Case Reports

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Basal cell carcinoma, papillary syringocystadenoma, apocrine adenoma and trichilemmoma on nevus sebaceous of Jadassohn

Carcinoma basocelular, siringocistoadenoma papilífero, adenoma apócrino e triquilemoma sobre nevo sebáceo de Jadassohn

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ABSTRACT

Described by Jadassohn and also known as organoid nevus, nevus sebaceous is considered a hamartoma that exhibits follicular, sebaceous, eccrine and apocrine malformations of varying degrees. Between 10% and 30% of patients with sebaceous nevi of Jadassohn are at risk of developing cutaneous or adnexal neoplasia during adulthood. The authors describe the case of a patient with nevus sebaceous of Jadassohn associated with multiple tumors (benign and malignant) of different strains, highlighting the importance of the dermatologist physician's knowing this entity and how to perform an examination of the scalp.

Keywords: Carcinoma, Basal Cell; Hamartoma; Neoplasms; Nevus sebaceous of Jadassohn

RESUMO

Descrito por Jadassohn e também conhecido como nevo organoide, o nevo sebáceo é considerado hamartoma que exibe má-formação folicular, sebácea, écrina e apócrina de graus variados. Durante a idade adulta, de dez a 30% dos pacientes com nevo sebáceo de Jadassohn têm risco de desenvolver neoplasia cutânea ou anexial. Relatamos caso de paciente com nevo sebáceo de Jadassohn associado a múltiplas neoplasias (benignas e malignas) de diferentes linhagens e ressaltamos a importância do conhecimento dessa entidade e do exame do couro cabeludo por parte do dermatologista.

Palavras-Chave: *Carcinoma basocelular; Hamartoma; Nevo sebáceo de Jadassohn; Neoplasias*

INTRODUCTION

Originally described by Jadassohn in 1895, it was only in 1932 that it was termed "sebaceous nevus", which was introduced by Robinson.[1,2] This condition is a relatively prevalent congenital hamartoma that classically develops through phases of growth and maturation, exhibiting follicular, sebaceous, eccrine and apocrine malformations of varying degrees.[3,4] Several HRAS and KRAS activating mutations have been reported in sebaceous nevi, allowing some authors to see them as a result of a proliferative state of total skin somatic mosaic.[4]

During adulthood, 10% to 30% of patients with nevus sebaceous of Jadassohn (NSJ) are at risk of developing cutaneous or adnexal neoplasia.[3,4]

The authors of the present case report a case of patient bearing NSJ associated to multiple neoplasms (benign and ma-

lignant) of different lineages, emphasizing the importance of knowing this condition and of undergoing examination of the scalp by a dermatologist.

CASE REPORT

A 65-year-old female patient complained of pruritus on the scalp for a month. On examination, it was possible to observe in the occipital region: a slightly erythematous verrucous plaque measuring around 2.5 cm on its longest axis (Figures 1 and 2). According to the patient, she bore an asymptomatic lesion on the scalp since birth, and that had been pruritic for a month. The authors decided to carry out an incisional biopsy of the lesion, which was sent to histological examination, evidencing a nodular basal cell carcinoma (Figure 3). The complete excision of the lesion was performed with safety margins. The histology revealed a sebaceous nevus associated with syringocystadenoma papilliferum, apocrine tubular adenoma, trichilemmoma (Figures 4 to 7), and scarring dermal fibrosis.

DISCUSSION

The NSJ, also known as organoid nevus, emerges more frequently in the scalp, however it can arise in the face and, less commonly, in the limbs.[3,5] It occurs in approximately 0.3% of individuals, with no gender preference. The lesion is usually present at birth and has the appearance of a well-defined plaque composed of multiple confluent yellow-orangish or yellow-brownish papules, predominantly in the scalp, where it progresses with alopecia at the site of the lesion.[3] It has a bimodal distribution: during puberty its surface becomes thickened and verrucous due to hormonal stimuli to the eccrine and apocrine components, while in adulthood the lesion may become nodular with the occurrence of ulcerations and crusts. The possibility of

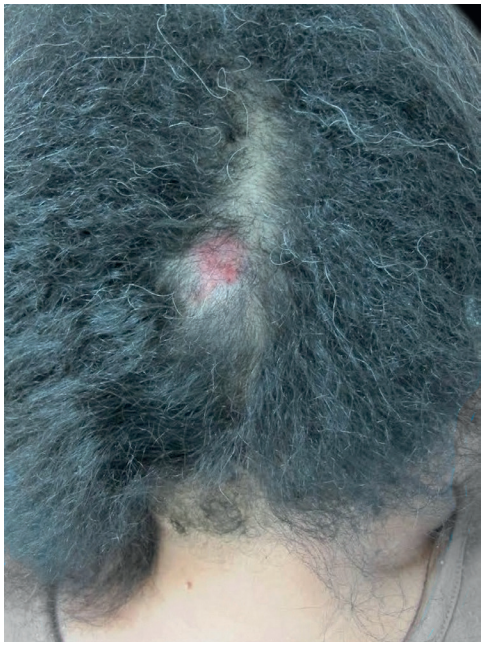

Figure 1: Occipital region: slightly erythematous verrucous plaque

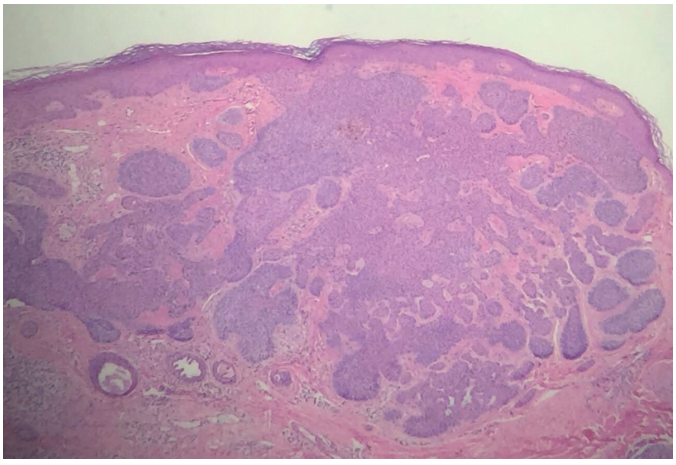

Figure 3: Basaloid epithelial proliferation forming blocks with peripheral palisade, compatible with basal cell carcinoma (Hematoxylin & eosin x40)

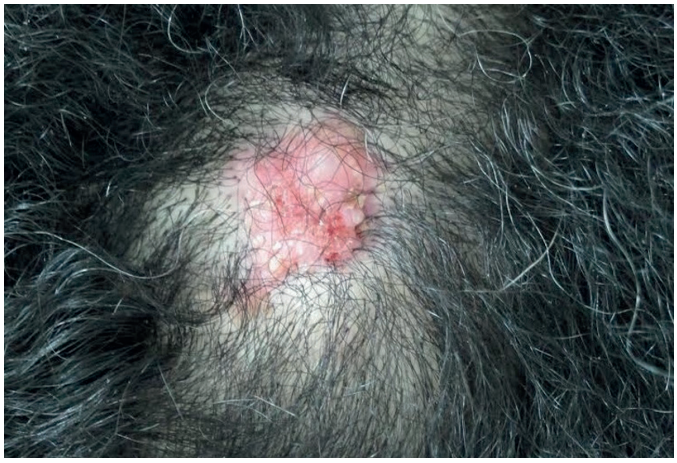

Figure 2: Occipital region: detail of the lesion

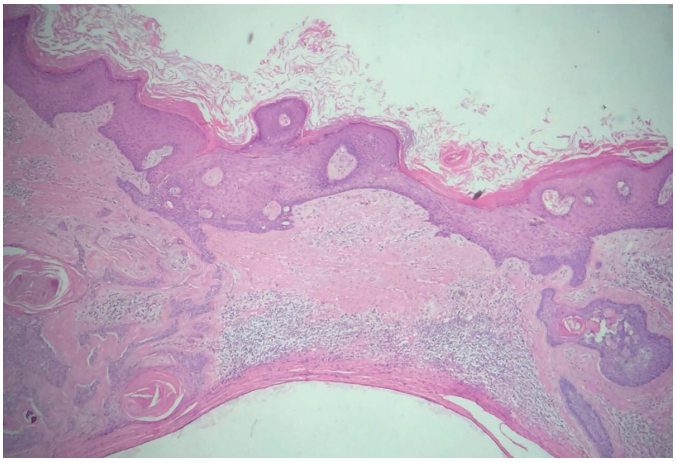

Figure 4: Cutaneous papillomatosis with hyperkeratosis and acanthosis, compatible with sebaceous nevus (Hematoxylin & eosin x100)

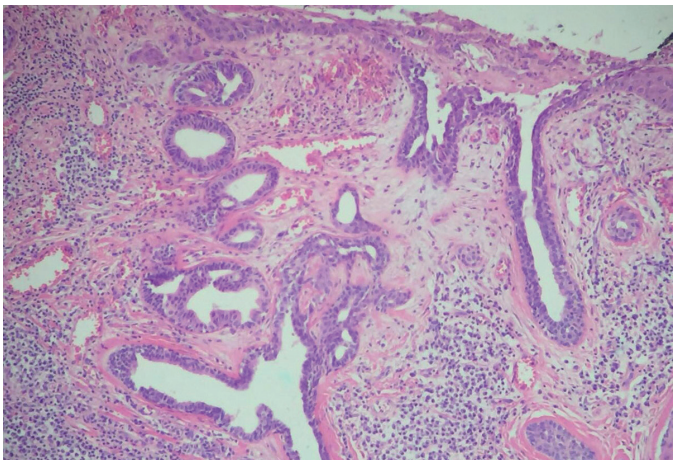

Figure 5: Glandular epithelial proliferation, compatible with syringocystadenoma papilliferum (Hematoxylin & eosin x100)

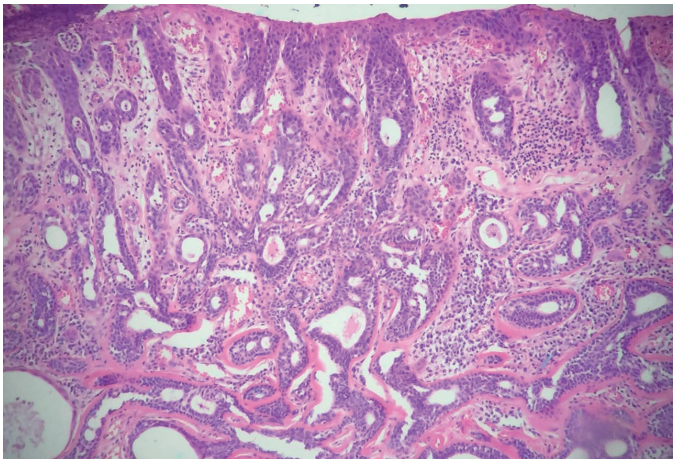

Figure 6: Glandular epithelial proliferation with characteristics typical of apocrine adenomatous lesion (Hematoxylin & eosin x40)

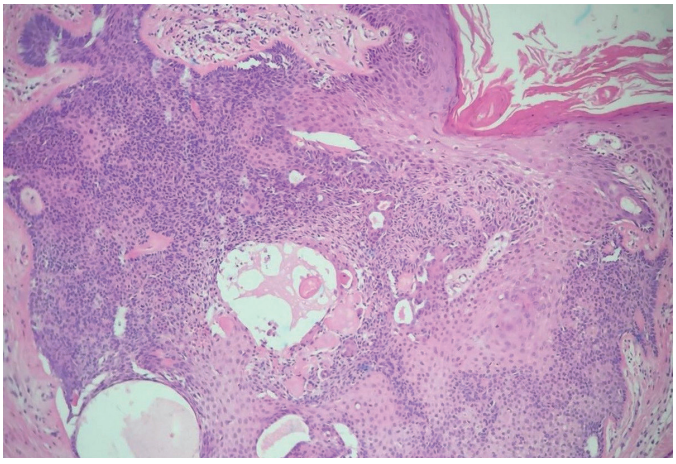

Figure 7: Basaloid epithelial proliferation connected to the epidermis, with presence of corneous cysts and clear cell's trichilemmoma-like components (Hematoxylin & eosin x100)

secondary neoplasms in this phase ranges from 10% to 30%, the main ones being basal cell carcinoma, papillary syringocystadenoma (both observed in the patient described in the present report), and trichoblastoma.[3]

Other tumors already described in association with NSJ include benign ones –trichilemmoma (also present in the described patient), trichoadenoma, nodular hidradenoma, apocrine hidrocystomas, syringoma, apocrine nevus, poroma, spiradenoma, keratoacanthoma, piloleiomyoma, osteoma, melanocytic nevus, seborrheic keratosis and keratoacanthoma; [4,6-8] and malignant ones – squamous cell carcinoma, sebaceous carcinoma, apocrine carcinoma, leiomyosarcoma, eccrine porocarcinoma, and melanoma.[6,9]

There is no consensus on the ideal approach. Some authors recommend early surgical excision (pre-pubertal) aimed at preventing malignant and aesthetically disfiguring transformations. Others, however, advocate a more conservative behavior. Future studies might identify molecular markers or genetic alterations that could indicate a greater risk of neoplastic transformation, thus avoiding unnecessary surgical interventions.[10]

The diversity of tumors of different lineages detected in a single lesion motivated the authors to prepare this report emphasizing the importance of the knowledge and development of this entity (NSJ), and calling attention to the relevance of the examination of the scalp during dermatological consultations. ●

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