



“A CASE REPORT: SMALL ROUND CELL CARCINOMA OF LUNG PRESENTED AS GROSS PLEURAL EFFUSION ADMITTED IN THE P.D.U. CIVIL HOSPITAL, RAJKOT, GUJARAT.”

Medicine

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ABSTRACT

Desmoplastic small round cell tumour (DSRCT) is a rare, aggressive and malignant tumour that is characterized by nests of small tumour cells surrounded by a cellular and vascular collagenous stroma and predominantly affects young adolescent males. This tumour most commonly originates in the abdomen; however, in rare cases, DSRCT can originate in other body regions. SRCT in the lung is extremely rare. Here we present an unusual case of pulmonary SRCT diagnosed in an old male on basis of CT guided biopsy and immunohistochemistry.

KEYWORDS

Small Round cell Tumour (SRCT), CT Scan, FNAC, Biopsy

INTRODUCTION

Small round cell tumour (SRCT) is a clinically and morphologically well-defined neoplasm^[1]. This highly aggressive malignant small cell neoplasm tends to affect adolescents and young adults and occurs predominantly in the abdomen, pelvis, and omentum^[1,2]. The average life span is less than two years. It was first described by Gerald and Rosai in the year 1989, when they noticed a distinctive type of small cell tumour, which predominantly involved the abdomen and affected young males^[3]. Gerald et al. in their study of 19 patients of desmoplastic small-round-cell tumour concluded that these tumours occurred predominantly in the abdomen, with the mean age of occurrence being 18.4 years, and had prediction for adolescent males. The tumour carries a characteristic t[11;22] [p13;q12] translocation, which leads to oncogenesis^[4]. They also noticed that these tumours had inconstant distant organ involvement with immunohistochemical reactivity for epithelial, neural, and muscle markers and a highly aggressive behaviour^[5]. This property along with its characteristic translocation, differentiates this tumour from other round cell tumours.

CASE REPORT

A 60year old male, chronic smoker, farmer by occupation, admitted to the department of pulmonary medicine for complaining of cough with mild mucoid expectoration, breathlessness, right sided chest pain, decreased appetite and weight loss for 1 months of duration. No complain of fever or hemoptysis. Patient had occasionally abdominal pain, non-colicky, non-radiating not associated with any bowel symptoms. Patient was physically active and stable. Patient does not have any comorbidities

EXAMINATION

Patient was thin old man, afebrile with normal pulse 74/min, BP-118/78mm Hg, RR-18/min and maintaining SpO2 97% on Room Air. Auscultatory finding suggestive of decrease air entry on right side of chest. No any enlarged cervical and axillary lymph nodes palpable on local examination. Digital clubbing not present.

INVESTIGATION

Haematological examinations revealed a haemoglobin of 10.2gm/dl, TLC-24900/cumm with neutrophilia and normal platelet count. other investigations RFT, LFT and coagulation profile were normal. Sputum cytology shows no malignant cell, Sputum AFB & CBNNAT shows negative results. Ultrasonography of chest shows right sided moderate to gross pleural effusion with underlying approx. 10*9cm mass lesion. Chest Thoracostomy performed on patient, pleural fluid was straw coloured with transudative picture. Analysis of pleural fluid showed 6-7 cells with a differential count of 80% polymorphs and 20% lymphocytes. The pleural fluid sugar level was 87 mg/ dl and protein level were 3.1 gm/dl. The Adenosine Deaminase (ADA) level in the pleural fluid was 6.31 U/L (Units/litre). Pleural fluid cytology was negative for malignant cells and did not show any organisms either. Total pleural fluid drain 3000ml. CECT Thorax shows fairly large ill-defined irregular marinated heterogeneous soft tissue density mass lesion size 113*95*152 cm seen at mediastinum encasing mediastinal great vessels, trachea and bronchi extending up to right hilar region.

Mass lesion markedly compress superior vena cava and intraluminal extension which inferiorly extend into right atrium. Lesion engulf all segmental bronchi of right lung field. Moderate pleural effusion on right side with fissural extension seen.

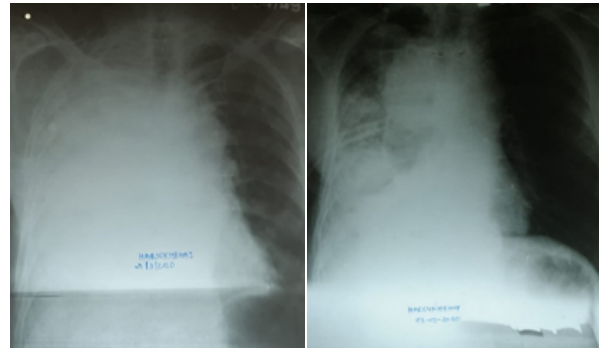


Figure-1: Chest X-ray PA view showing homogenous opacity on the right side involving all zones with obliteration of the costophrenic angle.

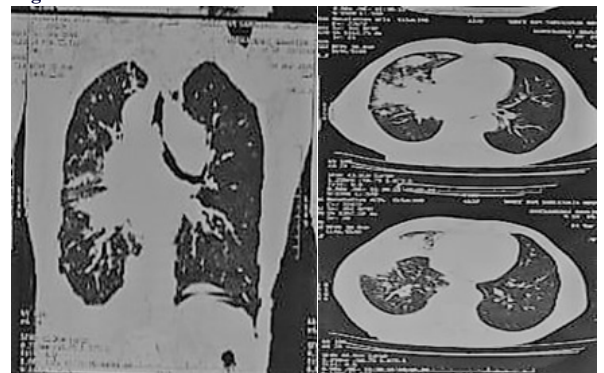


Figure-2 CT scan of thorax (CECT) images shows a mediastinal mass lesion of heterogenous density with right sided pleural effusion.

SUSPICION

On basis of clinical assessment, CT thorax and pleural fluid findings that patient might having following differential diagnosis^[7] of desmoplastic round cell carcinoma which are mention below-

- (1) Small cell mesothelioma
- (2) Primitive neuroectodermal tumour
- (3) Malignant non-Hodgkin's lymphoma
- (4) Small cell carcinoma

CONFIRMATORY INVESTIGATION

CT guided biopsy from right lung mass carried out which grossly shows grey white thread fragments like soft tissue bits.

Microscopically shows atypical mononuclear cells arranged in diffuse manner. They contain hyperchromatic rounded nuclei and scanty cytoplasm which confirmed Round cell carcinoma.

DISCUSSION

Desmoplastic small round cell tumour is a rare tumour that was reported by Gerald and Rosai^[1] in 1989, and it is said to be related to primitive neuroectodermal tumour. It is known to involve the formation of fusion protein of Ewing's sarcoma genes and Wilms' tumour genes, and it has been gaining attention in recent years. Many cases of DSRCT tend to occur in young men, and the site of occurrence is inside the abdominal cavity in approximately 90% or more of the cases, but there have been reports of occurrence in the pleural cavity. However, such reports of occurrence in the pleural cavity have been few^[2,3]. Based on the reports that Wilms's tumour gene expression is observed in these sites of occurrence and in developing mesothelia^[4], DSRCT is suspected of developing from undifferentiated mesothelia, but considering previous reports of occurrence in the brain and the variety of tumours, the possibility of derivation from more highly undifferentiated mesenchymal cells is being studied. Moreover, reports on occurrence in the pleural cavity also included occurrence in the mediastinum and occurrence in the pleura, but there have been no reports of cases presenting with a lung tumour, such as that observed in the present case.

It is treated with a multimodality approach. Those undergoing surgery for tumour resection has better survival, but most of the times the tumour is unresectable^[6].

CONCLUSION

In elderly group of patients with afebrile, chronic cough, coupled with dyspnea and weight loss with multicentric interstitial and parenchymal CT lesion with pleural effusion should raise suspicion of Bronchoalveolar carcinoma.

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