A Picture Says a Thousand Words

Hairless verrucous growth over scalp

Madhab Datta MBBS, Sayantani Chakraborty MD, Abheek Sil MBBS[®]

Department of Dermatology, Venereology, and Leprosy, R.G.Kar Medical College, Kolkata, West Bengal, India

Correspondence: Abheek Sil, Department of Dermatology, Venereology, and Leprosy, R.G.Kar Medical College, 1, Khudiram Bose Sarani, Kolkata 700004, West Bengal, India. E-mail abheek.sil@gmail.com

CASE DESCRIPTION

An 8-year-old boy presented with an asymptomatic yellow-brown coloured growth over the scalp since birth. The lesion progressed gradually, proportionate to the child's growth, from a small raised tan-coloured area to its current presentation. He had no history of birth trauma. Family history was non-contributory. Local examination revealed a solitary, annular, alopecic, tan-brown coloured verrucous plaque, measuring 6 cm × 4 cm, with a velvety surface on the left parieto-occipital region (Figure 1). Localized alopecia restricted to the plaque was evident. Remaining mucocutaneous and systemic examinations were unremarkable.

What is your diagnosis?

Figure 1. Annular tan-brown coloured verrucous plaque with velvety surface present over scalp (note the absence of hair within lesion).

DIAGNOSIS

Based on clinical features, a diagnosis of nevus sebaceus (NS) was established. His parents were counselled about the usual benign course of the condition but were also cautioned about the need for regular follow-ups to assess any secondary benign, or rarely, malignant transformation.

DISCUSSION

NS is a hamartoma with epithelial and non-epithelial origins. As it may contain any or all components of the skin, it is often referred to as an organoid nevus. Histologically, NS displays an array of abnormalities within sebaceous glands, sweat glands, and hair follicles, often with a lack of terminally differentiated hair follicles. At birth, lesions are only slightly raised and subtly discernible and commonly involve the scalp, face, or the neck occasionally. They follow a linear configuration along the lines of Blaschko, although it is difficult to appreciate when the size is small. On the scalp, it remains as an alopecic, slightly raised yellow-orange or tan-coloured plaque, with normal hair growth surrounding the lesion. With age, the nevus thickens slightly. At adolescence, due to hormonally driven sebaceous gland hyperplasia and maturation of apocrine glands, pubertal expansion characterized by progressive thickening occurs and the surface becomes pebbly or verrucous (1).

NS syndrome, on the other hand, is a neurocutaneous condition characterized by one, normally extensive, or multiple sebaceous nevi, along with associated systemic involvement like ocular abnormalities (e.g., coloboma and choristoma), epilepsy, cognitive delay, hypophosphatemic rickets, genitourinary and cardiovascular defects, and SCALP syndrome (sebaceous nevus syndrome, CNS malformation, aplasia cutis congenita, limbal dermoid, and pigmented nevus). Although rare, benign

and malignant (in 10% to 20% of cases) transformation including but not limited to trichoblastoma, syringocystadenoma papilliferum, trichilemmoma, desmoplastic trichilemmoma, sebaceous adenoma, apocrine adenoma, eccrine poroma, adnexal carcinomas and even basal cell carcinoma should be observed for (2).

Clinical distinction should be made from verrucous epidermal nevus (onset at birth/early childhood; verrucous, skin-coloured, dirty-grey or brown papule or plaque with preserved hair growth) and nevus comedonicus (closely arranged, grouped, often linear papules that have at their centre dilated follicular openings with keratinous plugs resembling comedones).

Although typically a clinical diagnosis owing to its characteristic clinical appearance and location, NS may be confirmed with a biopsy (in addition to rule out suspicion of neoplastic change). Observation and close follow-up are the ideal recommendations.

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