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Case Report

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A Rare Case of Pulmonary Carcinosarcoma with Unusual Presentation: A Diagnostic Dilemma

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ABSTRACT

Pulmonary carcinosarcoma is a rare malignancy of the lung and has poor prognosis than non-small cell lung cancer. Effective treatment has not been developed for non-resect able advanced stage. We report a case in a 51-year-old female who presented with a right sided massive pleural effusion. Pleural biopsy tissue histology and immunohistochemistry revealed carcinosarcoma.

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Introduction

Pulmonary carcinosarcoma is rare malignant subtype of non-small lung cancer (NSCLC) of the lung usually seen in less than 0.3% of all lung cancer [1]. Carcinosarcoma is defined by presence of sarcomatous elements, epithelial elements and poorly differentiated carcinoma [2]. Most common involved organs in carcinosarcoma are uterus, gastrointestinal tract (GIT), skin, lungs, and hypopharynx [3]. It was first described in 1908, there are only few case reports in literature suggesting its predilection towards male gender (fifth decade of life), smokers and asbestos exposure [3, 4]. Surgery is the treatment of choice with median survival time of 9 to 12 months after curative lung resection. However, medical treatment does not play any beneficial role.

In the most recent version of the World Health Organization (WHO) classification, carcinosarcoma is included in the category of sarcomatous neoplasms with a poorer prognosis than non-small cell lung carcinoma. defined by the presence of (squamous or adenocarcinoma) combined with sarcomatous elements such as rhabdomyosarcoma, osteosarcoma, or chondrosarcoma

Case Presentation

A 51-year-old smoker female presented in emergency department with the complaints of difficulty in breathing, dry cough and mild fever for 2 months duration. The patient denied of any past medical history, no history of malignancy in family. Her baseline blood investigations were unremarkable. On examination she was dyspnoeic and oxygen saturation was 91% on room air. Her chest X-ray showed right sided pleural effusion with right sided chest drain inside (figure 1), which was inserted from outside hospital in view of massive effusion. Her right sided pleural fluid was haemorrhagic with low adenosine d aminase (ADA), exudative with cytology negative for malignant cells. Computed tomography (CT) of thorax was done which showed a massive right sided pleural effusion with collapse of right lung. Her screening fibreoptic bronchoscopy (FOB) was negative for any endobronchial growth (figure 2). For further evaluation thoracoscopy was done on day-4 of admission and biopsy taken from costal pleura (figure 3) and sent for histopathological examination (HPE). CT abdomen done to rule out abdominal pathology, which showed suspicious mass in liver and left adrenal gland suggesting metastasis. Contrast enhanced magnetic resonance (CEMRI) imaging of brain was done which was normal. Meanwhile her pleural biopsy report was obtained on day 14 which showed spindle cell neoplasm with sarcomatous elements. For further confirmation immunohistochemistry (IHC) was done which was positive for TTF-1, cytokeratin (AE1AE3) and p40 in epithelial component suggesting carcinosarcoma. Patient received chemotherapy with gemcitabine and cisplatin for 3 cycles. Unfortunately, patient died after completion of her third cycle of chemotherapy due to severe sepsis, distant metastasis (brain) and acute respiratory distress (ARDS).

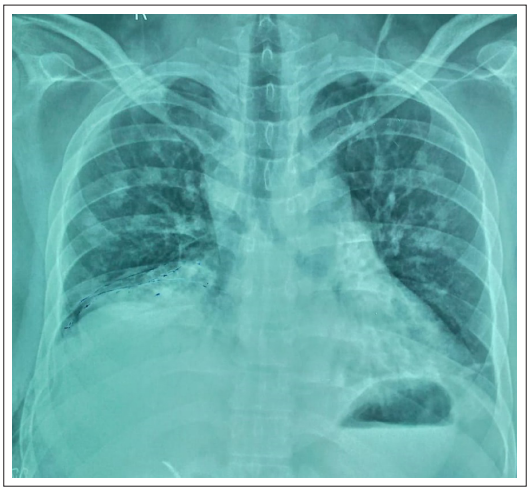

Figure 1: Chest X-ray showing right sided pleural effusion with right sided chest drain inside

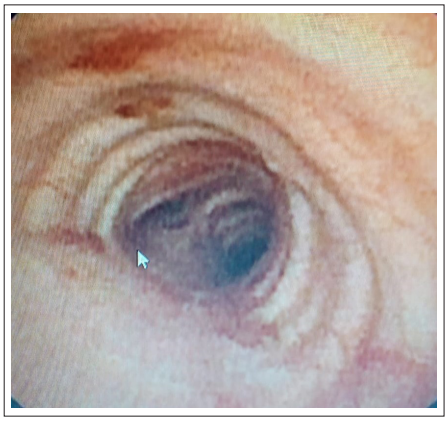

Figure 2: Fibreoptic bronchoscopy (FOB) showing normal trachea and carina no endobronchial growth seen in bilateral bronchial tree

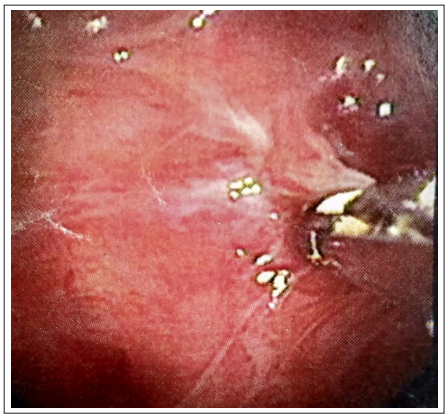

Figure 3: Showing thoracoscopy guided biopsy taken from costal pleura

Discussion

According to World Health Organization (WHO) classification of lung cancer 2004, five subtypes of carcinosarcoma include spindle cell carcinoma, pleomorphic carcinoma, giant cell carcinoma, carcinosarcoma, and pulmonary blastoma [5]. Usually presents at fifth to sixth decade of life with a male-to-female ratio of 4:1. Most common symptoms at presentations are breathing difficulties, fatigue, chest pain, fever, weight loss, and appetite loss. Common radiological findings include a solitary mass involving lung parenchyma, mediastinum and hilum. But pleural involvement is rarely seen at presentation [6]. However, our case we could not find any mass lesion lung parenchyma or mediastinum in chest CT imaging it should be also considered as one of presentation of carcinosarcoma in future. On microscopy most common findings are spindle cell elements or fibrosarcoma followed by chondrosarcoma, osteosarcoma, and rhabdomyosarcoma [6, 7]. The carcinomatous component is mostly squamous cell carcinoma (69%), then adenocarcinoma (20%) and the least common is large cell carcinoma seen in only 11% cases [3].

Carcinosarcomas are difficult to diagnose preoperatively because of heterogeneity especially in atypical presentation like in our case. Hence strong suspicion and extensive workup is required for early diagnosis and curative surgery. Only sarcomatous component can be major finding in biopsy masking the carcinomatous component, hence extensive biopsy deep lesion sampling is recommended from multiple sites to get exact diagnosis. The immunohistochemical stains used for epithelial elements are cytokeratin, P40, p63, TTF1 and Napsin for epithelial component, the sarcomatous component is negative for keratin. Other markers for rhabdomyosarcoma are desmin, myogenin, and MyoD1 and immunoreactive with S100 are positive in chondrosarcoma: however osteosarcoma shows osteocalcin stains the osteoid matrix [4]. The differential diagnoses of carcinosarcoma are carcinoma with desmoplastic stroma, malignant mesothelioma, and metastatic sarcoma [5]. The prognosis of the endobronchial carcinosarcoma is better than that for the peripheral carcinosarcoma [7]. Though therapy pulmonary carcinosarcoma is yet to be established due to the scarce incidence of this malignancy. However, curative intent complete surgical resection is the treatment of choice for pulmonary carcinosarcoma whenever possible. In case of advanced disease systemic chemotherapy is considered along with other supportive therapy with palliative intend. Distant metastasis is common to the lymph node brain, liver, adrenal gland, bone, kidney, and liver. There is a worse prognosis than for conventional non-small cell carcinoma especially in sarcomatous component due to its high tendency for distant metastasis [7].

Conclusions

Pulmonary carcinosarcoma is a rare lung tumour with worse prognosis that presents either as a solid mass involving the peripheral lung parenchyma, involving the endobronchial tree or pleural involvement with or without pleural effusion. Early diagnosis and surgical resection have favourable outcome [8].

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