Abstract

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Pulmonary interstitial glycogenosis associated with a spectrum of neonatal pulmonary disorders

Ernest Cutz
University of Toronto, Canada

Abstract

Primary or isolated pulmonary interstitial glycogenosis (PIG) is a rare disease presenting as tachypnea and hypoxemia during the perinatal period. A diffuse interstitial infiltrate with focal hyperinflation is visible on chest imaging. The biopsy findings include diffuse expansion of the interstitium by spindle-shaped cells with pale cytoplasm that, on electron microscopy (EM), are poorly differentiated mesenchymal cells containing abundant monoparticulate glycogen. This glycogenosis appears to be a transient abnormality, usually with a favorable prognosis. Recently, cases of PIG, some associated with other pulmonary or systemic abnormalities, have been described. The clinical significance and potential role of PIG changes remain unknown. We report 28 cases of PIG associated with a spectrum of pediatric pulmonary and cardiovascular disorders, including arterial hypertensive changes with and without abnormal alveolar development (n = 9), congenital heart disease (CHD; n = 4), hyperplasia of pulmonary neuroendocrine cells resembling neuroendocrine hyperplasia of infancy (NEHI, n = 5), congenital pulmonary airway malformation (n = 5), congenital lobar emphysema (n = 4), and Noonan syndrome (n = 1). In all cases, PIG was confirmed by positive periodic acid–Schiff (PAS) staining, immunopositivity for vimentin, and EM. Although some patients improved with age, 7 died of respiratory failure or complications of CHD, suggesting that PIG may be clinically significant when associated with other severe disorders. The association of PIG with a spectrum of mostly congenital lung disorders supports its origin as a developmental abnormality of interstitial fibroblast differentiation rather than a nonspecific reactive process.

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Biography

Dr. Cutz received his MD from Charles University Medical School, Prague, Czechoslovakia. He contined his post-graduate studies in the Department of Histology& Embryology at Charles University in Prague and at the Centre Anticancereux in Toulouse, France. He later moved to Toronto and joined the Department of Pathology at The Hospital for Sick Children, with work at the Toronto General Hospital, Princess Margaret and Wellesley Hospital. Cutz has an FRCP(C) Speciality cerification in Anatomical Pathology from Royal College of Physicians and Surgeons of Canada.