

## BRIEF ARTICLES

**Coexisting Basal Cell Carcinoma and Squamous Cell Carcinoma in Congenital Nevus Sebaceous**Jordan Rosen BS<sup>a</sup>, Katherine Nolan MD<sup>a</sup>, Noah Shaikh BS<sup>a</sup>, Les Rosen MD<sup>a</sup>, Martin Zaiac MD<sup>a</sup><sup>a</sup>Department of Dermatology and Cutaneous Surgery, University of Miami Miller School of Medicine, Miami, FL**ABSTRACT**

Nevus sebaceous is a congenital epidermal hamartoma characterized by hyperplastic changes to the epidermis and adnexa. Nevus sebaceous is associated with an elevated risk of cutaneous neoplasms, most often benign; however, malignant neoplasms, most notably basal cell carcinoma, can also present in these patients. Although a rare occurrence more often affecting adult patients, squamous cell carcinomas have also been reported to arise at the site of pre-existing nevus sebaceous. Herein we report a unique case of a patient with basal cell carcinoma and squamous cell carcinoma arising concurrently in the same nevus sebaceous.

**INTRODUCTION**

Nevus sebaceous (NS) is a congenital epidermal hamartoma that is present in less than 1% of newborns.<sup>1</sup> NS appears classically on the face or scalp as a waxy, yellow- or orange-colored linear or round plaque. Lesions can involve the epidermis, hair follicle, sebaceous glands, and apocrine glands. NS frequently increases in size during puberty.<sup>2</sup> As patients age, there is an increasing risk for the development of cutaneous neoplasms within NS.<sup>3</sup> The vast majority of NS-associated neoplasms are benign. Malignant neoplasms, most notably basal cell carcinoma (BCC), represent less than 1% of the cutaneous neoplasms arising from NS, and frequently affect those over the age of twenty.<sup>3,4</sup> We report the fourth case to our knowledge of BCC and SCC arising concurrently in the same NS.

**CASE REPORT**

A 56-year-old Hispanic female with no significant medical history presented to clinic regarding a lesion on her left cheek (Figure 1). The lesion had been present since birth but had begun to grow over the last several months. The patient also noted mild discomfort at the site of the lesion beginning around the same time that the lesion started to grow. Family history was negative for any skin cancers or other malignancies.

On examination there was a well-defined 1.4-cm x 0.8-cm pink, translucent, pearly ovoid plaque with arborizing telangiectasias overlying the mandible on the left cheek. Two biopsies were taken from the lesion edges and were sent for pathological examination. The first biopsy specimen

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revealed acantholysis, papillomatosis, dilated apocrine glands, large sebaceous lobules, and a lack of terminal hairs, all of which are consistent with NS (Figures 2-3). The second biopsy demonstrated nodular basaloid tumor nests, with a peripheral palisading pattern, fibromyxoid stroma, and focal retraction. Additionally, the second biopsy showed a prominent surrounding inflammatory infiltrate with eosinophils and peripheral trapping of elastic fibers. The findings in the second biopsy were

congruent with a superficial type BCC in association with a SCC (Figures 4-5).

Given the location of the lesion and the malignant features, the patient underwent Mohs surgery for complete removal of the NS. Pathological evaluation of the surgical specimen confirmed clear margins. At 6-month follow-up the surgical site is now well healed with no evidence of reoccurrence.



Figure 1. A 1.4x0.8cm pink, translucent, pearly, ovoid papule with arborizing telangiectasias involving the left cheek

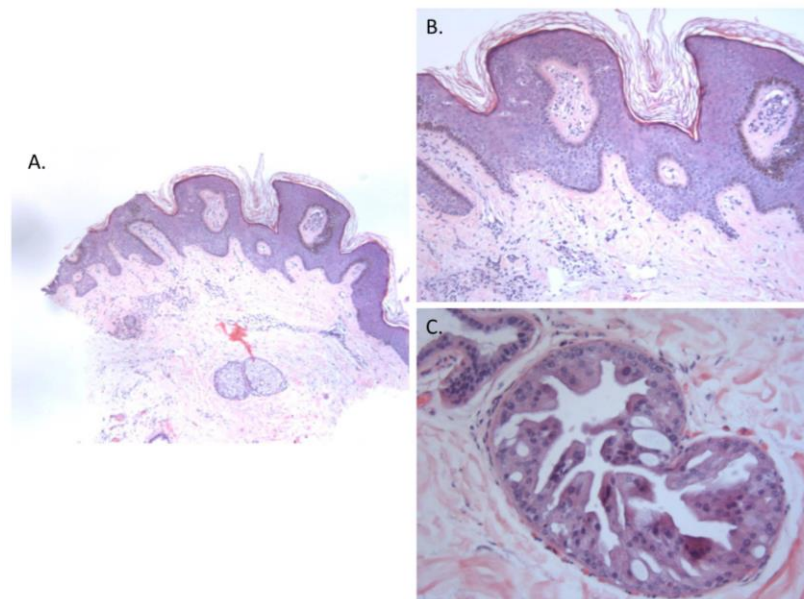
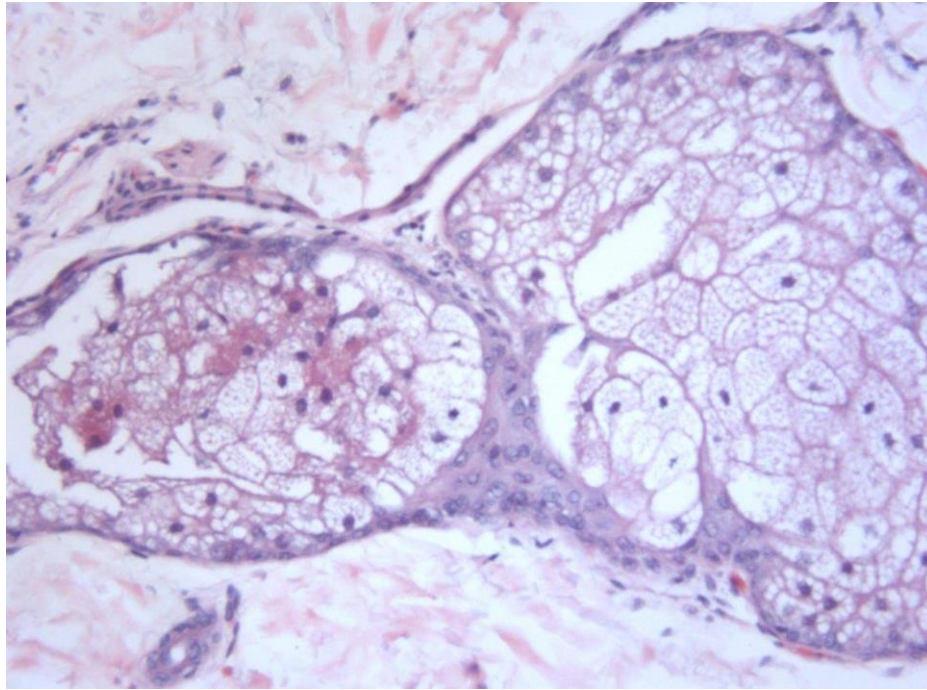
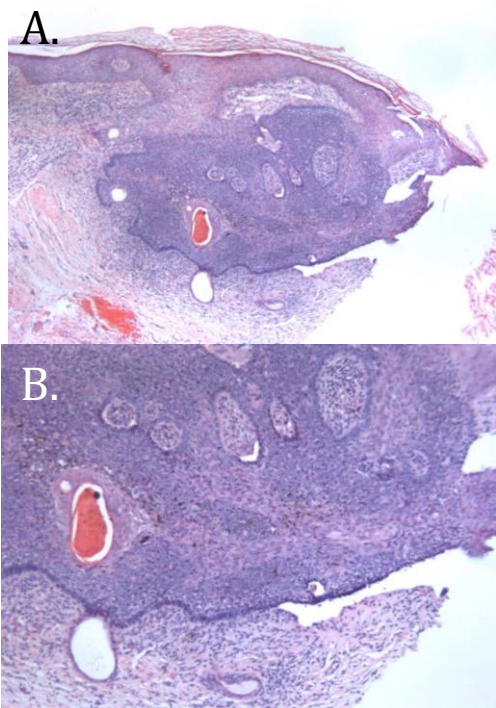


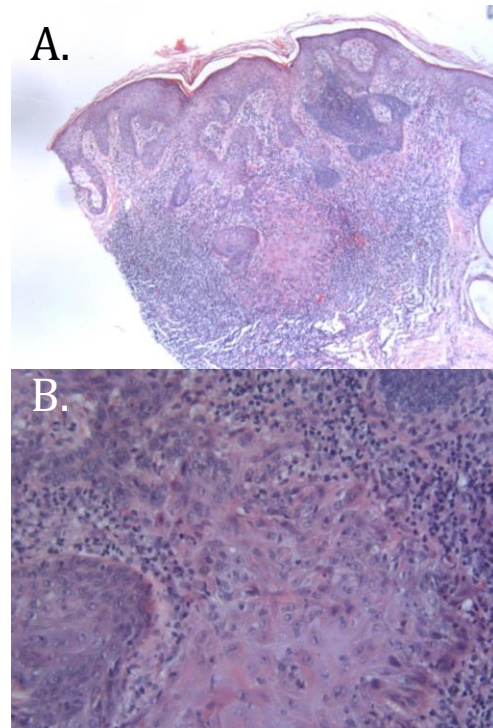
Figure 2. Classic features of nevus sebaceus are seen, including papillomatosis and acanthosis. (A) H&E 5x (B) H&E 10x (C) H&E 20x



**Figure 3.** Apocrine glands and large sebaceous lobules are present throughout the dermis (H&E 20x)



**Figure 4.** Within the nevus sebaceous there are characteristic findings of BCC with nodular basaloid tumor nests, peripheral palisading, fibromyxoid stroma and focal retraction artifact (A) H&E 5x (B) H&E 10x



**Figure 5.** There is SCC characterized by proliferation of atypical keratinocytes with glassy cytoplasm extending into the reticular dermis in association with an inflammatory cell infiltrate consisting of eosinophils, lymphocytes, and histocytes (A) H&E 5x (B) H&E 20x

## DISCUSSION

Although NS is considered a benign congenital lesion, there are important associations and considerations to take when caring for such patients. NS is classically thought to progress through three clinical stages as patients age. The first stage occurs throughout childhood and presents with a smooth yellow- or orange-colored plaque characterized by limited hair or sebaceous glands. The second stage, occurring during puberty and likely related to hormonal changes, is associated with a more verrucous appearing lesion and maturation of apocrine and sebaceous glands. Finally, the last stage is associated with the presence of benign and malignant cutaneous neoplasms.<sup>5</sup>

Ambiguity regarding NS and the risk of future malignancy has made it difficult for physicians to determine when to recommend surgical excision versus continued observation with follow-up. NS are often found on the scalp and, unsurprisingly, tumors arising from within NS occur more frequently on the scalp.<sup>6</sup> The most common tumor types that are found to arise within NS are benign; more specifically, between 90% and 97.5% of tumors developing within NS are benign.<sup>2,3</sup> Trichoblastoma and syringocystadenoma are the most common neoplasms that occur in NS.<sup>2,3</sup> The most common malignant tumor associated with NS is BCC, followed by SCC.<sup>2</sup> The mean age of patients presenting with BCC or SCC is 54 and 49 years of age, respectively.<sup>2</sup>

Additionally, there have been three similar cases of BCC and SCC arising concurrently within an NS. Unlike this case, which describes a lesion on the mandible, the other three cases describe lesions in other areas. The earliest case was described in

1970 in a 56-year-old woman who presented with an ulcerated lesion on the temple.<sup>7</sup> In 2005, Ball et al. reported a case of BCC and SCC developing from within an NS located on the scalp of a 35-year-old man.<sup>8</sup> Lastly, in 2007, Arshad et al. described a similar case of a 55-year-old man with SCC and BCC arising from a NS on the scalp<sup>9</sup>; despite wide excision and radiotherapy, the patient's SCC metastasized, leading the author to suspect increased aggression of SCC that arises from within NS.<sup>9</sup>

While, in this case report we describe an extremely rare case of simultaneous BCC and SCC in a single lesion of nevus sebaceous, there have been other reports of two distinct malignancies developing in the background of NS.<sup>10</sup> There have been two reported cases of simultaneous SCCs developing within a single NS.<sup>10,11</sup> Although uncommon for children to develop SCC within NS, Belhadjali et al. reported a case of an 11-year-old patient with NS containing two simultaneous SCCs in different stages of invasion.<sup>11</sup> In that case the lesion was excised and was not reported to have re-occurred.

In conclusion, this case describes the rare occurrence of simultaneous BCC and SCC in a single lesion of NS. While basaloid and adnexal neoplasms are most commonly thought of in relation to NS, this case highlights that SCC can also occur and be associated with more aggressive behavior. The association between BCC, SCC, and NS may also offer insight into the underlying pathogenesis of these three neoplasms.

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