

Pleuropulmonary blastoma in an adult patient: Report of a case

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ABSTRACT

Pleuropulmonary blastoma (PPB) typically presents in young children, most younger than 5 years, as dysontogenetic neoplasm. It has poor prognosis with three different subtypes: cystic (type I), combined cystic and solid (type II) and solid (type III). Our patient was admitted with dyspnea and was diagnosed at 25 years of age. He had upper right lobectomy and diagnosed as Type II PPB followed by alternating IE/VAC (Ifosfamide, etoposide/vincristine, dactinomycin, cyclophosphamide) polychemotherapy. Immunohistochemical studies showed vimentin positive while cytogenetic studies showed absence of trisomies 8 and 2 abnormalities. Our patient is presently alive and disease-free thirty months after diagnosis. It is not possible to obtain definite knowledge about the prognostic factors and survival rates in adult type PPB. Complete resection is proposed to be one of the important prognostic factors in literature. Longer follow up results in larger patient groups are required for obtaining definitive data. [Turk J Cancer 2007;37(4):158-161]

KEY WORDS: Pleuropulmonary, blastoma, adults

INTRODUCTION

Pleuropulmonary blastoma (PPB) is a rare pulmonary tumor usually observed in childhood under the age of 6 years (1). In 1988, Manivel et al. (2) described PPB in children as an entity that was distinct from the biphasic epithelial-stromal morphology of the classic adult type, and termed as PPB, which includes tumors that previously have been described as pulmonary blastoma, pulmonary sarcomas, embryonal sarcoma, pulmonary rhabdomyosarcoma. These tumors are rare dysontogenetic childhood tumors. The primitive sarcomatous features of the tumor are analogous to those of other dysontogenetic or dysembryonic tumors, such as Wilms tumor, hepatoblastoma, neuroblastoma, and embryonic rhabdomyosarcoma (3). PPB was classified into three groups by Dehner in 1995 as cystic (type I), mixed (type II), or solid (type III). Type I has a more favorable prognosis than type II and III (4). PPB is observed rarely in adults and there is no optimal treatment regimen that has been defined yet. In this report, a case of PPB is presented and reviewed briefly in the light of the literature.

CASE REPORT

A 25 years-old female patient with no history of smoking referred to our clinic with complaints of dyspnea and irritative cough that had been present for the previous 7-8 months. Although there were no pathologic findings in her physical examination, her chest radiography revealed

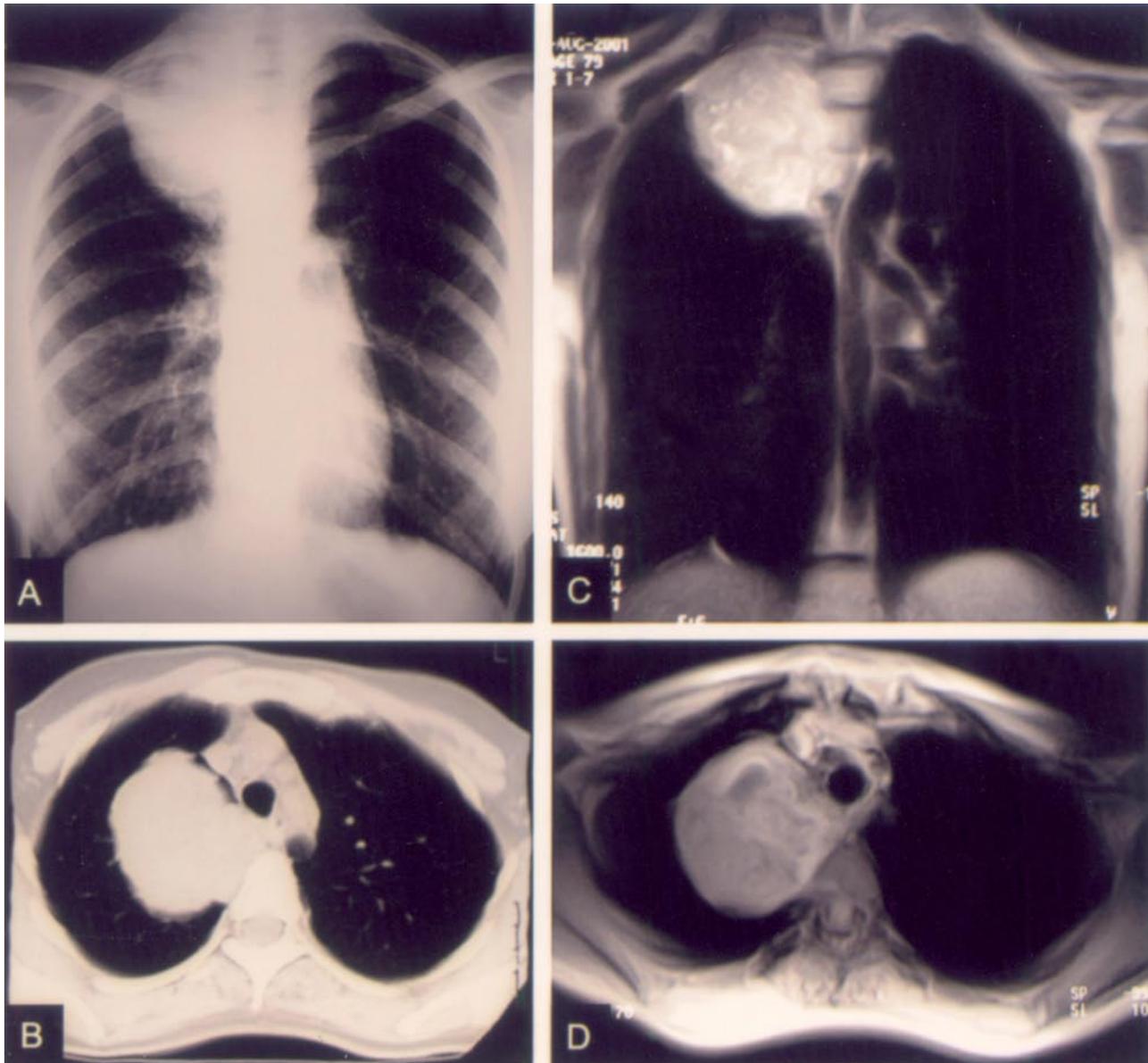


Fig 1 (A,B,C,D). The posterior-anterior chest X-ray, computed tomography and magnetic resonance imaging of the lung in our patient with pleuropulmonary blastoma. (A): The tumoral lesion located in the upper zone of right lung in the neighborhood of upper mediastinum is shown in the posterior-anterior chest X-ray; (B): In the computed tomography the contour of the tumoral lesion seems to be smooth located near the vertebra, which could be demonstrated separately from the vertebrae; (C,D): T1A and T2A MRI images. The lesion seems to be isointense in T1A and slightly hyperintense in T2A images. The lesion with the heterogeneous parenchyme can be easily separated from neighborhood tissues

a smooth-contoured opacity with a 7-8 cm diameter in the upper-right zone (Figure 1A). Computed tomography of the thorax revealed a 7.2x5.9 cm mass lesion in the medial part of upper-right lobe and extending to the right paratracheal area (Figure 1B, C, D). There was no distant metastasis. The patient was operated via a right thoracotomy and upper right lobectomy including tumor with clear margins was performed

Pathological examination revealed an 8 cm spheroid shaped tumor, yellow-orange color in cut surface with regular contour (Figure 2). A tumoral tissue with large necrosis area and with slightly irregular edges was detected in the pulmonary parenchyme under light microscopy. The tumor consisted of cells with ovoid or circular shapes, vesicular nuclei, some distinguishable nucleoli, but with atypical indistinct cytoplasmic bordered cells, pseudo-

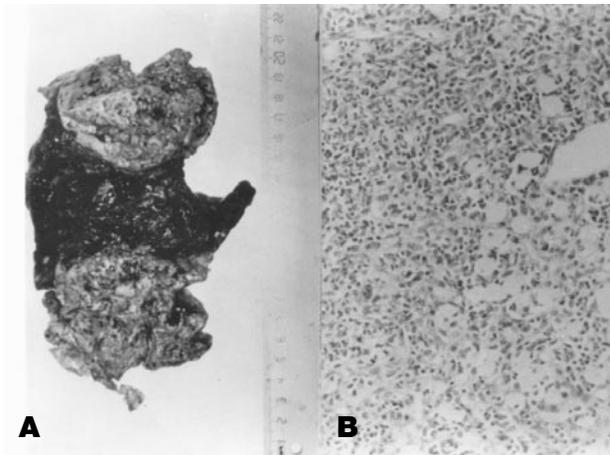


Fig 2 (A,B). (A): Macroscopy of the lesion shows well demarcated with heterogeneous necrotic and hemorrhagic appearance; (B): Microscopical examinations revealed that the tumoral lesion occurred spherical, oval shaped cells with hyperchromatic nuclei and scant cytoplasm and histiocytes with foamy cytoplasm scattered within these cells (H&E, x 200)

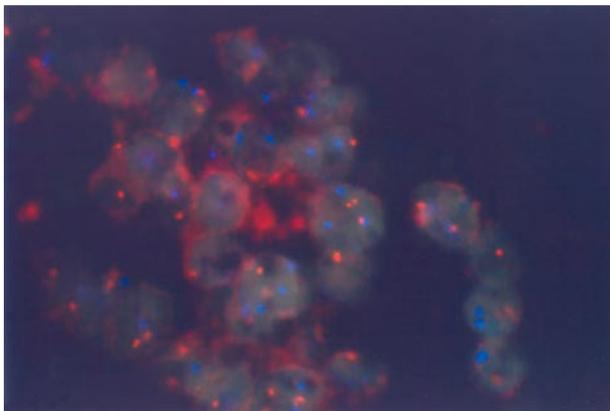


Fig 3. No anomaly of trisomies 8 and 2 was detected with FISH method

glandular and alveolar structures, narrow sequences forming cavities and diffused in form. Pleomorphism and multinucleasia were noted in tumoral cells. Also, lymphocytic infiltration was observed in peritumoral areas, with lipid macrophage accumulation in some areas. In the immunohistochemistry; positive staining was detected for vimentin as well as factor VIII-related antigen and anti-smooth muscle antigen (SMA) at the tumor cells' walls, in histiocytes with lysozyme and negative staining with S-100 protein (S-100), placental alkaline phosphatase (PLAP), neuron-specific enolase (NSE), leukocyte common antigen (LCA), carcinoembryonic antigen (CEA), neurofilament, alpha-fetoprotein (AFP), desmin, synaptophysin and cytokeratin. In tumor cell, cytoplasm was PAS positive stained (Figure 2). These pathological examinations confirmed the diagnosis of PPB. We investigated chro-

mosomal anomalies of chromosom 2 and 8 for trisomy with florescence in situ hybridization method (FISH) in darkness (5). Although one hundred cells were counted, trisomy in chromosome 2 and chromosome 8 was not detected (Figure 3). Adjuvant ifosfamide, etoposide/vincristine, dactinomycin, cyclophosphamide (IE/VAC) alternating regimen was administered. The patient completed the sixth chemotherapy cycle and then was put on a routine follow-up programme. The patient continues to be in a disease-free state at the thirty-sixth month.

DISCUSSION

PPB is mainly distinct from classic adult-type pulmonary blastoma by the absence of malignant epithelial elements (6). In pediatric patients the lesion is a true dys-embryonic neoplasm of thoracopulmonary mesenchyma, without malignant epithelial cells. Pulmonary blastoma with carcinomatous elements has only been described in adults (2). Engagement of parietal pleura and diaphragm can often be observed in type II and III, which have poorer prognoses (7). Type II is similar to Wilms tumor morphologically and therefore, it is sometimes incorrectly called "extra-renal Wilms tumor" (8). Three pathologic types based on gross and microscopic features have been defined in the following manner: type I is a purely cystic lesion that may be mistaken clinically and pathologically for a congenital lung cyst; type II is a cystic and solid lesion with areas of thickening and/or nodularity with or without a relationship to the cysts; and type III is a purely solid tumor consisting of friable, gelatinous to mucoid, lobulated tissue often accompanied by hemorrhage and necrosis (3). In immunohistochemical studies; the most common findings were vimentin positivity and S-100 protein positivity in cartilagenous foci positivity and desmin positivity in areas with rhabdomyoblastic differentiation. The only typical characteristic of tumor is vimentin positivity (9). In our patient, pathological evaluation of tumor was compatible with type II PPB. Cytogenetic studies of childhood cases, chromosome anomalies of trisomy 8 and 2 karyotypic abnormality has been detected (5,10). But in our adult case, interestingly karyotypic abnormalities such as trisomies 8 and 2 were not demonstrated.

Although surgery can yield positive results in type I lesions, surgical success is rather more limited in type II and III (1). The most important prognostic factor is the to-

tal excision of the mass with clear margins. Postoperative radiotherapy is recommended in cases with incomplete resections (11). Cyclophosphamide, doxorubicin, ifosfamide, etoposide, vincristine are commonly used agents in the treatment of PPB. The most common combination is vincristine, dactinomycin, cyclophosphamide (VAC) regimen. This treatment should be reserved for sarcomatous lesions in particular (12). Our patient received a total of 6 cycles of IE/VAC regimen postoperatively. Toxicities of the regimen were consisted of Grade 3/4 leukopenia in two cycles with one febrile neutropenic episode.

Although metastases may not be present at diagnosis in these patients, brain, liver and bone metastases can develop in addition to local recurrence at the follow-up (13). Recurrence and distant metastases, most commonly in the central nervous system, can be observed in approximately half of the patients (12). The poor prognostic factors of the disease are origin of cystic pulmonary disease, tumor

size exceeding 5 cm, and mediastinal and/or pleural invasion. Priest et al. (7) suggested that 2 years and 5 years survivals were 62% and 42% respectively, despite aggressive treatment in type II and III PPB. Our case has been followed for thirty-sixth months without local recurrence and metastases.

CONCLUSION

PPB is a rare lung cancer group mostly detected in the childhood and trisomy 2 and trisomy 8 is observed in cytogenetic studies. Our case is an interesting one as it is seen in adulthood and without the presence of trisomy 8 and trisomy 2. Surgical excision with clear margins seems to be cornerstone of the treatment of this tumor. Efficacy of the adjuvant chemotherapy should be determined in large case series especially in patients with type II-III disease.

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