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Case Report

Primary Primitive Neuroectodermal Tumor (PNET) - A Rare Mediastinal Neoplasm in Elderly

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Abstract

Primary primitive neuroectodermal tumors (PNETs) occur most frequently as tumors of bone and soft tissues of children or young adults. These are small round-cell malignant neoplasms which show varying degrees of neuronal differentiation and are uncommon in adults. Mediastinal mass is a rare presentation of this tumor. We here describe a case of mediastinal PNET in 80 years old lady diagnosed by histopathology and immunohistochemistry.

Keywords: *Primary Primitive Neuroectodermal Tumors; Homer-Wright Rosette; Askin Tumor; Ewings Tumor; Immunohistochemical*

Abbreviations

PNETs: Primary primitive neuroectodermal tumors; PET: positron emission tomography; SUVmax: maximum standardized uptake value, IHC: Immunohistochemical

Introduction

Primary primitive neuroectodermal tumors (PNETs) are small round-cell malignant tumors which show varying degrees of neuronal differentiation. They are considered as a member of EWING family of neoplasms, most frequently arising from the bones and soft tissues in young adults and adolescents [1]. They have also been uncommonly described in other body locations such as testis, kidney, ovary, uterus and pancreas [2]. Occurrence of intrapulmonary and mediastinal PNET, termed as Askin Tumor when localized to thoracopulmonary region, is seen in children and young adults but very rare in elderly [3]. It has been most commonly reported arising from lung and we have recently reported pleural PNET [4] and now we are reporting a case of mediastinal PNET tumor in an 80 years old female.

Case Report

A 80-year-old housewife, non smoker, non alcoholic presented with gradually progressive breathlessness and cough with expectoration for the past 10 years. She was being treated on the lines of obstructive airway disease. Her condition worsened in the past 4 months during which she developed non-radiating, diffuse, left sided chest pain, increased breathlessness and weight loss of around 8 kg.

On examination her vital parameters were stable and lung auscultation revealed bilateral basal crepitations and rhonchi. Hematology and blood chemistry work up were within normal limits. X-ray and HRCT chest revealed a mass lesion in upper mediastinum abutting the chest wall with multiple cystic bronchiectatic lesions in bilateral lungs fields (Figure 1).

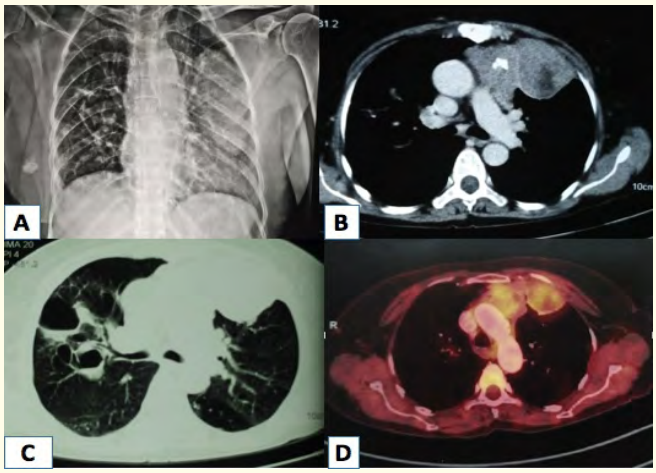

Figure 1: *A) X-ray chest showing a mass lesion in left lung field. B and C) Ct chest revealed a mass lesion in upper mediastinum abutting the chest wall with multiple cystic bronchiectatic lesions in bilateral lungs fields. D) PET-CT showing heterogeneously enhancing, marginated, lobulated, solid-cystic mass in anterior mediastinum extending into left hemithorax.*

The patient underwent a positron emission tomography (PET) CT for disease which revealed FDG avid (maximum standardized uptake value [SUVmax]-5.2) heterogeneously enhancing, marginated, lobulated, solid-cystic mass in anterior mediastinum extending into left hemithorax, abutting great vessels of thorax and infiltrating the pericardium, with right supraclavicular, left internal mammary and mediastinal lymph nodes. No other site of primary was found anywhere else in body on PET avidity. The findings were suggestive of invasive thymoma/ thymic carcinoma/ Askin tumor (Figure 1). Fiberoptic bronchoscopy revealed normal bronchial anatomy. No intrabronchial mass lesion or mucosal abnormality was observed. A CT guided tru-cut biopsy of the lesion was done with no post procedural complications. Histopathology of mass revealed small round discohesive tumor cells with finely granular chromatin and no significant mitosis or necrosis. Small Homer- Wright rosette structures were also seen (Figure 2).

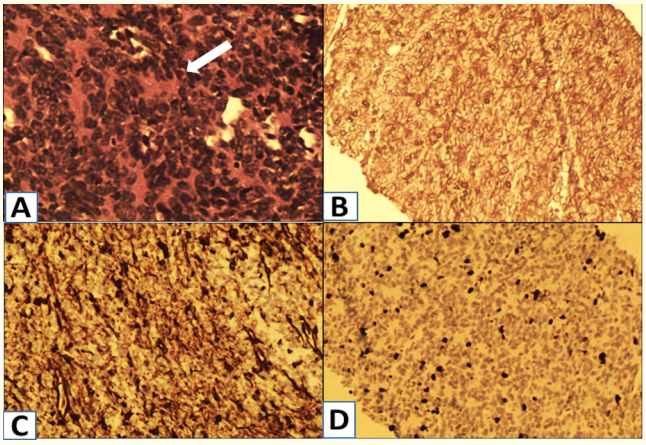

Figure 2: *A) H and E staining of slide showing highly cellular neoplastic tissue. Tumor cells are small round discohesive with finely granular chromatin. Small homer wright rosette like structures also seen (White arrow). B) Showing positive CD99 stain. C) Showing vimentin stain positive. D) Showing KI67 positivity.*

Immuno-histochemical (IHC) findings of tumor cells revealed strong positivity for CD99 and focal positivity for vimentin, synaptophysin, chromogranin and KI67 (20%) (Figure 2), while being negative for LCA, CD56 and thyroid transcription factor (TTF1) ruling out lymphoma and small cell carcinoma. The biopsy and immunohistochemistry findings confirmed PNET of mediastinum. The patient's condition was discussed in the tumor board and palliative chemotherapy was started and presently doing well on follow-up.

Discussion

Askin tumour was first defined by Askin and Rosai [3] Once considered as distinct entities, primitive PNET, Ewing's sarcoma and Askin tumors are now considered as members of the Ewing's family of tumors and when localized to the thoraco-pulmonary region are termed as Askin tumours [3].

These are small round-cell malignant neoplasms of neuroectodermal origin and are rare posterior sulcus or chest wall tumors occurring in the adolescent or young adult patients [4]. These tumors are more common in males (M: F = 1.8: 1) with an average age of presentation at 28.2 years (range 8 - 56 years) [5]. Mostly arising from peripheral lung parenchyma, they present with non specific pulmonary symptoms like cough, fever, dyspnea, hemoptysis, and chest pain [6].

Radiologically, differentiating these tumors from other thoracic neoplasms is difficult. The differential diagnosis of PNET of the lung includes small cell carcinoma and other small round-cell tumors such as malignant lymphoma, granulocytic sarcoma, rhabdomyosarcoma, classical neuroblastoma, and synovial sarcoma [7]. Histopathology and IHC are required to confirm the diagnosis. Nests of small cells and Homer-Wright rosettes with an acidophilic positivity of neurofibrillar elements are characteristic histopathological findings in PNET [8]. The biopsy in our patient showed infiltration with round and spindle-shaped tumor cells with characteristic Homer Wright rosette (Figure 2).

Several immunohistochemical (IHC) markers and antibodies such as O13, HBA-71, and 12E7 (the MIC2 gene product) that recognizes the cell surface antigen, defined by the cluster of CD99, facilitate the diagnosis [9]. In our case TTF1markers were negative on IHC, which ruled out adenocarcinoma. Vimentin and CD-99 were positive, so a diagnosis of sarcoma was considered. IHC expression of the MIC2 gene product (CD-99) in our case is helpful in separating PNET from other small round cell tumors such as sarcomas [4]. Further IHC revealed CK negativity, ruling out sarcoma.

PNET is a highly malignant tumor with a very poor prognosis. This tumor is extremely rare in old age with few case reports are described in literature. The treatment modalities for this tumor includes various combinations of radical surgical resection, neoadjuvant and adjuvant chemotherapy, and irradiation. The 2-year survival rates are 33%, 66%, and 33%, respectively [10].

This report constitutes the first case of PNET arising from the anterior mediastinum. In patients with mediastinal mass, primary PNET of the mediastinum should be considered as differential diagnosis of primary lung cancer and IHC must be performed in all small round cell tumors.

Conflict of Interest

Nil.

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