A Case of Pulmonary Interstitial Glycogenosis

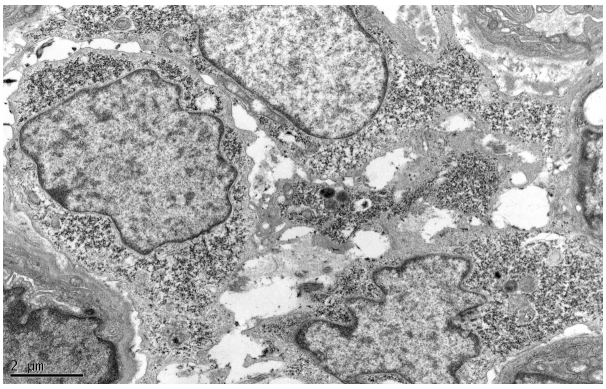
J.K. Brealey[1]

[1] Electron Microscopy Unit, Anatomical Pathology, SA Pathology, Adelaide, Australia.

Abstract

A 2-month-old infant with an undiagnosed lung disease since birth underwent a lung biopsy for investigation. Light microscopy identified round to oval-shaped bland-looking mesenchymal cells within widened alveolar septa. The cells had a foamy appearance, and no significant inflammation was identified. Immunohistochemistry did not reveal any additional significant abnormality. By electron microscopy, the alveoli displayed widened septa that contained increased numbers of interstitial mesenchymal cells bearing abundant glycogen, and other cytoplasmic structures in these cells were very sparse. The myelinosomes with type 2 pneumocytes were interpreted as being within normal limits, and no micro-organisms were identified. With the aid of electron microscopy, a diagnosis of pulmonary interstitial glycogenosis (PIG) was made.

PIG was first described in 2002 and is one of many children's interstitial lung diseases (chILD). The main ultrastructural feature of PIG is abundant glycogen within interstitial mesenchymal cells. Infants with PIG typically present with respiratory distress and hypoxemia at or shortly after birth. PIG is often found in association with other cardiopulmonary disorders. The pathogenesis of PIG is unknown; one hypothesis is it represents a disorder of lung maturation.



Interstitial cells bearing abundant glycogen within a widened alveolar septum.