PAEDIATRIC DERMATOLOGY

EPIDERMAL NEVUS SYNDROME ASSOCIATED WITH NEUROLOGICAL, SKELETAL AND CUTANEOUS INVOLVEMENT WITH GRAFT OF GIANT COMMON WART

F Kettani [1] - F Hali [1] - K Baline [1] - S Chiheb [1]

Chu Ibn Rochd, Department Of Dermatology's Diseases, Casablanca, Morocco [1]

Background: Epidermal nevus syndrome (ENS) describes a heterogeneous group of congenital disorders characterized by the presence of epidermal nevi associated with systemic involvement, in particular the central nervous system, eye and skeleton. We report a new case.

Observation: An 11-year-old boy, born of a non-consanguineous marriage, had an extensive epidermal nevus on the right side of the face, neck, trunk and upper right limb from birth. Since the age of 8 months, he was followed for rebel epilepsy with an average of 3 seizures a week. The clinical examination also revealed poliosis, dyschromic lesions of the knees, lumbar hyperlordosis and mental retardation. A hyperkeratotic plaque measuring 10 cm long axis was noted in front of the right ear. The histological study confirmed the diagnosis of epidermal nevus with common wart. In front of the neurological symptomatology as well as the facial and cervical localization of the nevus, a cerebral CT, a MRI and an electroencephalogram were realized revealing no anomalies. The rest of the malformation report was normal.

Key message: We report a new case of ENS associating an extended right hemicorporeal epidermal nevus with neurologic, skeletal and cutaneous involvement. ENS is a sporadic neuroectodermal disorder in which epidermal nevus may be present at birth or appear in infancy and may be accompanied by various central nervous system, bone, and eye abnormalities. Neurological involvement occurs in one-third to one-half of cases, especially in the case of facial or cervical localization of nevus at type of mental retardation, convulsions, ventricular dilation, hemiparesis, leptomeningeal hemangioma, cranial pair involvement or brain tumor. It has been reported that seizures are often difficult to control with sometimes non-contributive brain imaging. The incidence of development of common warts on epidermal nevus is currently unknown.