PAEDIATRIC DERMATOLOGY

A RARE CASE PATAU SYNDROME WITH APLASIA CUTIS CONGENITA

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Background: Aplasia cutis congenita (ACC) is a rare malformation of skin development, characterized by the loss of skin layer from the epidermis, dermis, subcutaneous, or event reach the duramater. This disorder is common associated with other congenital syndrome, but rarely reported with Patau Syndrome (PS). PS or also known as trisomy 13 is the third most common autosomal trisomy. One characterized by multiple congenital anomalies such as of microphthalmia or anophthalmia, cleft lip and/or palate, polydactyly, and ACC.

Observation: An aterm baby girl was born with ulcers on the scalp since birth, with polydactyly on the fingers and toes and associated with rapid respiratory rate without wheezing. Her mother had history of previous abortions for two times, but there was no abnormality during this pregnancy. On orthodontic graphic examination showed Large Ventricular Septal Defect (VSD) sub aortic. Chromosome examination revealed trisomy 13 in this patient. The lesion was treated with wound dressings (WD) of hydrocolloids and improved on the 11th day of observation, unfortunately the patient died due to sepsis and respiratory failure.

Key message: ACC often healed and enclosure with proper treatment. However, other symptoms caused associate congenital disorder, such as PS that could threaten patient.