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Sebaceous Carcinoma, Syringocystadenoma Papilliferum, and Sebaceoma Arising in a Nevus Sebaceus of Jadassohn: A Case Report and Review of the Literature

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ABSTRACT

Nevus sebaceus (NS), also known as nevus sebaceus of Jadassohn, is a congenital hamartoma of the pilosebaceous unit, most commonly located on the head and neck. Its natural course is characterized by progressive morphological changes and the potential development of secondary tumors, which are usually benign and more rarely malignant, particularly in adulthood.

We report the case of a 45-year-old man with a congenital nevus sebaceus of the scalp complicated by the exceptional association of three secondary tumors: syringocystadenoma papilliferum, sebaceoma, and sebaceous carcinoma. The patient presented with a recently enlarging, mildly painful, and oozing nodule arising within the pre-existing lesion. Clinical and dermoscopic examination revealed features suggestive of malignant transformation. Histopathological analysis confirmed the diagnosis of sebaceous carcinoma associated with syringocystadenoma papilliferum and sebaceoma within the excision specimen. Immunohistochemical staining for mismatch repair proteins showed preserved expression, ruling out Muir-Torre syndrome. At 12-month follow-up, there was no evidence of local recurrence or metastatic disease.

To our knowledge, this is the first reported case of the simultaneous occurrence of sebaceous carcinoma, syringocystadenoma papilliferum, and sebaceoma within a single nevus sebaceus. This case underscores the importance of lifelong surveillance of nevus sebaceus and provides dermoscopic clues that may aid in the early detection of malignant transformation.

KEYWORDS :

nevus sebaceus; sebaceous carcinoma; syringocystadenoma papilliferum; sebaceoma; dermoscopy; Muir-Torre syndrome

MAIN ARTICLE

INTRODUCTION

Nevus sebaceus (NS), or verrucous sebaceous hamartoma, is a malformation that is most often congenital, present at birth in 72% of cases, primarily affecting the pilosebaceous unit [1]. Also known as nevus sebaceus of Jadassohn, this term was first introduced in 1895. With a prevalence of 0.3% among neonates, 95% of these lesions are typically located in the head and neck region [2]. NS is characterized by hyperplasia of the sebaceous glands, apocrine glands, hair follicles, and epidermis, presenting as a raised, flesh-colored or pink-orange verrucous plaque.

During its evolution, NS progresses through three distinct phases. The final phase is marked by the development of various tumors, most commonly benign and rarely malignant. In a landmark study of 707 patients with NS, Idriss and Elston reported that 22.5% of patients developed secondary tumors, with trichoblastoma (7.4%) and syringocystadenoma papilliferum (5.2%) being the most common benign neoplasms, while malignant tumors occurred in only 2.5% of cases [3]. Sebaceous carcinoma arising in NS is exceptionally rare; a comprehensive review by Izumi et al. in 2008 identified only 14 cases in the literature prior to their series of 10 additional cases [4].

We report the exceptional case of a sebaceous carcinoma, a syringocystadenoma papilliferum, and a sebaceoma arising within a scalp NS and emphasize, through this observation, the necessity of regular clinical and dermoscopic surveillance of these lesions to enable early management. To our knowledge, this is the first reported case of the simultaneous occurrence of these three specific tumor types within a single NS.

CASE PRESENTATION

A 45-year-old man with Fitzpatrick skin phototype IV and no significant past medical history presented to the dermatology outpatient clinic for a recent increase in size of a lesion arising on a congenital nevus of the scalp.

During the clinical interview, the patient reported that he had had a yellow-orange alopecic plaque since birth, which had remained stable until puberty. During adolescence, the plaque increased in size, becoming more prominent with a verrucous surface. The recent appearance of a mildly tender nodule that had been progressively enlarging over the past six months, along with the discharge of serosanguinous fluid, prompted the patient to seek dermatological consultation. He had no history of trauma, radiation therapy, burns, or immunosuppressive therapy, and no family history of cutaneous or internal malignancy.

On physical examination, an alopecic plaque of the scalp was noted, pink-orange in color, located in the frontoparietal region and slightly lateralized to the right, measuring 8 × 3 cm, with well-defined borders and a surface that was verrucous in some areas and mammillated in others. At the posterior aspect of the lesion, a round, sessile, firm tumor was identified, measuring approximately 4 × 3 cm, covered with yellowish crusts and a central hemorrhagic area (Figure 1A–B). Palpation of the tumor elicited mild tenderness.

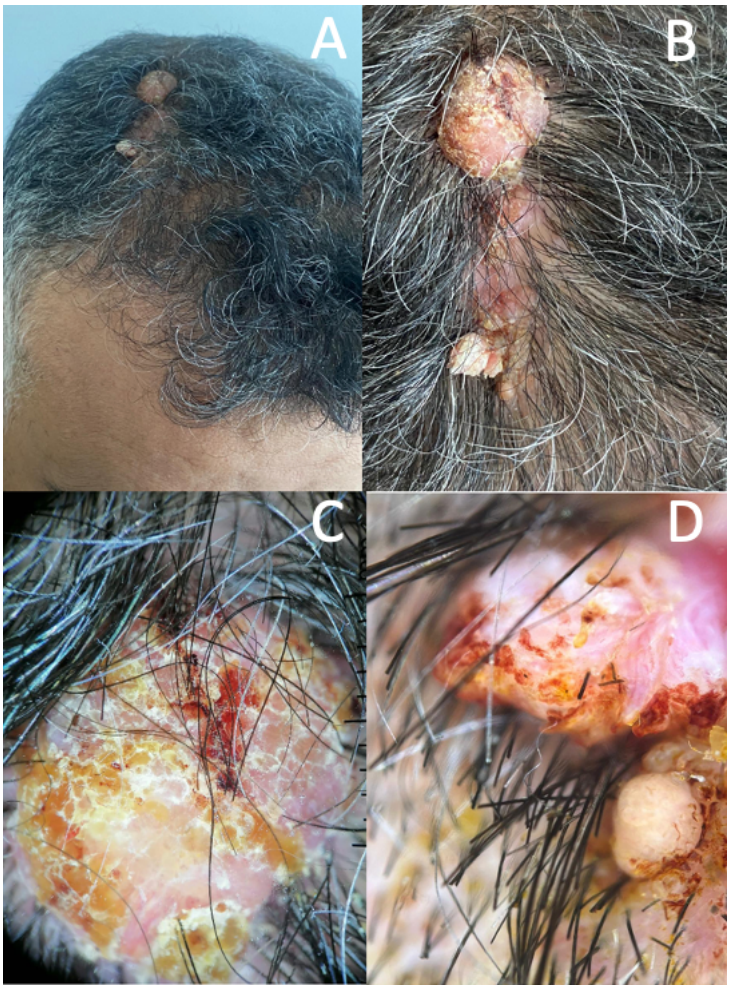

Figure 1: A–B: Clinical photographs showing an orange alopecic plaque with an irregular surface and a posterior round tumor. C–D: Dermoscopic images revealing a pink-orange background, yellowish crusts, hemorrhagic areas, structureless yellow zones, and peripheral irregular linear vessels.

An excisional biopsy of the posterior tumor was performed, revealing a sebaceous carcinoma (Figure 2).

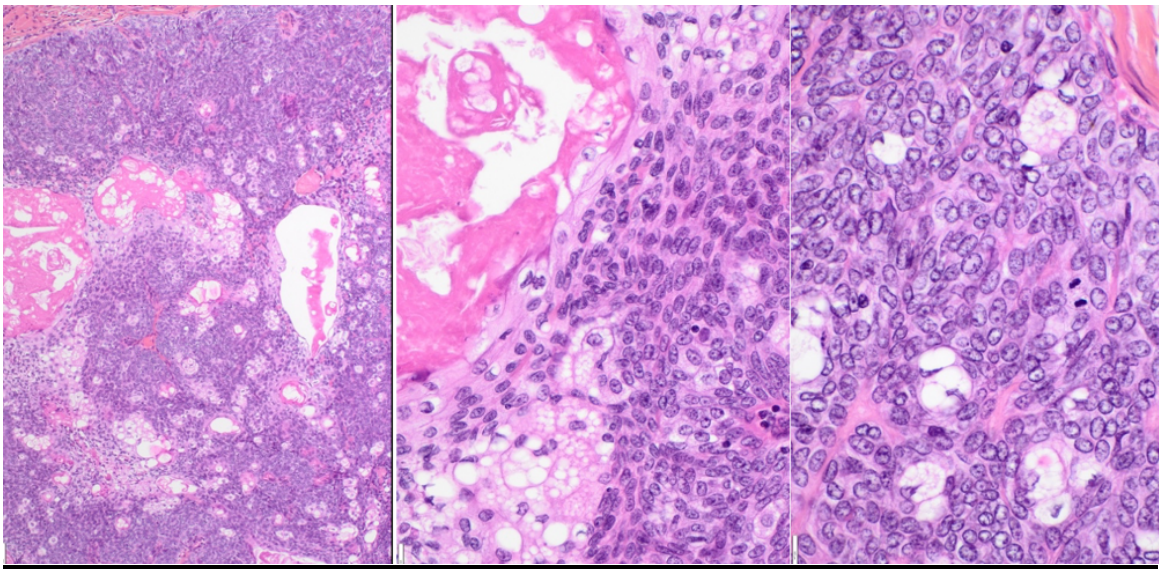

Figure 2: Histopathological examination (H&E stain). A: Low-power view (×40) showing a dermal tumor with lobular architecture arising within a nevus sebaceus. B-C: High-power view (×200) showing large sebocytes with foamy cytoplasm and marked cytonuclear atypia, numerous mitotic figures, consistent with sebaceous carcinoma.

The workup was completed with ultrasonography of the regional lymph node basins and a cranial computed tomography (CT) scan, neither of which showed evidence of locoregional extension. Given the presence of multiple sebaceous neoplasms, Muir-Torre syndrome was considered in the differential diagnosis. Immunohistochemical staining for mismatch repair proteins (MLH1, MSH2, MSH6, PMS2) showed preserved expression, effectively ruling out this diagnosis.

Following these investigations, the patient underwent wide local excision of the entire lesion with 1.5-cm margins (Figure 3).

Histopathological examination of the excision specimen revealed, in addition to the sebaceous carcinoma (measuring 3.2 × 2.8 cm), a sebaceoma (1.5 × 1.2 cm) and a syringocystadenoma papilliferum (2.1 × 1.8 cm). All surgical margins were clear.

At 12-month follow-up, there was no evidence of local recurrence or metastatic disease. The patient continues to undergo regular clinical surveillance.

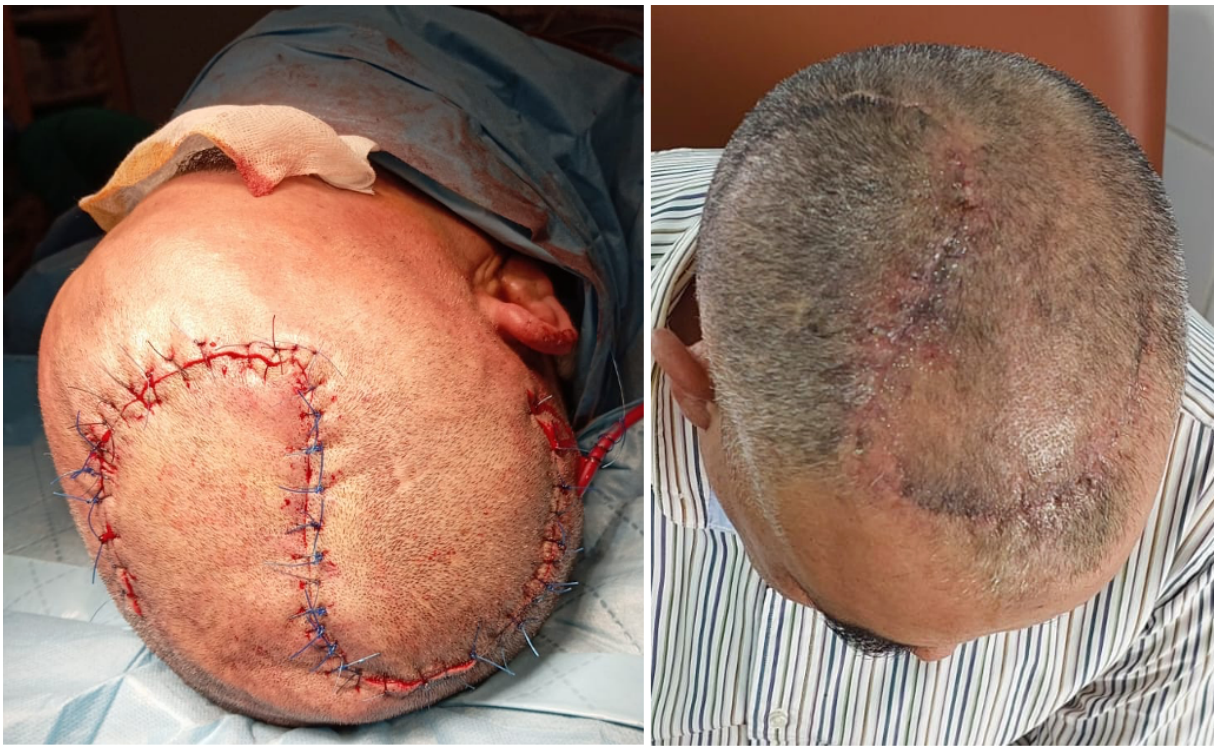

Figure 3: Postoperative clinical photographs following wide surgical excision of the scalp lesion with 1.5-cm margins. The surgical defect was closed primarily.

DISCUSSION

NS is a benign tumor that appears at birth or in early childhood, with a preferential location on the cephalic extremity. It occurs sporadically, sometimes in association with neurological, ophthalmological, or other abnormalities (epidermal nevus syndrome) [5].

Its evolution is characterized by three successive stages:

- First stage: occurring during childhood, where the NS presents as a barely elevated, asymptomatic, yellow-orange or pink alopecic plaque, measuring on average 2 to 3 cm in its greatest dimension.
- Second stage: at puberty, under hormonal influence, the NS increases in size and its surface becomes verrucous, mammillated, or comedonal.
- Third stage: in adulthood, approximately one-third of patients develop tumors on the basis of this hamartoma, with more than 40 different histological types having been described. [6]

In the majority of cases, the tumors that develop on NS are benign, most commonly trichoblastomas and syringocystadenoma papilliferum. Other benign tumors may also arise, including trichilemmoma, sebaceoma, melanocytic nevus, seborrheic keratosis, keratoacanthoma, and other adnexal tumors [3,7].

Malignant transformation is much rarer, most often involving basal cell carcinoma (BCC) and, less frequently, squamous cell carcinoma (SCC). In a recent multicenter study by Ye et al. analyzing 497 cases of secondary tumors arising in NS, 90.3% were benign and only 9.7% were malignant, with BCC being the most common malignancy [8]. The occurrence of

sebaceous carcinoma arising in a NS is exceptional. A comprehensive literature review reveals approximately 30 reported cases to date [4,9,10].

Table 1. Published cases of sebaceous carcinoma arising in nevus sebaceus

Author (Year)	Age/Sex	Location	Associated tumors	Treatment	Follow-up	Outcome
de Giorgi et al. (2003) [11]	67/F	Scalp	None	Wide excision (1 cm)	24 months	No recurrence
Miller et al. (2004) [9]	45/F	Scalp	BCC, trichoadenoma, trichoblastoma, SCAP	Complete excision	Not reported	No metastasis
Matsuda et al. (2005) [12]	53/F	Cheek/neck	None	Wide excision (1.5 cm) + parotidectomy	Not reported	No recurrence
Kazakov et al. (2007) [10]	57-71/F (5 cases)	Scalp, face, nuchal	Sebaceoma, SCAP, trichoblastoma	Excision	Variable	No recurrence
Izumi et al. (2008) [4]	Mean 67.7/F (10 cases)	Scalp (8/10)	Trichoblastoma, sebaceoma, SCAP	Excision	1-7 years	No recurrence
Crandall et al. (2012) [13]	29/F	Scalp	None	Wide excision	Not reported	Not reported
Present case (2025)	45/M	Scalp	SCAP + Sebaceoma	Wide excision (1.5 cm)	12 months	No recurrence

BCC: basal cell carcinoma; SCAP: syringocystadenoma papilliferum

The present case illustrates the exceptional association of three types of secondary tumors—syringocystadenoma papilliferum, sebaceoma, and sebaceous carcinoma—arising within a congenital NS. This combination has not been previously reported in the literature.

Syringocystadenoma Papilliferum

Syringocystadenoma papilliferum (SCAP) is a relatively common benign tumor that occurs preferentially on the scalp, arising in a pre-existing NS in approximately 40% of cases [5].

Other locations also correspond to those of NS. Clinically, the initially flat or slightly verrucous lesion becomes vegetating, mammillated, congested, and often oozing after puberty. Transformation into syringocystadenocarcinoma is possible but exceptional [14].

Dermoscopy of SCAP is characterized by exophytic papillary structures, a central depression, erosions, crusts (or ulceration), as well as hairpin and polymorphous vessels observed in most cases. Yellow-white areas and an erythematous background intermingled with vascular structures have also been described in SCAP arising on NS [5]. The diagnosis relies on histological examination showing an irregular cystic cavity communicating with the surface through one or more orifices lined by hyperplastic epidermis. The cavity is filled with irregular villous projections lined by a bilayered epithelium.

Sebaceoma

The term "sebaceoma" was proposed by Troy and Ackerman to designate a particular sebaceous tumor, previously described as basal cell carcinoma with sebaceous differentiation or sebaceous epithelioma [15]. These benign tumors typically present as solitary, flesh-colored to yellowish papulonodules on the face or scalp, but they may be multiple, particularly in the setting of Muir-Torre syndrome or in association with NS. Their growth is relatively slow, and they generally do not recur after treatment. Histologically, the tumor is composed of irregularly shaped basaloid cells with differentiation toward sebaceous cells. In a sebaceoma, basaloid cells must outnumber differentiated sebocytes [15].

Sebaceous Carcinoma

Sebaceous carcinoma (SC) is a malignant tumor arising from the sebaceous glands, characterized by aggressive behavior and a high metastatic potential [16]. It typically occurs in elderly patients on the upper eyelid. Exceptionally, it may complicate a NS; approximately 30 cases have been published, all arising in the cephalic region, with a slight female predominance and most often after the age of 50, with only two cases described before the age of 30 [4,13].

Dermoscopic Features

Dermoscopy plays an increasingly important role in the surveillance of NS. While SCAP typically shows exophytic papillary structures with hairpin vessels, sebaceoma is characterized by yellow globules and a crown of vessels, and sebaceous carcinoma may display polymorphous vessels, yellow structureless areas, and ulceration [1,17].

In a study by Cheng et al. analyzing 15 cases of sebaceous carcinoma, the majority of tumors (66.67%) presented a polymorphic vessel pattern, with other features including whitish-pink areas (80%), yellowish structures (73.33%), and yellowish structureless areas (60%) [1]. Yellowish structures were the main dermoscopic findings differentiating SC from squamous cell and basal cell carcinomas ($P < 0.001$), whereas purplish globules, shiny white blotches and strands, and whitish-pink areas distinguished SC from other sebaceous tumors ($P < 0.05$) [1].

In our case, the presence of irregular linear vessels, hemorrhagic crusts, and a pink-orange background prompted biopsy, which confirmed malignancy. These findings suggest that any dermoscopic change from the baseline appearance of a NS, particularly the appearance of polymorphous vessels or ulceration, should raise suspicion for malignant transformation.

Muir-Torre Syndrome

Given the presence of multiple sebaceous neoplasms, Muir-Torre syndrome (MTS) must be considered in the differential diagnosis. MTS is an autosomal dominant disorder characterized by the association of sebaceous tumors with visceral malignancies, most commonly colorectal cancer, in the spectrum of Lynch syndrome [18]. However, SC arising in NS does not appear to be associated with MTS [4,10]. In our patient, the absence of personal or family history of visceral malignancies and the preserved expression of mismatch repair proteins on immunohistochemistry effectively ruled out this diagnosis.

Treatment and Prognosis

There is no established consensus regarding therapeutic management of SC arising in NS; complete excision is the treatment of choice for most teams. Margins of 1 to 1.5 cm have been proposed by several authors [11,12]. Mohs micrographic surgery may provide superior outcomes with lower recurrence rates, as suggested by Su et al. in a study comparing Mohs surgery to standard excision for localized SC [19]. Radiotherapy may be considered in cases of locoregional extension [16].

SC arising in NS appears to carry a better prognosis than SC at other locations. In the series by Izumi et al., no cases of recurrence, lymph node metastases, or distant metastases were observed during a follow-up period of 1 to 7 years [4]. To date, no cases of metastasis or recurrence have been described with this entity. However, given the limited number of reported cases, it is difficult to draw definitive conclusions, and long-term follow-up remains essential.

Prophylactic Excision

The question of prophylactic excision of NS remains controversial. Historically, prophylactic excision of NS was advocated, generally before puberty, to prevent the development of malignant tumors [20]. However, recent publications suggest that the risk of malignancy has been significantly overestimated in the past, possibly due to erroneous histological interpretation of trichoblastomas and basaloid hyperplasia as BCC, and do not justify such an approach [21,22].

In a recent large retrospective study of 953 cases, Zhou et al. identified 15.1 years as the cut-off age for tumor presence in resected NS lesions and recommended that monitoring should begin at 10 years and resection be considered before 15.1 years as tumor risk increases [23].

In young adults, prophylactic excision of nevus sebaceus may be considered, particularly when regular follow-up is difficult to ensure or is declined by the patient. In the absence of surgery, regular clinical surveillance is essential. Any suspicious change should prompt dermoscopic examination and excisional biopsy.

Clinical Implications

This case highlights several important points for clinicians managing patients with nevus sebaceus:

1. NS has pluripotent differentiation capacity and can give rise to multiple tumor types simultaneously, as illustrated by the exceptional association of three distinct neoplasms in our patient.
2. Regular clinical and dermoscopic surveillance is recommended, particularly after puberty and especially after the age of 30 when the risk of malignant transformation increases.
3. Any new nodule, ulceration, rapid growth, or bleeding within a NS should prompt dermoscopic examination and excisional biopsy.

4. Dermoscopic features suggestive of malignancy include polymorphous vessels, irregular linear vessels, yellowish structureless areas, and ulceration.
5. Sebaceous carcinoma arising in NS appears to have a favorable prognosis compared to other locations, but long-term follow-up is essential.
6. Complete surgical excision with adequate margins (1–1.5 cm) remains the treatment of choice.
7. Muir-Torre syndrome should be considered in patients with multiple sebaceous neoplasms, and immunohistochemical staining for mismatch repair proteins is recommended.

CONCLUSIONS

The morphology of NS changes considerably from childhood to adulthood. There is a potential for transformation of NS into both benign and malignant tumors, particularly after the age of 30. Among these, sebaceous carcinoma is an extremely rare but serious complication, underscoring the importance of ongoing surveillance for individuals with NS. This vigilance is crucial, as early detection can have a significant impact on the management of these patients.

To our knowledge, this is the first reported case of the simultaneous occurrence of sebaceous carcinoma, syringocystadenoma papilliferum, and sebaceoma within a single nevus sebaceus, highlighting the remarkable pluripotent nature of this hamartoma and the importance of complete histopathological examination of excised specimens.

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REFERENCES

1. Cheng CY, Su HJ, Kuo TT. Dermoscopic features and differential diagnosis of sebaceous carcinoma. J Dermatol. 2020;47(7):755-762.
https://doi.org/10.1111/1346-8138.15384

2. Kamyab-Hesari K, Seirafi H, Jahan S, et al. Nevus sebaceus: a clinicopathological study of 168 cases and review of the literature. Int J Dermatol. 2016;55(2):193-200.
https://doi.org/10.1111/ijd.12845

3. Idriss MH, Elston DM. Secondary neoplasms associated with nevus sebaceus of Jadassohn: a study of 707 cases. J Am Acad Dermatol. 2014;70(2):332-337.
https://doi.org/10.1016/j.jaad.2013.10.004

4. Izumi M, Tang X, Chiu CS, et al. Ten cases of sebaceous carcinoma arising in nevus sebaceus. J Dermatol. 2008;35(11):704-711.
https://doi.org/10.1111/j.1346-8138.2008.00550.x

5. Hagan WE. Nevus sebaceus of Jadassohn: the head and neck manifestations. Laryngoscope. 1987;97(8 Pt 1):909-914.
https://doi.org/10.1288/00005537-198708000-00004

6. Chepla KJ, Gosain AK. Giant nevus sebaceus: definition, surgical techniques, and rationale for treatment. Plast Reconstr Surg. 2012;130(2):296e-304e.
https://doi.org/10.1097/PRS.0b013e3182589df2

7. Hsu MC, Liau JY, Hong JL, et al. Secondary neoplasms arising from nevus sebaceus: a retrospective study of 450 cases in Taiwan. J Dermatol. 2016;43(2):175-180.
https://doi.org/10.1111/1346-8138.13070

8. Ye Q, Wu Q, Jia M, Li FZ, Fang S. Secondary tumors arising from nevus sebaceus: a multicenter collaborative study and literature review. Dermatology (Basel). 2023;239(1):140-147.
https://doi.org/10.1159/000526143

9. Miller CJ, Ioffreda MD, Billingsley EM. Sebaceous carcinoma, basal cell carcinoma, trichoadenoma, trichoblastoma, and syringocystadenoma papilliferum arising within a nevus sebaceus. Dermatol Surg. 2004;30(12 Pt 2):1546-1549.
https://doi.org/10.1111/j.1524-4725.2004.30552.x

10. Kazakov DV, Calonje E, Zelger B, et al. Sebaceous carcinoma arising in nevus sebaceus of Jadassohn: a clinicopathological study of five cases. Am J Dermatopathol. 2007;29(3):242-248.
https://doi.org/10.1097/DAD.0b013e3180339528

11. de Giorgi V, Massi D, Brunasso G, et al. Sebaceous carcinoma arising from nevus sebaceus: a case report. Dermatol Surg. 2003;29(1):105-107.
https://doi.org/10.1097/00042728-200301000-00021

12. Matsuda K, Doi T, Kosaka H, et al. Sebaceous carcinoma arising in nevus sebaceus. J Dermatol. 2005;32(8):641-644.
https://doi.org/10.1111/j.1346-8138.2005.tb00814.x

13. Crandall M, Satter EK, Hurt M. Extraocular sebaceous carcinoma arising in a nevus sebaceous during pregnancy. J Am Acad Dermatol. 2012;67(3):e111-113.
https://doi.org/10.1016/j.jaad.2011.09.019

14. Bruno CB, Cordeiro FN, Soares FES, Takano GHS, Mendes LST. Dermoscopic aspects of syringocystadenoma papilliferum associated with nevus sebaceus. An Bras Dermatol. 2011;86(6):1213-1216.
https://doi.org/10.1590/S0365-05962011000600027

15. Dua Y, Miller C, Ramakrishnan A. Basal cell carcinoma and syringocystoadenoma papilliferum arising in naevus sebaceous-a narrative review. ANZ J Surg. 2026. doi:10.1111/ans.70646.
https://doi.org/10.1111/ans.70646

16. James WD, Elston DM, Treat JR, Rosenbach MA, Neuhaus IM. Epidermal nevi, neoplasms, and cysts. In: Andrews' Diseases of the Skin. Elsevier Inc.; 2020.

17. Kyllo RL, Brady KL, Hurst EA. Sebaceous carcinoma: review of the literature. Dermatol Surg. 2015;41(1):1-15.
https://doi.org/10.1097/DSS.0000000000000152

18. Satomura H, Ogata D, Arai E, Tsuchida T. Dermoscopic features of ocular and extraocular sebaceous carcinomas. J Dermatol. 2017;44(11):1313-1316.
https://doi.org/10.1111/1346-8138.13905

19. Moscarella E, Argenziano G, Longo C, et al. Clinical, dermoscopic and reflectance confocal microscopy features of sebaceous neoplasms in Muir-Torre syndrome. J Eur Acad Dermatol Venereol. 2013;27(6):699-705.
https://doi.org/10.1111/j.1468-3083.2012.04539.x

20. Su C, Nguyen KA, Bai HX, et al. Comparison of Mohs surgery and surgical excision in the treatment of localized sebaceous carcinoma. Dermatol Surg. 2019;45(9):1125-1135.
https://doi.org/10.1097/DSS.0000000000001780

21. Chun K, Vázquez M, Sánchez JL. Nevus sebaceus: clinical outcome and considerations for prophylactic excision. Int J Dermatol. 1995;34(8):538-541.
https://doi.org/10.1111/j.1365-4362.1995.tb02948.x

22. Barkham MC, White N, Brundler MA, Richard B, Moss C. Should naevus sebaceus be excised prophylactically? A clinical audit. J Plast Reconstr Aesthet Surg. 2007;60(11):1269-1270.
https://doi.org/10.1016/j.bjps.2007.03.038

23. Lee YJ, Han HJ, Kim DY, Yoo CY, Lim JS. Malignant transformation of nevus sebaceous to basal-cell carcinoma: case series, literature review, and management algorithm. Medicine (Baltimore). 2022;101(31):e29988.
https://doi.org/10.1097/MD.0000000000029988

24. Zhou W, Cao Y, Wu C, et al. Should be removed or taken biopsy in early stage: a retrospective study of 953 cases of nevus sebaceous from a single center. J Dermatol. 2025. doi:10.1111/1346-8138.17941
https://doi.org/10.1111/1346-8138.17941