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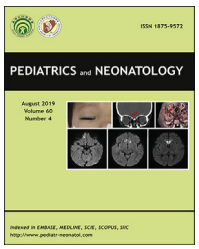

Images

New features of aplasia cutis congenita type 5 — Skin atrophy associated with respiratory insufficiency and multiple intestinal atresia caused by the early death of twin fetus

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A 1-day-old female infant was transferred to our NICU with a diagnosis of a congenital skin defect. She was born from a twin pregnancy complicated with intrauterine twin death in week 12 of pregnancy. Two previous pregnancies had ended early in spontaneous miscarriages. The infant presented hypersensitivity to stimuli, symmetrical skin defects covering 30% of the body, most widely distributed in a butterfly/H-pattern on the trunk, with thigh and elbow marks, and a single horizontal lesion underneath the clavicles and upon the sagittal suture. There was no association with Blaschko's lines nor with dermatomes. The chest-restricting lesions caused mild respiratory insufficiency; a CT scan revealed congenital emphysematous lung lesions. Due to aggravated ileus signs, a laparotomy was performed that revealed multiple small bowel atresia (type I, IV). Skin lesions were managed conservatively and healed properly. The neurologic examination and an EEG test showed no abnormalities, and the patient was discharged home after 40 days.

Based on the clinical presence, a diagnosis of aplasia cutis congenita type 5 was made. This is a non-genetic condition with a typical repeatable location of wounds, occurring when the following three factors are met: 1) multiparous pregnancy, 2) placental vascular twin anastomoses, and 3) early twin death (fetus papyraceus).[1] The exact mechanism remains unknown, but possible ischemia can occur due to 1) altered hemodynamics — a rapid hypotension in the survivor caused by vessels' relaxation in the dying twin[2] and 2) thrombotic events from the necrotic tissues.[3]

The reason for the repeatable location of wounds remains a mystery; an acute ischemic event would result in distal necrosis. We rather state a hypothesis of prolonged ischemia affecting areas of rapid growth and maximum tensile forces. Our case report provides a new insight into the pathogenesis of ACC type 5, since we report for the first time a suspected coincidence of fetus papyraceus with skin, multiple bowel, and lung hypoplasia, confirming its vascular genesis (see Fig. 1).

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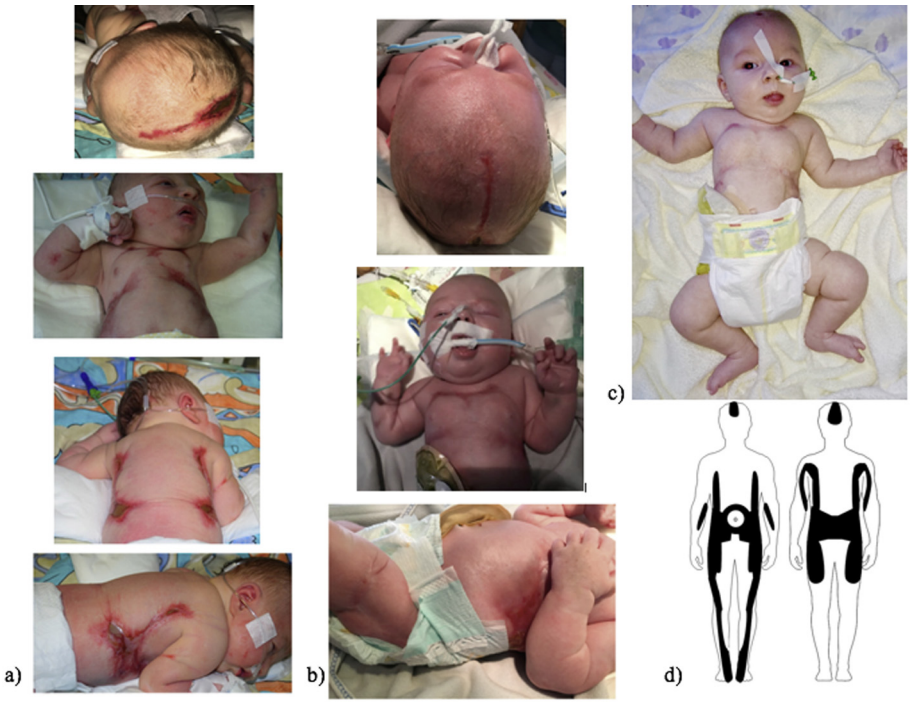

Figure 1 Skin Changes: a) on admission; b) on day 12; c) in the 3rd month of life. d) the distribution pattern of ACC type 5 (reprinted from The Journal of Pediatrics, Vol 153 (6), Schaffer JV, Popiolek DA, Orlow SJ, Symmetric Truncal Aplasia Cutis Congenita following Multifetal Reduction of a Sextuplet Pregnancy, 860—863, Copyright 2018, with permission from Elsevier).

Conflicts of interest

None declared.

Acknowledgements

Written informed consent was obtained from the parents for the publication of this case report and any accompanying images.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.pedneo.2018.09.004.