GENETICS AND GENODERMATOSES

COMEDONIC NEVUS SYNDROME: CASE REPORT.

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Background: Comedonic nevus syndrome is a rare entity with few reported cases, characterized by the presence of comedonic nevus in association with extracutaneous manifestations. We report a case of Comedonic Nevus Syndrome, with exuberance of cutaneous lesions and congenital cataract.

Observation: An 15-year-old girl presented to us with lesions similar to comedones from birth, congenital cataract and visual acuity decrease. Dermatological examination characterized by grouping of comedones arranged in plates and linearly on the face, upper trunk, upper and lower limbs, following Blaschko lines. Dermatoscopic examination shows follicular openings filled by keratin plates. Anatomopathological examination of the dorsal lesion was compatible with comedonic nevus. We opted for clinical follow-up and multidisciplinary follow-up with ophthalmologist and neurologist.

Key message: Comedonic nevi is characterized by a rare, poorly diagnosed entity that can occur as an isolated or combined lesion with other congenital malformations. The individual lesion or syndrome presents as a grouping of comedones, arranged linearly or in plaque, from small centimeters until involvement of extensive body surface, can be uni or bilateral and systematized. Histologically characterized by elongated and dilated follicular infundibulum. The main extracutaneous manifestations are skeletal, ophthalmologic, and central nervous system. The usual course is persistent and unchanged, and may occur with periods of secondary infection. The treatment is ineffective, being therapeutic options proposed dermabrasion, manual extraction of comedones, the application of topical keratolytic agents, surgical treatment - depending on the size of the lesion. Therapeutic abstention remains the most common attitude.