [8] and "primary pulmonary EMC" [9]. Lung myoepithelioma with extensive myxoid changes may be another differential diagnosis of this case. However, most cases of myoepithelioma were S-100 protein+ and/or SMA+ [4,9].

Here by we present case of young adult clinically manifested with short duration of cough with blood tinged sputum ,CT imaging shows multiple canon ball lesions of the lungs and histopathologically by CT guided biopsy of lung tissue consistent with features of Atypical Chondro myxoid neoplasm and there was no primary found to be associated.

II. CASE PRESENTATION

A 38 year old Indian male with no previous co morbidities presented with history of cough dry in nature since few weeks, occasionally with blood stained sputum increased during last 3 days, associated with right sided pleuritic chest pain. He gave the history of weight loss 5-6 kgs in span of 4 months. Clinical examination was unremarkable. During initial work up CXR showed multiple well defined nodules and masses in both lungs, largest noted in right upper lobe, Young Adult with Canon Ball Lung Metastasis and Unknown Primary: A Case of Primary Pulmonary Myxoid Sarcoma suggestive of metastasis; Superior mediastinum widening due to lymphadenopathy ; left sided minimal pleural effusion noted figure citation1. Further followed with CT Chest which was suggestive of a). Both lung shows diffuse metastasis of varying size giving a Cannon Ball appearance, largest noticed in right upper lobe measuring 3.5 x 3.2 cms; b). Multiple pleural metastatic lesions also noticed with left side pleural effusion; c). Multiple mediastinal and hilar lymph node masses+; d). Oesophagus displaced laterally by nodal mass figure citation 2.

CT Guided Biopsy of Lung tissue showed features of atypical chondromyxoid sarcoma, morphological features resembles those seen in extra skeletal myxoid chondrosarcoma. Patients vitals where stable and spo2 of 96 on room air, other systemic examination was in normal limits .To Rule out the primary cause pan CT was done and there

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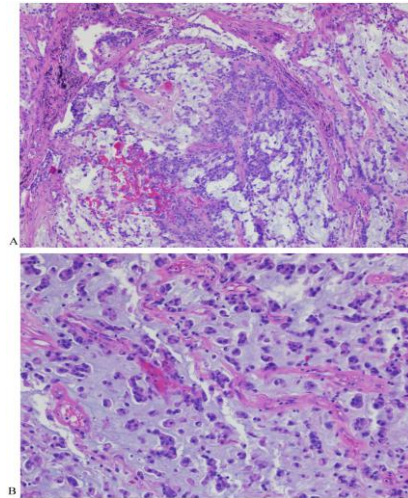
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was no foci present except right upper lobe lung mass. Upper and lower endoscopy was normal.

Images:



Low power magnification (10 ×): multilobulated mass with fibrous septa and myxoid background (figure A). High power magnification (20 ×): atypical epithelioid cells with round to oval nuclei, moderate nuclear atypia arranged in cords,



(Figure C)

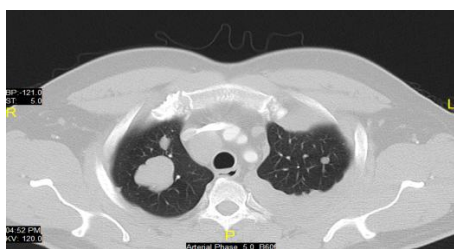


figure D

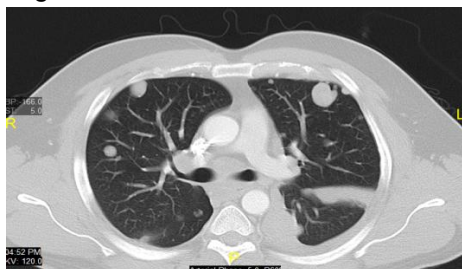


figure E

III. DISCUSSION:

Extra skeletal Myxoid Chondrosarcoma (EMC) is a rare soft tissue sarcoma, which primarily occurs deep in extremities, especially in the skeletal muscle or tendon, recognized as a

distinct pathologic entity by Stout and Verner in 19531. In 1992, Saleh et al. [3] reassessed the clinical features of EMC in a series of 10 patients and showed the "indolent but resilient" nature of this neoplasm.

Primary pulmonary malignancies are most commonly of epithelial origin. Tumours of other histologic differentiation have been described in association with the airway and bronchi; however primary tumours with cartilaginous or osteoid features originating in the lung are extremely rare and typically indicate metastatic disease. While cases of extra skeletal myxoid chondrosarcoma of the extremities are extremely infrequent, descriptions of this tumour histology arising from the lung is one of the rare and isolated to case reports in the literature. The histogenesis of EMC is still a subject of controversy. However, chondroblastic differentiation had been supported by ultrastructural [10-15] and histochemical [16-18] studies. And also cytogenetic and molecular analyses have shown that EMC is a distinct entity with the characteristic translocation t(9;22) involving the EWS gene (22q12) and the TECgene (9q22) in a majority of the cases. The classic histologic appearance with multiple myxoid lobules, separated by fibrous septa, and tumour cells arranged in cord or strands in a netlike or linear interconnected pattern like a "string of pearl" [19]. Immuno-histochemical, most EMC are positive for vimentin and focally positive for S100, negative for cytokeratin (except EMA) and GFAP.

Primary chondrosarcomas of the lung are extremely rare with only isolated case reports found in the literature. The largest series reported by Kalhor et al. highlights four cases-two of myxoid character and two hyaline type [20]. In 2012, two case reports were published describing one EMC [21] in a 51-year old female with severe anaemia and primary pulmonary myxoid sarcoma with EWSR1 CREB1 fusion, resembling EMC [3]. In November 2015 there was another rare case reported of 69 year old male with left lung upper lobe mass, which was finally diagnosed histopathological as Extra Skeletal Pulmonary Myxoid Chondrosarcoma [22].

With the present case given its rarity, atypical aggressive feature of this tumour involving both the lung where the primary source was not identified and manifesting in very short duration of time. It's difficult to state regarding the treatment and further prognosis of the described patient. The aggressive activity of chondrosarcoma depends on the tumour histology and is widely variable. EMC are generally low grade chondrosarcoma, but recent studies have demonstrated high potential for metastasis and local recurrence suggesting a more intermediate malignancy of this tumour [23].

Chondrosarcomas are notoriously resistant to conventional chemotherapy and radiation therapy, and resection remains the main stay of treatment. But in our case there are multiple lesions involving both lungs which rules out the resection mode of treatment. In advanced or metastatic tumours, recent studies with novel chemotherapy regimens have not demonstrated increased survival [24]. Further efforts are needed in this area to define a more effective strategy. The overall prognosis of ECM is difficult to estimate given the rarity of the tumour. Considering the rarity of this case with multiple lesions and the poor response of chondrosarcomas to

chemotherapy and radiation, no further therapy is planned in our patient.

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