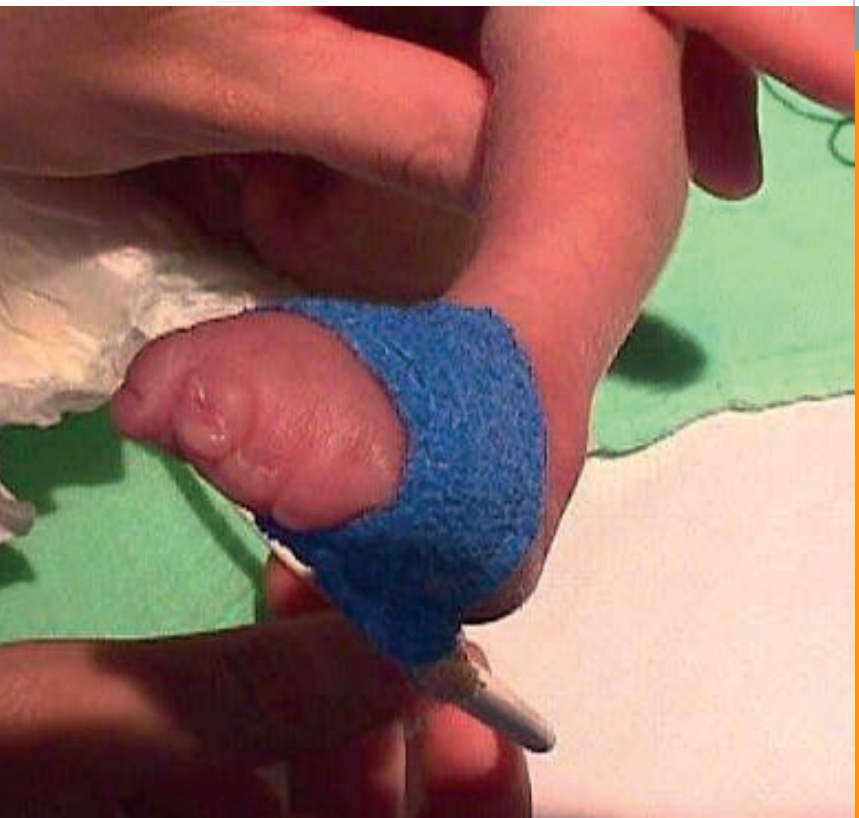
SWISS SOCIETY OF NEONATOLOGY

Adams-Oliver Syndrome



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Berger TM, Neonatal and Pediatric Intensive Care Unit,
Children's Hospital of Lucerne, Switzerland

This female infant was born to a 26-year-old G1 by Cesarean section at 32 3/7 weeks of gestation due to non-reassuring fetal heart rate and absent fetal movements. Cord-pHs were 7.24/7.36 and Apgar scores were 5 at 1, 8 at 5, and 9 at 10 minutes, respectively. NP-CPAP was initiated in the delivery room because of respiratory distress and she was transferred to our institution.

Pregnancy had been complicated by intrauterine growth restriction. Maternal TORCH serologies were normal. Amniocentesis was performed at 28 weeks of gestation and revealed a normal karyotype.

On admission to the NICU, the infant was noted to be symmetrically growth retarded with a birth weight of 1120 g (< P 3), a length of 38.5 cm (P 3-10) and a head circumference of 27.3 cm (< P 3). Her skin had a mottled appearance. She had an area of alopecia on the parieto-occipital area of her head (Fig. 1), several periumbilical skin lesions and scars (Fig. 2) as well as hypoplastic fingers (Dig III-IV right hand) and toes (Dig III-IV right foot, Dig II-V left foot) (Fig. 3).

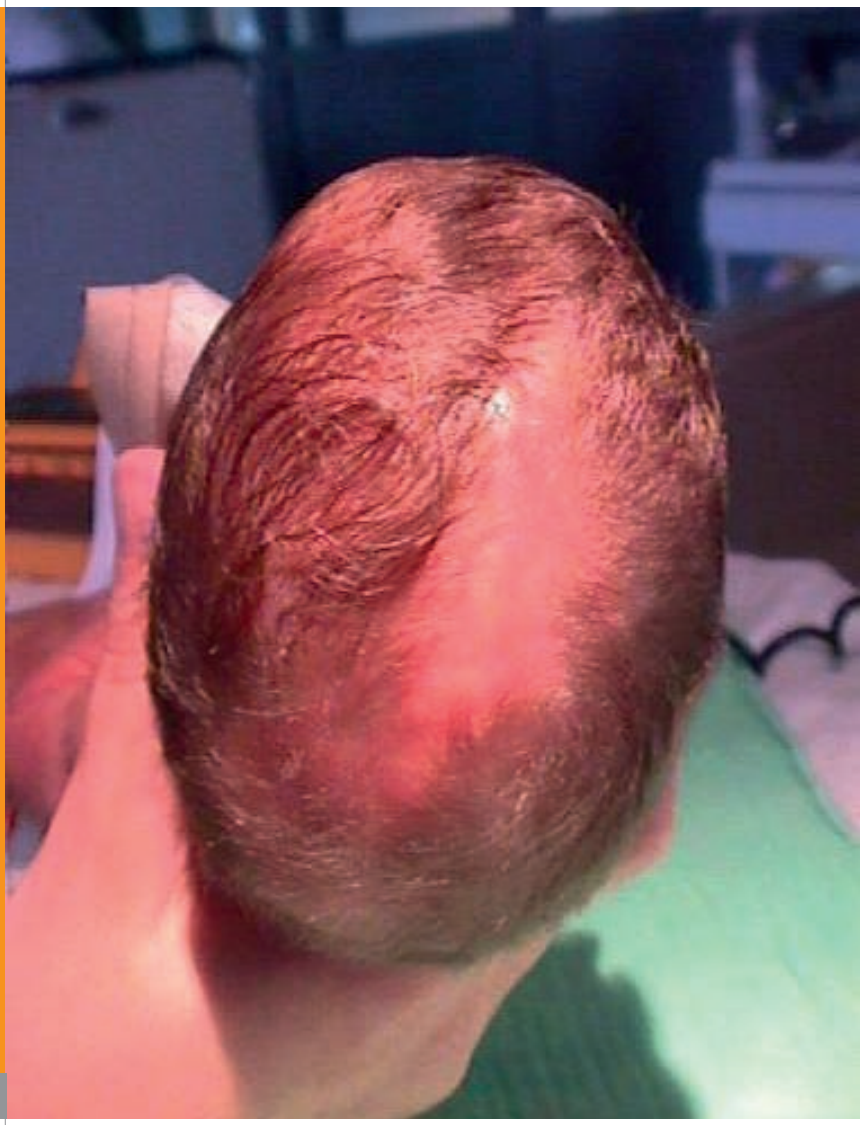

Fig. 1

Alopecia on right side of the scalp.

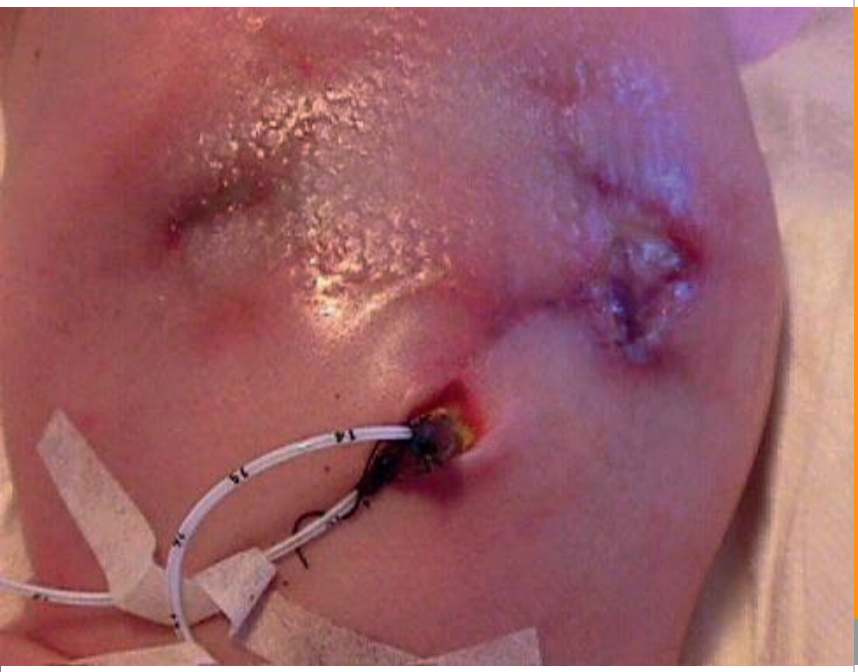

Fig. 2

Periumbilical skin lesions.

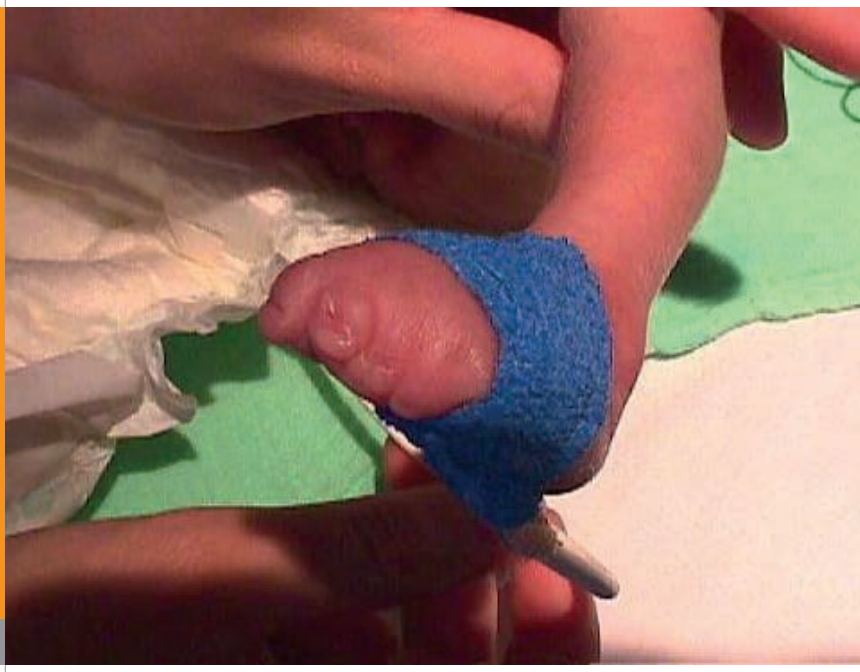

Fig. 3

Hypoplastic toes.

The following additional investigations were performed: cerebral ultrasound examination revealed small but distinct subependymal calcifications; CMV in the urine was negative; fundoscopy was remarkable for very fine, narrow retinal blood vessels; echocardiography was normal.

Based on these findings, a diagnosis of Aplasia cutis congenita or Adams-Oliver syndrome was made. No other cases of this autosomal dominant disorder could be detected in the family. The infant's recovery from her respiratory distress syndrome was uneventful and she was transferred to a hospital closer to her parents' home at a corrected gestational age of 36 weeks.

DISCUSSION

In 1945, Adams and Oliver described eight members of a family with this disorder (1). The syndrome is characterized by mild growth deficiency, aplasia cutis congenita over the posterior parietal region, with or without an underlying defect of bone, as well as variable degrees of terminal transverse limb defects. Cutis marmorata is also frequently seen. Ulcerations of the abdominal skin (2), and intracranial calcifications (3), as seen in our patient, have also previously been described. In the majority of cases, an autosomal dominant inheritance was observed. The underlying pathophysiologic mechanism for this syndrome remains unknown, but the defects may result from impaired circulation in "watershed" areas during a critical period of development (4).

The prognosis is excellent in the majority of cases. Larger scalp defects are more likely to be associated with underlying bone defects, and if the superior sagittal sinus or dura are exposed, there is an increased risk of hemorrhage (2) and infection.

REFERENCES

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CONTACT Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch