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Primary Pulmonary Primitive Neuroectodermal Tumor with CNS Metastasis

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Abstract:

We describe a rare case of primitive neuroectodermal tumor (PNET) originating in the lung of a 20 year old male. A lung mass was detected on a chest radiograph showing opacity with pleural effusion. CT thorax showed mediastinal mass while hypodense mass was seen in right parietal convexity of brain in CT brain. We received multiple grey brown to dark brown friable masses largest of size 13x11x5 cm and other small masses altogether measuring 16x5x8 cm. Cut sections of all masses were grey white and hemorrhagic. Primary pulmonary PNET was reported on histopathology with CNS biopsy also showing similar appearance.

Key words: Bone Neoplasms, Neuroectodermal Tumors, Brain, Sarcoma, Pleural Effusion, Humans.

Introduction

A peripheral primitive neuroectodermal tumor (PNET) is a malignant tumor that usually occurs in bones and soft tissues during childhood or adolescence. PNET constitutes a family of malignant neoplasms comprising small, undifferentiated neuroectodermal cells. PNET tumors arising in the thoracopulmonary region are commonly referred to as Askin tumour [1,2]. A PNET originating in the lung parenchyma is extremely rare and to our knowledge only few cases have been described in practice. We report an aggressive and rapid progressive case of primary pulmonary PNET with CNS metastasis.

Case Report

A 20 year old male was admitted with complaints of chest pain and dyspnoea since 10 days. The chest pain was retrosternal in location and it was non-radiating nor related to the meals. This was accompanied by headache and sudden onset weakness in the left upper limb and lower limb. He also had a history of sudden collapse, 1 month prior to admission. He was a non-smoker and non-alcoholic. He was apparently in good health and reported that he had no systemic disease. On examination it was found he was an averagely built person and had neither clubbing nor lymphadenopathy. Also, all the routine investigations were found to be in the normal limits.

His chest X-ray showed opacity with pleural effusion in right lung. His CT chest showed mediastinal mass with right pleural based lesions [Fig.1a,b]. Moderately enhancing extra-axial hypodense mass lesion in right parietal convexity with midline shift towards left was seen in CT brain [Fig.2a,2b].

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Pleural fluid cytology revealed lymphocytes and reactive mesothelial cells. The differential diagnosis of thymic carcinoma, neuroendocrine carcinoma, malignant lymphoma and hemangioendothelioma were considered.

The patient was taken up for thoracotomy with excision of the tumor. Intraoperatively, it was observed that there was a large mass in the mediastinum with invasion of great vessels and mediastinal lymphnodes. Hence the tumor debulking was done to the extent possible. Complete excision of the tumor could not be done as it was infiltrating the great vessels and pericardium.

We received multiple grey brown to dark brown friable mediastinal masses largest of size 13x11x5 cm and other small masses all together measuring 16x15x8 cm. Cut section of all masses were found to be grey white and hemorrhagic [Fig.3]. The neoplasm was composed of sheets and cords of small hyperchromatic cells with round to oval nuclei containing coarsely granular chromatin and indistinct nucleoli showing scanty cytoplasm. A few Homer-Wright rosettes were also observed with extensive necrosis [Fig.4a-c].

Panel of IHC markers like CD34, LCA, chromogranin, cytokeratin and EMA [Fig.5a] were done which were all found to be negative except CD99 [Fig.5b] which showed rich positivity. Our final diagnosis was primary pulmonary primitive neuroectodermal tumor extending to mediastinum with metastasis into CNS in view of microscopic and immunohistochemistry of the specimen along with CD99 positivity.

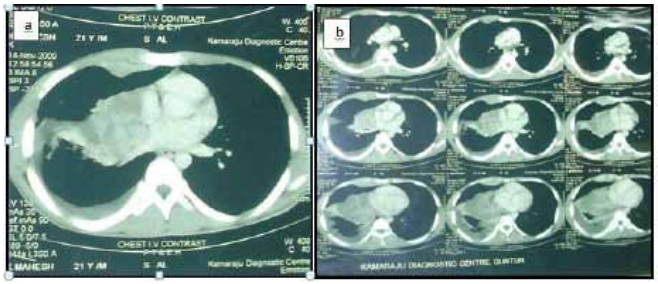

Fig.1(A,B): CT chest plain and contrast showing mediastinal mass lesion with right sided pleural based lesions.

Discussion

PNET are rare tumors occurring in the posterior sulcus or chest wall of adolescent or young adult patients. They are believed to develop from the peripheral nerves such as intercostal nerves. Lesions of PNET are typically painful. Invasive thoracic tumors may either develop or invade on the chest wall, lung or mediastinum [3]. They are generally soft and fleshy with hemorrhagic and necrotic areas. These tumors were classified by Askin and Colleagues as thoracic autonomic neurogenic tumors. A similar

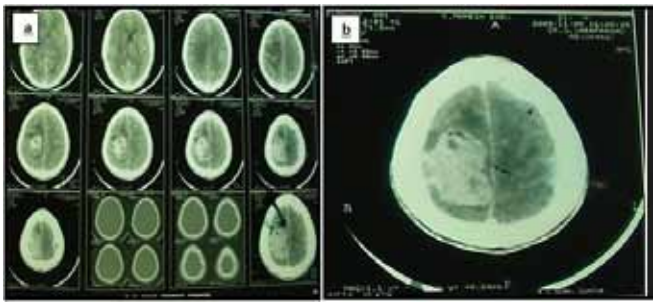

Fig.2(A,B): CT brain: Moderately enhancing extraaxial hypodense mass lesion.

Fig.3: Photograph of the resected specimen showing a lobulated, solid, greyish yellow intra-pulmonary mass with focal necrosis.

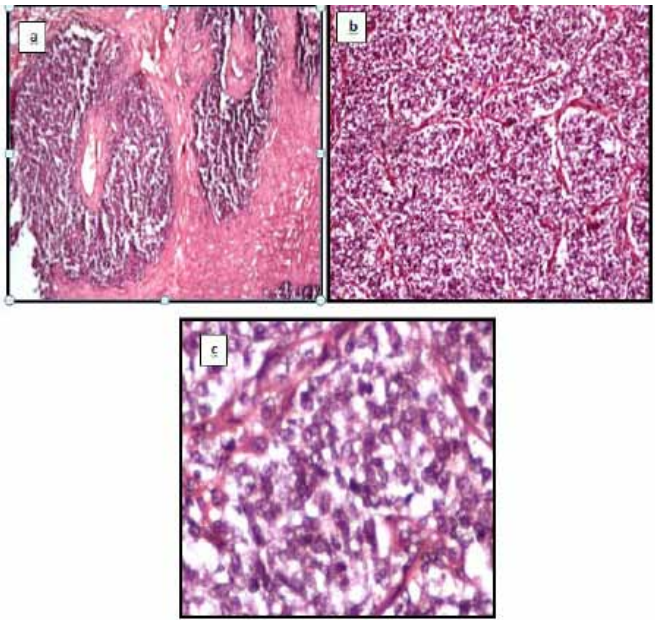

Fig.4(A): Photomicrograph showing tumor cells arranged in Homer-Wright rosette like configuration. **(B & C)**: The tumor cells have round hyperchromatic nuclei with coarsely granular chromatin and indistinct nucleoli.

chromosomal translocation occurring in both Ewings sarcoma and PNET lesions suggest that those tumors are closely related. Typically, Askin tumour develops as a solitary mass or multiple masses in the thoracic area [4]. In thoracic area, these tumors are invasive and prone to destroying bone. Once resected, they recur with extremely high frequency. The most common recurrence sites are the skeleton and CNS. Characteristic PNET pathological findings include compact nests of small cells and Homer-Wright pseudorosettes with an acidophilic core of neurofibrillary character [5,6]. An abnormality of chromosome 22 (e.g., reciprocal translocation of the long arms of chromosomes 11 and 22 or deletion of the long arm of 22) is characteristically present [7].

Primitive neuroectodermal tumour lesions are aggressive and are usually lethal. Once diagnosis of PNET has been made, early wide excision of the

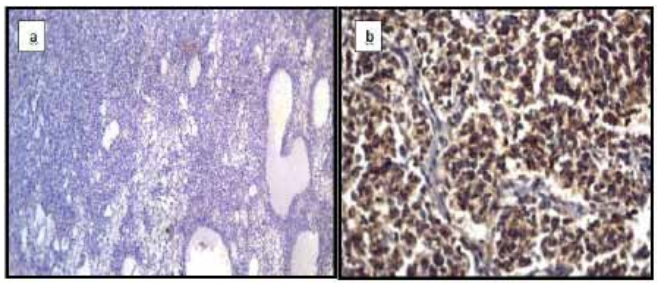

Fig.5(A,B): Photo shows diffused negativity for EMA and strong positivity for CD 99.

tumor along with multimodality chemotherapy and radiotherapy should be undertaken to offer any hope of a long term cure.

The present case describes one of the few reported cases of PNET in an adult. It is unusual for the age of presentation and short duration of unique symptom that is acute onset thoracic pain. Plain chest radiograph and CT scan were diagnostic for pulmonary tumor. Open thoracotomy with excision of the removable tumor proves the diagnosis of PNET. The tumor had invaded the great vessels in the thorax and the mediastinal lymph nodes due to the advanced nature of the tumor.

In summary, we have described a rare example of primary pulmonary PNET which was suggested by a large, heterogenous, enhanced, unilateral lung mass with amorphous calcification and was confirmed by a generous biopsy that yielded an adequate specimen for pathological diagnosis.

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