Epidermal Nevus Syndrome – A Neuro-cutaneous Marker

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A 13 years old girl presented with linear, slightly raised verrucous nevus present over right half of the body since birth. She presented with seizures at the age of 13 yrs. She had no h/o headache, vomiting, fever and any other focal neurological deficit. Her intelligence quotient was normal.

On dermatological examination, the child showed linear, verrucous lesions over the both upper and lower limbs on the right side of the body (Fig. 1). The lateral aspect of the trunk showed S-shaped verrucous lesions and transverse bands over the anterior and posterior aspect of the trunk (Fig. 2). All the lesions presented along the lines of Blaschko. The nail examination of the right hand and right foot showed ridging, splitting, discoloration and dystrophy. Based on the above clinical findings, it was diagnosed as linear epidermal nevus or otherwise known as nevus unius lateralis. Her EEG showed focal epileptiform activity over the right temperoparietal region which is ipsilateral to the nevus (Fig. 3). MRI brain was normal. X-ray chest, ECG, echocardiogram, ultrasonogram of the abdomen were normal.

Epidermal nevus syndrome (ENS) refers to several disorders that are commonly presenting with epidermal nevus and associated neurological manifestations like seizures or hemimegalencephaly.1 Approximaely 50-80 percent of the patients with ENS have neurological deficit. The location of the nevus appears to correlate with the likelihood of neurological symptoms. The seizures occur in more than fifty percent of the patients with ENS. The EEG abnormalities (focal epileptiform discharges) occur ipsilateral to the nevus.2 This case highlights the importance of side of skin lesion and the laterality of seizure focus.

References