A Rare and Massive Primitive Neuroectodermal Tumour

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Introduction

Primitive neuroectodermal tumours was originally described by Askin et al. [1]. It is commonly found in children and young females. It frequently appears as a chest wall mass and grows rapidly to involve the neighboring pleura. The histogenesis of this rare tumour is uncertain and it is believed to be arising from the intercostal nerves. It exclusively arises from one hemi thorax and progresses rapidly to involve the chest wall. Involvement of the mediastinum in the adult is extremely rare in the literature and till now only five such cases in a span of twenty years have been reported from a tertiary care centre of India [2]. We herein report the sixth case of an anterior mediastinal PNET in an adult female which had progressed rapidly to a huge mass involving the entire left hemithorax and measuring the heaviest PNET reported so far in the literature, (Table 1).

Case Report

A 63-year-old female, cook by profession from Kolkata, India presented with dry cough and progressive shortness of breath, left-sided severe chest pain, loss of weight and low-grade fever for 2 months. The patient was a non-smoker, non-alcoholic and non-diabetic. There was no history of similar illness in the family. General examination revealed pallor and there was no clubbing or peripheral lymphadenopathy.

On examination of the respiratory system features of left hemithorax volume enlargement were present and mediastinum was grossly shifted to the contralateral side. Percussion note was dull over thoracic volume enlargement were present and mediastinum was pallor and there was no clubbing or peripheral lymphadenopathy.

Chest X-ray postero-anterior view revealed left-sided homogenous opacity involving all three zones with loss of costophrenic and cardio phrenic angles along with contralaterally shifted mediastinum suggesting left-sided massive pleural effusion; but on thoracentesis, only 20 ml hemorrhagic fluid could be aspirated from the left infrascapular area. Pleural fluid study yielded brownish-colored, predominantly lymphocytic, exudative fluid with high Adenosine De-aminase (ADA) [Total cell count- 190/cmm, lymphocyte- 90%, neutrophil-5%, ADA-143 IU, protein-4.2 g/dl]. No acid-fast bacillus was found in sputum smear microscopy. Contrast-enhanced CT (CECT) scan showed a huge non-homogenous mass of heterogeneous density occupying the entire left hemi thorax, possibly arising from the anterior mediastinum, causing mediastinal shift to the right (Figure 1). CT-guided Fine Needle Aspiration Cytology (FNAC) suggested it to be a small round-cell tumour (PNET). Fibreoptic bronchoscopy showed no intraluminal growth. A diagnosis of PNET arising from the anterior mediastinum was made, a rare entity.

Patient was planned for surgery after the routine blood investigations which did not reveal any abnormalities. Patient was put on right lateral decubitus position with left hemithorax up and a left posterolateral thoracotomy was done. Left hemi thorax was opened through the fifth left intercostal space. There was a huge 25×15 cms mass occupying the whole of the left hemithorax which was firm to hard in consistency with minimal adhesions to the surrounding pleura. The heart and trachea were pushed to the right. After releasing the adhesions and dissecting it from the neighboring structures the tumour was removed en bloc. The tumour was very heavy and it weighed 9.2 kilograms, the heaviest PNET tumour reported so far in literature (Figure 2). The tumour from the anterior mediastinum, causing mediastinal shift to the right (Figure 1). CT-guided Fine Needle Aspiration Cytology (FNAC) suggested it to be a small round-cell tumour (PNET). Fibreoptic bronchoscopy showed no intraluminal growth. A diagnosis of PNET arising from the anterior mediastinum was made, a rare entity.

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Conclusion

In our report we present an adult female with a 25x15 cms PNET tumor occupying the entire left hemithorax arising from the anterior mediastinum and occupying the entire left hemithorax and weighing 9.2 kilograms, the heaviest and the largest reported in literature so far. Although ES/PNET arising from mediastinum is extremely rare [6-8], it should be considered in the differential diagnosis of mass in the hemithorax in adult patients and may benefit from an aggressive multimodality treatment.

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