## *Brief Reports*

# Epidermal Nevus Syndrome

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The association of epidermal nevus with other developmental defects, particularly of the central nervous system, eye and skeletal system is well recognized. It has now become clear that a wide variety of histological types of epidermal nevus may occur in such cases. We report one such case.

### Case Report

A female baby born by LSCS, to a 24 year old primigravida with no history of antenatal problems and consanguinity, at a peripheral hospital in Bangalore was referred to M.S. Ramaiah Medical College Hospital. There was no family history of similar structural defects. The birth weight was 1.5 Kg and the gestational age was 32 weeks. She was very sick on admission with respiratory distress, which started a few hours after birth.

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The skull was asymmetrical with flattening of the head on the right side. The right eye as well as the cornea was small with narrow palpebral fissure compared to the left eye. The medial half of the right upper eyelid was partially developed. The palpebral conjunctiva was thickened and there was bilateral corneal haziness. The right ear lobule was large and had a crumpled appearance. The hair on the right side of the scalp were long, fuzzy, differing in texture with patchy alopecia.

The striking features were linear tan colored verrucous skin lesions over the face, particularly on the right side extending to the scalp. Similar skin lesions were present on the right upper and lower limbs. The nails were normal *(Fig- 1)*

There was poor peripheral circulation, irregular heart beats and central cyanosis. The baby had congestive cardiac failure as evidenced by hepatomegaly, edema, tachypnea and tachycardia. No murmur was heard. Respiratory system examination revealed bilateral crepitations. The baby was lethargic, hypotonic and had sluggish neonatal reflexes. Eye grounds were normal. There was no focal deficit.

The hemogram and chest X-ray were normal. ECG revealed multiple atrial ectopics with poor right ventricular forces. Echocardiography, CT Scan of the brain and skin biopsy could not be done.

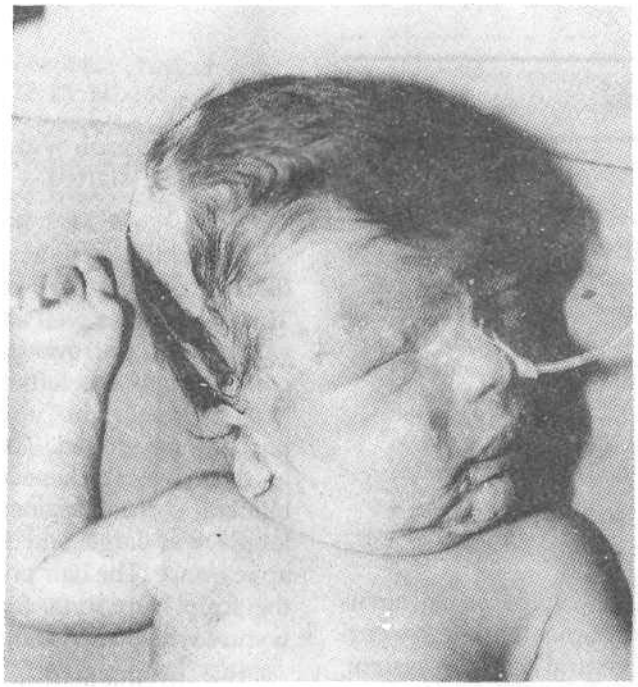

*Fig. 1. Clinical photograph of the neonate showing verrucous epidermal nevi predominantly over the face and trunk with patchy alopecia on the right side of the scalp.*

The baby continued to have respiratory distress with attacks of apnea and cyanosis. She was treated for congestive heart failure with no significant response. The baby also developed multiple generalized tonic-clonic seizures which responded poorly to anticonvulsants. She deteriorated and died after 96 hours of admission.

## Discussion

Epidermal nevus syndrome is a neurocutaneous disorder in which the epidermal nevi are associated with other abnormalities, particularly of the central nervous system, eye and skeleton(l).

It is now recognized that a wide variety of histological types of epidermal nevus may provide for the cutaneous component of this syndrome(2). The skin lesions are mostly sebaceous nevi or verrucous epidermal nevi. These lesions are very closely related and many authors regard them as variants(3). Epidermal nevi occuring on the head and neck are likely to be sebaceous nevus, while those occuring elsewhere are more likely to be verrucous epidermal nevus(4). The propositus reported had skin lesions predominantly over the face, scalp and limbs. The slight verrucosity of the lesions and the extensive distribution suggest that they are verrucous epidermal nevi, though this would be difficult to delienate it from sebaceous nevi, due to lack of histopathology in this case.

Verrucous epidermal nevi are circumscribed hamartomatous lesions which may be present at birth or may appear during childhood. In the newborn period they are velvety streaks or plaques, single or multiple, tending to become more pigmented or verrucous over the years. Significant enlargement could occur at puberty.

Ten per cent of cases have other developmental anomalies(4). Many neurological abnormalities have been identified in the epidermal nevus syndrome, including hemimegalencephaly, gyral malformations, mental retardation and seizures(5). The baby reported had multiple seizures on the fourth postnatal day, which was resistant to therapy.

Ocular abnormalities like colobomata of the eyes, choristomas have been reported(6). A variety of skeletal, cardiac and genito-urinary abnormalities have been described(3). Congenital heart disease with congestive heart failure and arrhythmias could have been the cause of death in the baby reported.

These lesion§ are potentially premalignant with a 15-20% risk of tumor especially basal cell carcinoma(7), hence the early surgical removal of these lesions should be considered.

It seems likely that epidermal nevus reflects somatic mutations occuring in embryonic or fetal life, most of them sporadically.

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