

UNILATERAL DELAYED CUTANEOUS MATURATION

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Abstract. A newborn infant with hemicorporeal hypopigmentation, alopecia, and nail hypoplasia is reported. The infant had associated mental retardation, microphthalmia, optic nerve dysplasia, and ductus arteriosus. During the first years of life the pigmentary defect apparently resolved, and the hair grew. The nails remained hypoplastic. The infant expired at age 27 months from cardiopulmonary complications.

Hemicorporeal congenital ectodermal defects are rare. Diffuse unilateral epidermolytic hyperkeratosis associated with limb deformities has been studied by Rossman et al. (3) and Cullen et al. (1). Shear et al. (4) found congenital unilateral psoriasis in association with ectromelia and central nervous system anomalies. Others (4-7) have reported unilateral pigmentary changes and limb blood pressure asymmetry. We have recently had the opportunity to study a child with unilateral delayed epidermal, pigmentary and appendageal maturation associated with cardiac and central nervous system abnormalities and believe it may be of interest to report our findings here.

REPORT OF A CASE

The patient was born after 25 weeks of gestation and weighed 1910 g. His mother is a 20-year-old Caucasian woman, gravida 1. There was no family history of cutaneous disease or congenital abnormality. Soon after birth the infant showed signs of congestive heart failure: tachypnea, costal retractions, tachycardia and hepatomegaly; and on his 18th day of life he was transferred to a hospital where a patent ductus arteriosus was ligated.

At birth, the infant had an absence of hair and nails and decreased pigmentation on the left side of the body as well as a patchy, erythematous dermatitis on the left side of his neck, ears, scalp and right foot. Two large patches of alopecia were present on the left frontoparietal and temporal areas of the scalp. Several isolated hairs were seen in the otherwise bald areas.

When the infant was 3½ months old a cutaneous punch

biopsy was performed from a hypopigmented area on the left side of his back. Histological examination of a section from the biopsy showed a flattened epidermis with absence of most of the rete ridges and containing a number of vacuolated epidermal cells. Sweat ducts and glands were seen but no pilosebaceous structures were present in the biopsy specimen. A scattered infiltrate consisting of lymphocytes and a few plasma cells was present throughout the dermis. At this time the child had developed several hypoplastic nails of the left hand and foot. The cardiovascular problem had stabilized and the infant's growth was considered normal for his age. At 4 months of age some hair appeared in the bald areas of the scalp. At one year, the child was found to be physically and mentally retarded. The nails on his left hand and foot remained rudimentary and the diffuse hypopigmentation of the left side of his body had now become spotty. An ophthalmologic examination revealed right microphthalmia with secondary ptosis and nystagmus, as well as probable right optic nerve dysplasia leading to right 6th nerve palsy. At age 17 months he exhibited the developmental age of a 5-month-old child. He was lost to follow-up thereafter, but an interview with the mother revealed that the child had died at 27 months. He had never developed the ability to sit unsupported or feed without help and could not be toilet trained. Hair had grown and completely filled the alopecic areas on the left side of his scalp and almost completely normal pigmentation had appeared on the previously hypopigmented left half of his body.

DISCUSSION

A review of the literature does not reveal any similar combination of delayed unilateral epidermal maturation in combination with cardiac and neurologic maldevelopment. Falek et al. (2) reported two siblings with congenital heart disease, unilateral left rudimentary extremities, and scaly skin over the left side of the body. The cardiac defect involved multiple developmental anomalies, the most striking of which was a single ventricle present in both infants. The abnormalities of the limbs involved all the tissues,

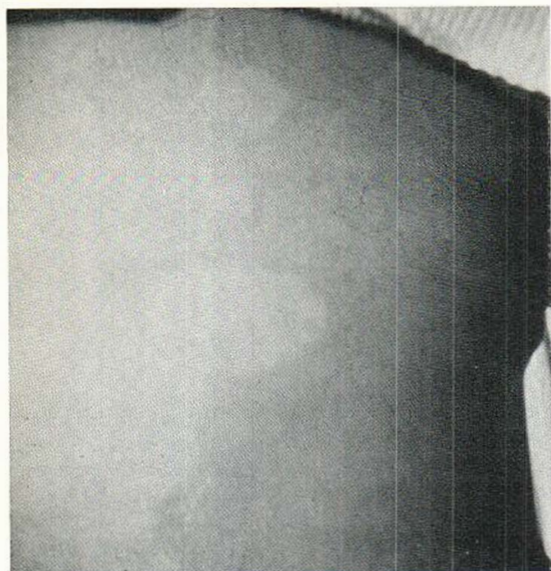


Fig. 1. At 3 months of age, view of the infant's thorax from the back. The leucodermic left hemithorax present since birth.

including bone. Unilateral ichthyosiform dermatitis was present in that patient but the hair was normal as was the cutaneous pigmentation. The children reported by Falek et al. died soon after birth so that the neurologic developmental pattern was not established.

Zlotnikoff (6) described two cases of unilateral pigmentation mosaicism including hyperpigmented

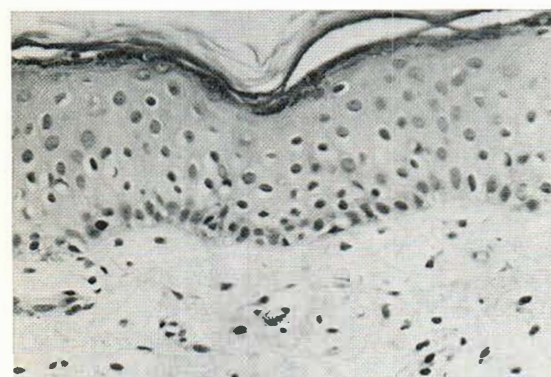


Fig. 2. Histological section from biopsy of leucodermic areas on the back. The epidermal ridges are somewhat flat. Two cells which may be melanocytes are seen in the basal layer. There are some vacuolated cells scattered through the epidermis. A scattered infiltrate of lymphocytes and a few plasma cells are present in the dermis. There is no evidence of pilosebaceous structures.

patches, baldness, differences in pupil coloration, perspiration, neural reflexes and asymmetrical limb blood pressures. The cardiac and neurologic malformations seen in our case were not included in the description of his patients. In 1948 Zellweger & Uehlinger (5) described a case of unilateral cutaneous anomalies associated with hypotrophic development of the right half of the body. None of the cases reported by these authors demonstrated delayed maturation of the epidermal appendages, nor the peculiar combination of delayed and/or aberrant multisystem development.

The congenital nature of the disorder in our patient is certain, but its hereditary nature cannot be ascertained. The number of cases previously described which vaguely resemble the changes found in our patient are too few and the data too sparse to determine whether our patient represents a new syndrome or whether his disorder is a variant of those previously described and discussed above. The constellation of findings is striking enough, however, to have attention drawn to them.

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