A Case of Epidermal Nevus Syndrome

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We report a case of epidermal nevus syndrome. The patient was a 26-year-old female who had numerous linear verrucous plaques on her neck, upper back and anterior chest. Biopsy of these lesions revealed epidermal hyperkeratosis, acanthosis and papillomatosis. Variable sized cafe-au-lait spots were scattered around the verrucous nevi. The other associated findings were claw hand deformity and epilepsy. Electroencephalogram showed very irregular, random and slow waves confined to the right temporooccipital area.

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Epidermal nevus syndrome was originally described by Solomon and his associates[1] in 1968. This syndrome is characterized by a high incidence of congenital skeletal and central nervous system abnormalities with epidermal nevi.[2] The presence of extenisive epidermal nevi should be considered an indicator for a careful history and search for other organic anomalies. From the time of the original description, many additional case reports have been published.[3-7]

REPORT OF A CASE

A 26-year-old female was referred to our hospital for treatment of multiple verrucous plaques which were present from birth. The family history was non-contributory. At two years of age, she began having focal seizures.

Physical examination revealed extensive linear plaques involving the postauricular area, neck, back and anterior chest(Fig. 1).

Individual lesions were verrucous and yellow-brown in color. Pea to nut-sized cafe-au-lait spots were scattered around the verrucous nevi (Fig. 2). A claw hand deformity of the left hand was also found(Fig. 3). The specimen taken from the anterior chest area showed hyperkeratosis, papillomatosis and acanthosis (Fig. 4).

Routine laboratory studies were within normal limits. The chest and skull roentgenograms were unremarkable. Skull roentgenogram and cranial C.T. scan were normal. However, the electroencephalogram demonstrated very irreg-

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ular, random and slow waves confined to the right temporo-occipital area(Fig. 5).

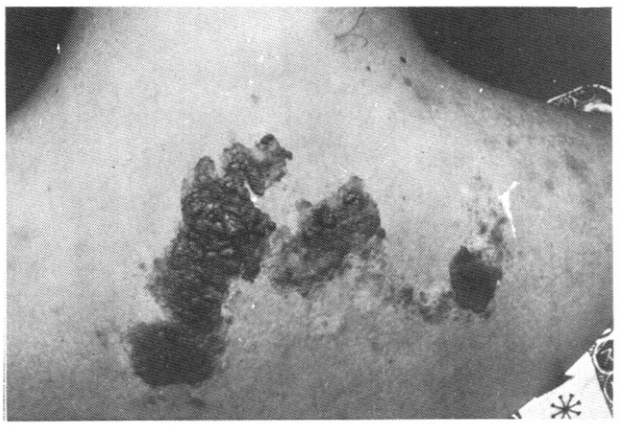

Fig. 1. Multiple, dark brown verrucous lesions on the back.

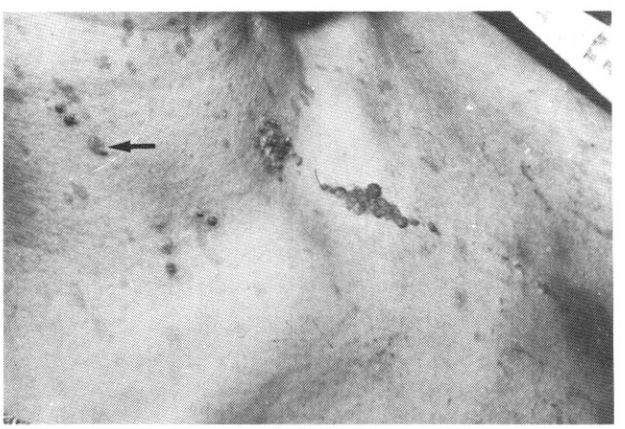

Fig. 2. Cafe-lait spots(➡)on the anterior chest.

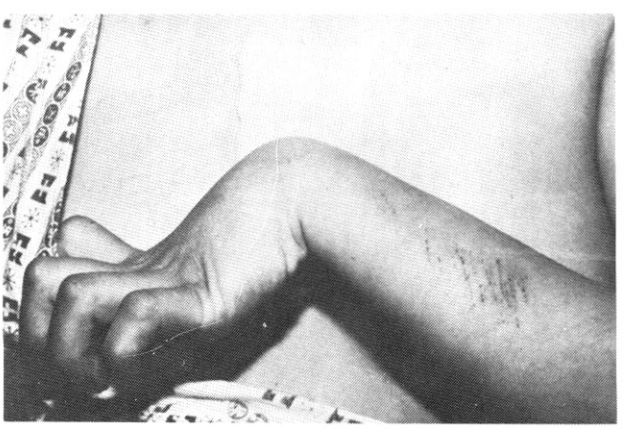

Fig. 3. Claw deformity of the left hand.

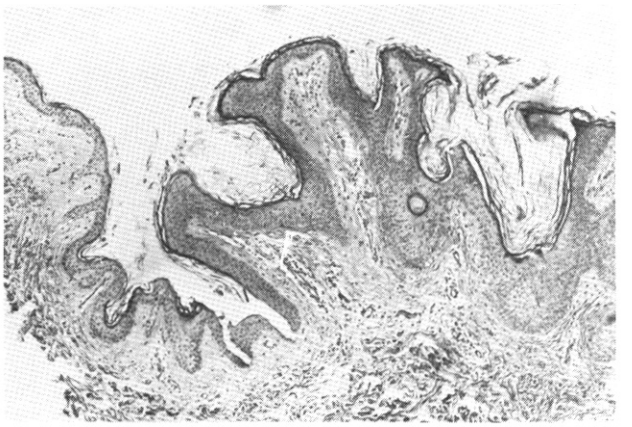

Fig. 4. Histopathologic section from the nevus of the anterior chest hyperkeratosis, acanthosis and papillomatosis(H & E stain, ×100)

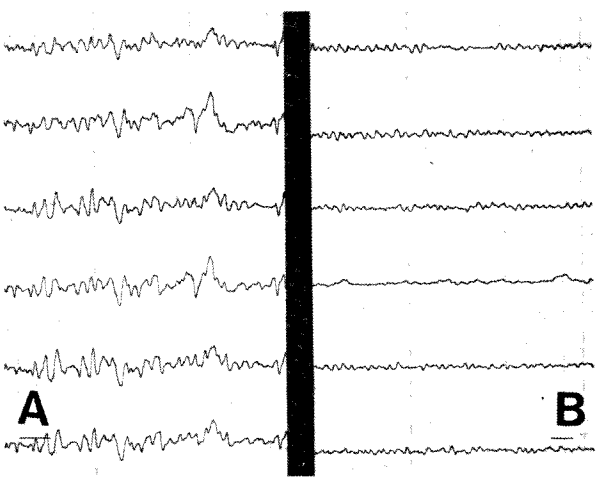

Fig. 5. Electroencephalogram shows very irregular, random, slow and abnormal waves(A) compared to the normal pattern(B).

DISCUSSION

The epidermal nevus syndrome refers to the association of epidermal nevi with anomalies in other organs. It is a rare type of neurocutaneous disorder characterized by verrucous skin rashes of the scalp and neck, seizures with early onset in life, mental retardation, and skeletal, ocular or other organic abnormalities. It has been reported under different names including organoid nevus syndrome,[4] linear sebaceus nevus syndrome[2] and neurocutaneous syn-

drome.[5] However, all epidermal nevi are hamartomas, or proliferations of elements normally found in the basal layer of the embryonic epidermis. Therefore these nevi have been classified according to their prominent component, resulting the terms nevus verrucous(keratinocyte) and nevus sebaceus(sebaceous gland). The generic term "epidermal nevus is used to encompass all these entities, and the term "epidermal nevus syndrome" proposed by Solomon et al[1] describes— the association of epidermal nevi with abnormalities in other organs.

The histogenesis of this tumor is still disputed. Faulty migration and development of embryonic tissus has been implicated because the sebaceus glands and hair follicles fail to migrate away from the epidermis from which they are derived. Others have postulated that the anomaly may be a result of a developmental error in separation of the ectoderm from the neural tube.[8]

The various cutaneous associations of epidermal nevus syndrome have been thor-oughly described by Solomon et al[1] in many individual case reports.[3-9] The most commonly associsted cutaneous lesions are cafe au lait spots, congenital hypopigmented macules and nevocellular nevi. Rarer lesions include nevus flammeus, capillary hemangioma, blue nevi, chondroma, dermatomegaly and nevus hy-pertrichosis.

The most common histopathologic pattern of epidermal nevus is hyperkeratosis, papillomatosis and acanthosis with elongation of rete ridges.[10] Our case showed similar findings. Other findings include psoriasiform, epidermolytic hyperkeratosis, seborrheic keratosis-like and verrucoid nevus comedonicus, acrokeratosis verruciformis-like and mixed histologic patterns.[3, 10]

Rogers et al[3] reported skeletal defects in a series of 131 patients, including genu valgum, clinodactyly, skull asymmetry, microcephaly, calcaneo valgus and intoeing gait. Those anomalies occurred in 15% of the patients. The case presented here had a claw hand deformity, which has not been described in previous reports.

There are numerous reports of neurologic disorders in the epidermol nevus syndrom reviewed by many authors.[1, 2, 3, 9, 11] Solomon et al[1] observed that patients with epidermal lesions affecting the head and face had a higher frequency of CNS involvement ; that is, mental retardation, epilepsy, and hemiparesis. EEG finding of our patent showed very irregular, random, slow and abnormal waves arising form the ipsilateral side of the brain. Other findings reported include hyperkinesia, hypsarrhythmia, nystagmus, hydrocephalus and generalized hypotonia.

Abnormalities in other organ systems are much less common : Ocular, cardiac and renal problems were rarely reported.

There is also evidence that there may be an increased frequency of development of malignancy at young age. The most common associated malignant tumors include Wilm's tumor, nephroblastoma, salivary gland adenocarcinoma, ameloblastoma, transitional cell carcinoma of the urinary bladder and adenocarcinoma of the breast and stomach.[2,16-19]

Appropriate treatment includes early accurate diagnosis of these lesions by histopathologic study and careful physical examination with valuation to define any associated disorders. The various treatment modalities for epidermal nevi include liquid nitrogen, podophylin application, deep shave excision, dermabrasion and surgical excision.[8]

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