Primary Pulmonary Rhabdomyosarcoma in Childhood: Clinico-Biologic Features in Two Cases With Review of the Literature—Erratum

To the Editor: In 1996, we reported on two cases of rhabdomyosarcoma (RMS) of the lung arising from normal lung tissue without previous cystic malformations [1]. The histology of these two cases was recently reviewed by the consultants of the International Pleuropulmonary Blastoma Registry. Together with those consultants, we want to amend the original report. After pathologic review, these cases are now classified as Type III pleuropulmonary blastoma (PPB) rather than primary pulmonary RMS. Although these patients received initial complete surgery, postoperative chemotherapy and local radiotherapy, the clinical evolution of the tumor was fatal in both cases. One of these children died from brain metastases, a unique and common complication of PPB [2]. The other child died from respiratory distress after relapse. Microscopically, both tumors contained a rhabdomyosarcomatous element, but both also had blastematous and spindle cell sarcomatous areas. The rhabdomyosarcomatous areas in PPB are identical to embryonal RMS. The presence of high grade blastemal elements, cartilage, spindle cell sarcoma and/or diffuse, marked anaplasia should suggest a diagnosis of PPB for lung masses in children 6 years of age or under [3]. Recently, a similar correction was reported in Pediatric Blood and Cancer and the importance of identifying PPB was stressed [4]. The treatment approaches for PPB and RMS are similar, but unlike RMS, PPB has a propensity for brain metastasis [2]. In addition, PPB is a strikingly familial cancer with genetic implications for others in the immediate and extended families [5]. We appreciate the opportunity to correct the histopathology of these two cases as the importance of the distinction between PPB and RMS is now more evident 12 years after the original report.

Amalia Schiavetti, MD*
Department of Pediatrics
University ''Sapienza''
Rome, Italy

Paolo Indolfi, MD
Department of Pediatrics
Second University of Napoli
Naples, Italy

D. Ashley Hill, MD
Division of Anatomic Pathology
Lauren V Ackerman Laboratory of Surgical Pathology
Barnes-Jewish and St Louis Children's Hospitals
Washington University Medical Center
St. Louis, Missouri

John R. Priest, MD
International Pleuropulmonary Blastoma Registry
Children's Hospitals and Clinics of Minnesota
St. Paul, Minnesota

REFERENCES

1. Schiavetti A, Dominici C, Matrunola M, et al. Primary pulmonary rhabdomyosarcoma in childhood: Clinico-biologic features in two cases with review of the literature. Med Pediatr Oncol 1996;26: 201–207.
2. Priest JR, Magnuson J, Williams GM, et al. Cerebral metastasis and other central nervous system complications of pleuropulmonary blastoma. Pediatr Blood Cancer 2007;49:266–273.
3. Dehner LP. Pleuropulmonary blastoma is THE pulmonary blastoma of childhood. Semin Diagn Pathol 1994;11:144–151. Review.
4. Pai S, Eng HL, Lee SY, et al. Correction: Pleuropulmonary blastoma, not rhabdomyosarcoma in a congenital lung cyst. Pediatr Blood Cancer 2007;48:370–371.
5. Priest JR, Watterson J, Strong L, et al. Pleuropulmonary blastoma: A marker for familial disease. J Pediatr 1996;128:220–224.

*Correspondence to: Amalia Schiavetti, Department of Pediatrics, University ''Sapienza'', Viale Regina Elena, 324, Rome 00161, Italy. E-mail: amalia.schiavetti@uniroma1.it

Received 4 July 2008; Accepted 7 July 2008

DOI 10.1002/pbc.21723
Published online 18 September 2008 in Wiley InterScience (www.interscience.wiley.com)