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Case Report

Nevus sebaceus of Jadassohn: an unusual case report with review of literature

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ABSTRACT

A 16-year-old girl presented with a new, rapidly growing, raised, hypopigmented, cerebriform plaque on the scalp. The skin lesion was brown coloured and with a waxy aspect. The diagnosis 'nevus sebaceus' was established. Nevus sebaceous of Jadassohn is a hamartoma with a combination of abnormalities of the epidermis, hair follicles, and sebaceous and apocrine glands. Herein, we report a case along with detailed review of literature of this unusual disease.

Keywords: Nevus sebaceus of Jadassohn, Skin tumor, Benign

INTRODUCTION

Nevus sebaceus of Jadassohn (NSJ) is a relatively uncommon, predominantly sebaceous hamartoma, with an estimated incidence of less than 1/100,000.[1] NSJ presents as a waxy, hairless, verucous plague located on the scalp, face, or neck. Historically, there is an association of bacel cell carcinoma arising within NSJ.[2] NSJ was thought to be caused by sporadic genetic mutations until 1982, when Monk et al presented the first report of a genetic association of NSJ in a mother and daughter.[3]

CASE REPORT

A sixteen-year-old girl presented to the dermatology OPD with complaints of a new, rapidly growing, raised, hypopigmented, cerebriform plaque over the scalp (Figure 1). Biopsy from the growth was performed and sent to us for histopathological examination. We received a single, skin covered, grey-brown tissue bit measuring 0.8cm. Microscopically, H & E stained sections studied showed epidermis and dermis. Epidermis was lined by stratified squamous epithelium showed papillomatous hyperplasia (Figure 2). Underneath, dermis shows multiple mature sebaceous glands and dilated infundibula of hair follicles (Figure 3). Thus a diagnosis of Nevus sebaceus of Jadassohn was made. The patient did not opt for surgical excision of the lesion and is asked to be on regular follow-up to look for any malignant change in the lesion.

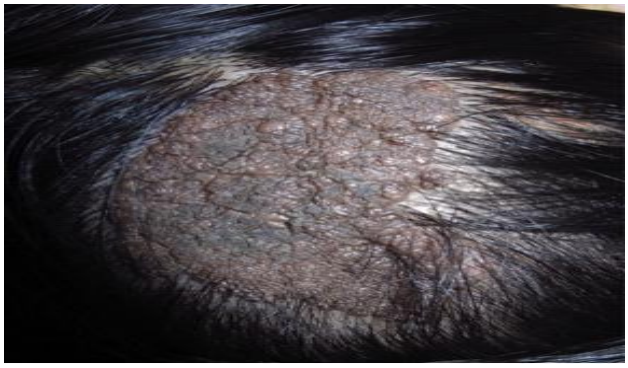

Figure 1: Clinical photograph of nevus sebaceus on scalp.

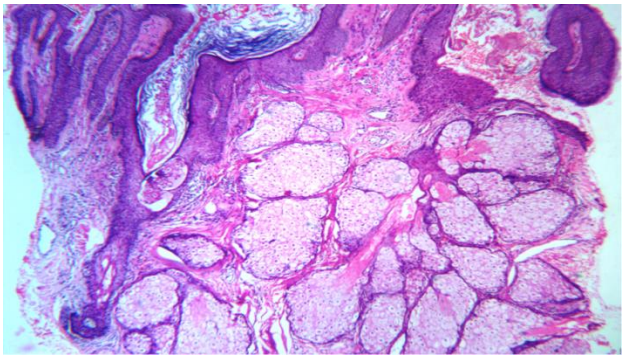

Figure 2: Low power magnification of the biopsy, H&E stain.

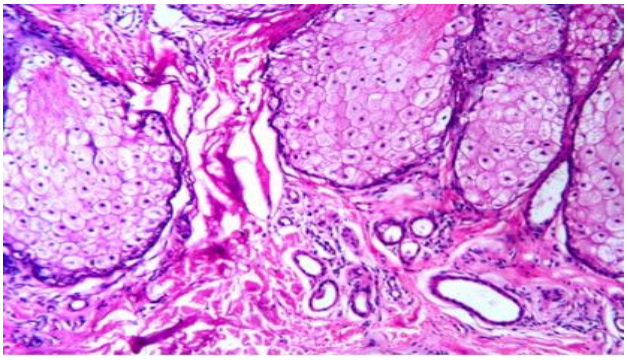

Figure 3: Higher magnification of the biopsy, H&E stain.

DISCUSSION

Certain patients with extensive nevus sebaceus may also develop epilepsy, mental retardation, other neurologic defects, or skeletal deformities, causing a neuroectodermal syndrome termed shimmel penning or nevus sebaceus syndrome.[4]

There is significant overlap with keratinocytic epidermal nevi and the epidermal nevus syndrome, and it is likely that these syndromes represent different phenotypes arising from the same underlying mutations in HRAS and KRAS.[5]

Previously it was held that the basaloid neoplasms in nevus sebaceus represented basal cell carcinoma. However, this view was revised in several critical reviews stating that most of these basal cell carcinomas actually represented trichoblastomas.[6,7]

Interestingly, the majority of basaloid tumors arising in nevus sebaceus are negative for PHLDA1, a follicular stem cell marker. PHLDA1 is generally positive in trichoepithelioma and trichoblastoma and negative in primary basal cell carcinoma. These findings indicate glycogen as an indication of pilar differentiation.[8]

Other malignant tumors that are known to occur include apocrine carcinomas, malignant eccrineporoma, keratoacanthoma, proliferating trichilemmal tumor, leiomyosarcoma, microcystic adnexal carcinoma, sebaceous carcinoma, and mucoepidermoid carcinoma.[9-17]

CONCLUSION

Although rare, NSJ should be kept in the differential diagnosis of all scalp lesions and close follow up with detailed family history should be procured to detect high risk groups and malignant transformation.

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