PAEDIATRIC DERMATOLOGY

A RARE CASE OF EXTENSIVE APLASIA CUTIS WITH ABSENT SEPTUM PELLUCIDUM

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BACKGROUND: The patient is a premature neonate born at 29 weeks gestation with birthweight of 1283g via emergency Cesarian section for maternal chorioamnionitis. The mother had presented at 22 weeks getation with premature rupture of membranes and greenish vaginal discharge. Bacterial cultures from the discharge were negative but she was treated empirically with antibiotics. The mother also had gestations diabetes on diet control. At birth, poor Apgar scores were noted requiring intubation and admission to neonatal intensive care unit. Medical issues included severe respiratory distress syndrome, pulmonary hypertension, chronic lung disease, large patent ductus arteriosus, neonatal jaundice and left inguinal hernia.
A few weeks after birth, the baby was noted to have a 4 by 5 cm patch of atrophic skin over the fronto-temporal scalp associated with extensive alopecia (30% scalp surface) and scalp seborrheoic dermatitis. Neurological and other systemic examination normal. Cranial ultrasound showed absent septum pellucidum.

OBSERVATION: The scalp seborrheic dermatitis resolved with topical treatment. Mild motor delay was noted although neurological examination was normal. She was referred for physiotherapy. At 17 months review, the scalp atrophic patch measured 10cm x 8cm with minimal hair growth and an overlying small yellow calcified nodule. Surgical options were explored by the plastic surgeon including serial excisions, tissue expander, scalp rotation flap and hair transplants.

KEY MESSAGE: Aplasia cutis congenita, localised absent of skin, most commonly affects the vertex of the scalp, less commonly on the face, trunk or extremities. It is usually an isolated finding, and can be single or multiple, with variable size. Associated abnormalities include underlying bone involvement, cerebrovascular defects and neural tube defects, especially if a “hair collar sign” is present. We report a rare association with absent septum pellucidum.