

Pleuropulmonary Blastoma With Cervical Lymph Node Metastasis at Presentation

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Pleuropulmonary blastoma is a rare childhood neoplasm accounting for less than one percent of all primary malignant lung tumors of children less than six years of age. Metastasis to CNS, orbit and iris, bone, contralateral lung and rarely adrenal glands, liver, kidney and pancreas has been described. This report presents a rare case of pleuropulmonary blastoma with cervical lymph node metastasis at the time of presentation.

Key words: Child, Metastasis, Pleuropulmonary blastoma, Tumor.

Pleuropulmonary blastoma is a rare, dysembryonic childhood neoplasm first described in eleven children by Manivel, *et al.* [1] in 1988. The tumor is a dysembryonic neoplasm of thoracopulmonary mesenchyme and arises from the lung, pleural surface or both [2]. The exact histogenesis is unknown. Pleuropulmonary blastoma is classified into type I (cystic), type II (cystic and solid) and type III (predominantly solid) lesions [3]. Metastasis to cervical lymph node is very rare [4].

CASE REPORT

A 9 year old boy presented to the pediatric OPD with persistent cough, left sided chest pain and cervical lymphadenopathy. Imaging and bronchoscopic findings were suggestive of left sided consolidation with collapse and mediastinal lymphadenopathy. Computerized tomography of the chest revealed a large heterogenous mass lesion occupying the whole of left hemithorax with areas of punctuate, coarse calcification. Lytic areas with sclerosis were seen in D3 vertebral body, suggesting metastasis.

Section of the cervical lymph node showed effacement of nodular architecture with a rim of preserved lymphoid tissue at the periphery. The node was infiltrated by a biphasic tumor comprising of blastemal and sarcomatous elements. The blastemal cells were present in nests and cords, had scanty

cytoplasm with hyperchromatic and pleomorphic nuclei, granular chromatin and inconspicuous nucleoli. These cells blended with the surrounding sarcomatous areas comprising of spindle shaped cells in fibrous stroma exhibiting fascicular pattern (Solid Type III pattern). Sections from the lung biopsy obtained on exploratory thoracotomy showed type II pattern with solid and cystic areas. Histopathology of the resected lung tumour was consistent with type II pattern of pleuropulmonary blastoma. Immunohistochemistry for vimentin, desmin, and smooth muscle actin were positive in the interstitial cells present in the solid areas, further confirming the diagnosis of pleuropulmonary blastoma. Cytokeratin and Epithelial membrane antigen were strongly positive in the lining epithelium in the cystic areas.

DISCUSSION

Priest, *et al.* [4] studied the clinicopathological correlates in 50 patients with pleuropulmonary blastoma, The most common presentation was respiratory distress. Other symptoms included fever, chest or abdominal pain, cough and malaise and common sites for metastasis were the central nervous system (11 patients), orbit and iris [5]. Other sites of metastasis included bone, adrenal, liver, kidney, pancreas and rarely ovary and lymph nodes [4]. Two patients had recurrences in the contralateral chest. None showed metastasis to the lymph nodes. Our case presented with cervical lymphadenopathy,

which is a rare presenting feature [4].

The lymph node and lung biopsies showed two different morphological patterns – Type II and Type III. In the lung biopsy Type II cystic areas were lined by cuboidal epithelium. However, biopsy from cervical lymph node showed predominantly type III solid areas characterized by blastemal cells and sarcomatous areas arranged in a fascicular pattern. Since the morphology of solid Type III lesion exhibits blastemal and stromal components [4]; hence this lesion must be distinguished from adult type pulmonary blastoma which also consists of epithelial and stromal components. Skeletal muscle and cartilaginous differentiation have also been described but was not seen in our case. Transition from Type I lesion to Type III lesion has been previously described by Wright, *et al.* [6].

Five year survival for type II and III lesions is approximately 42% after multimodality therapy. Patients with pleural, mediastinal and extrapulmonary involvement have worse prognosis. Treatment of pleuropulmonary blastoma is radical surgery followed by chemotherapy and radiotherapy or both, because response to radiotherapy is not so good. The patient was treated with VAC

chemotherapy and follow up has so far been uneventful.

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