CASE REPORTS

EPIDERMAL NEVUS SYNDROME

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A seven-year-old male child had epidermal nevus syndrome manifesting as delayed mile-stones, micro-cephaly, short stature, central nervous system abnormalities like ocular palsies with nystagmus, cutaneous manifestation of nevus unius lateris and a bone cyst at the lower end of right femur.

Key words : Epidermal nevus syndrome.

Epidermal nevus may be visible at birth or may develop within the first few months or years of life.[1] It affects both sexes equally, and only very rarely it occurs in more than one family member. Initially, the epidermal nevus may appear as a discoloured, slightly scaly patch, but with maturation, it becomes more thickened, verrucous and hyperpigmented.[1] The several morphological types include pigmented papilloma often in a linear distribution, unilateral hyperkeratotic streaks (nevus unius lateris) involving a limb or perhaps a portion of the trunk, velvetty hyperpigmented plaques, and whorled or marbled hyperkeratotic streaks over extensive areas of the body.[2] Some cases of epidermal nevus are associated with abnormalities of the other organs especially the central nervous system and the skeletal system. This combination has been designated as the epidermal nevus syndrome.[2]

Case Report

A seven-year-old male child had delayed developmental mile-stones and skin manifestations confined to the left side of the body. He was the second child of non-consanguineous parents, born at full term by normal delivery. At the time of his birth, the parents noted hyperpigmented patches on the left side of the body. Within a few days, the lesions became hyperpigmented, thickened and later verrucous. At times the lesions became pruritic, followed by infection and crusting.

The child attained head control at the age of five months, turned at the age of 10 months, stood with support at the age of 2 years and walked without support at the age of 4 years. The child started to speak 2 or 3 words with meaning at the age of 4 years. There was no family history of similar complaints.

The child was pale, moderately built and nourished with a head circumference of 50 cm, height of 93 cm, and the ratio of upper segment to the lower segment 1.1:1.

Verrucous hyperpigmented lesions in patches, whorls and linear streaks were present confined to the left side of the body, including the trunk and limbs (Fig. 1). Examination of the cardiovascular system revealed an ejection systolic murmur in the pulmonary area. Both heart sounds were normal and there was no cardiomegaly. Examination of the central nervous system revealed below average intelligence, limitation of lateral movement of the left eye, nystagmus of both eyes with normal fundus. Motor system examination showed normal

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Fig. 1. Verrucous hyperpigmented lesions confined to the left side of the body.

tone with exaggerated deep tendon reflexes and plantar flexure response. Examination of other systems showed normal findings.

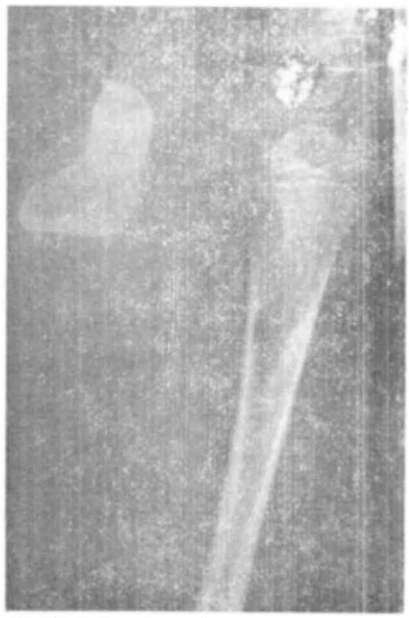

Fig. 2. Cystic lesion in the lower end of right femur.

His haemoglobin was 10.6 gm%, total and differential counts, peripheral blood smear, urine examination, X-ray chest and E.C.G. were normal. A skeletal survey showed cystic changes at the lower end of the right femur (Fig. 2). Other long bones and spine were normal. Skin biopsy from a verrucous lesion showed hyperkeratotic epidermis and papillomatosis and acanthosis.

Comments

Reports on epidermal nevus syndrome are rare. Micro-cephaly and delayed mile-stones indicate CNS abnormalities, along with features of the verrucous nevus.

Correlation between the different manifestations is not clearly understood. The inheritance could be by an abnormal dominant gene with very low penetrance.[2] Extensive studies on epidermal nevus have been done by Solomon et al.[2-4] Diamond and Amon[5] suggest that patients with epidermal nevus must be observed over an extended period of time for the development of earlier than anticipated neoplasms. Deep shave excision or deep dermabrasion may ablate the skin lesions permanently.[2]

References

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