

# Secondary Neoplasm Arising in Nevus Sebaceous of Jadassohn: A Case Report Emphasizing Early Recognition and Evaluation

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## Abstract

**Background:** Nevus sebaceous of Jadassohn (NSJ) is a congenital hamartomatous lesion involving epidermal, sebaceous, and adnexal structures. The lesion typically appears at birth or early childhood and remains stable for years. Secondary neoplastic transformations, either benign or malignant, may occur during adolescence or adulthood. **Case:** A 15-year-old girl presented with a yellowish plaque on the vertex of her scalp, first noticed at the age of five. Over time, the lesion enlarged, and a bluish-gray nodule developed two weeks prior to presentation, accompanied by localized pain. Dermatological examination revealed a cerebriform yellow-orange plaque with bluish nodules and alopecia. Dermoscopy showed yellow globules, a homogeneous yellow-orange background, and blue-gray ovoid nests. Histopathology confirmed features consistent with NSJ, including epidermal hyperplasia, papillomatosis, sebaceous glands opening into the epidermis, immature hair follicles, and ectopic apocrine glands. In addition, basaloid cell proliferation with peripheral palisading and melanin pigment suggested a secondary neoplasm. The overall findings are consistent with pigmented trichoblastoma, although the possibility of a pigmented basal cell carcinoma cannot be definitively ruled out. **Discussion:** The findings indicate neoplastic transformation arising within NSJ. Secondary tumors may develop even in adolescence, making early recognition crucial. Clinical changes such as rapid growth, color change, or pain should raise suspicion and prompt histopathological confirmation. Identifying secondary neoplasms influences management decisions, including the need for surgical excision. **Conclusion:** This case highlights the need for early recognition and thorough evaluation of NSJ lesions, particularly in adolescence, to detect and manage secondary neoplastic changes effectively.

**Keywords :** Excision, Nevus Sebaceous, Secondary Neoplasm, Transformation, Trichoblastoma

## Abstrak

**Latar Belakang:** Nevus sebaceous of Jadassohn (NSJ) adalah lesi hamartoma kongenital yang melibatkan struktur epidermis, sebacea, dan adneksa. Lesi biasanya muncul sejak lahir atau masa kanak-kanak awal dan tetap stabil selama bertahun-tahun. Transformasi neoplastik sekunder; baik jinak maupun ganas, dapat terjadi pada masa remaja atau dewasa. **Kasus:** Seorang anak perempuan usia 15 tahun datang dengan plak kekuningan di vertex kulit kepala yang pertama kali disadari pada usia lima tahun. Seiring waktu lesi membesar dan dua minggu sebelum datang muncul nodul biru abu-abu yang disertai nyeri lokal. Pemeriksaan dermatologis menunjukkan plak kuning jingga berbentuk serebriform dengan nodul kebiruan dan alopecia. Dermoskopi menunjukkan "yellow globules", "homogeneous yellow orange background", dan "blue gray ovoid nests". Histopatologi menunjukkan gambaran sesuai NSJ berupa hiperplasia epidermis, papillomatosis, kelenjar sebacea yang bermuara ke epidermis, folikel rambut imatur, dan kelenjar apokrin ektopik. Selain itu

ditemukan proliferasi sel basaloid dengan palisading perifer dan pigmen melanin yang mengarah ke neoplasma sekunder. Temuan keseluruhan konsisten dengan trichoblastoma berpigmen, walaupun kemungkinan basal cell carcinoma berpigmen tidak dapat sepenuhnya disingkirkan. **Diskusi:** Temuan ini menunjukkan adanya transformasi neoplastik pada NSJ. Tumor sekunder dapat muncul bahkan pada masa remaja sehingga pengenalan dini menjadi sangat penting. Perubahan klinis seperti pertumbuhan cepat, perubahan warna, atau nyeri harus menimbulkan kecurigaan dan mendorong konfirmasi histopatologi. Identifikasi neoplasma sekunder memengaruhi keputusan tata laksana termasuk kebutuhan eksisi bedah. **Kesimpulan:** Kasus ini menekankan pentingnya pengenalan dini dan evaluasi menyeluruh pada lesi NSJ, khususnya pada remaja, untuk mendeteksi dan menangani perubahan neoplastik sekunder secara efektif.

**Kata kunci :** Eksisi, Nevus sebaceus, Neoplasma Sekunder, Transformasi, Trichoblastoma

## I. INTRODUCTION

Nevus sebaceus of Jadassohn (NSJ) is a congenital hamartomatous lesion of the skin characterized by disorganized proliferation of epidermal, follicular, sebaceous, and apocrine elements. The lesion typically presents as a well-demarcated, yellowish, hairless plaque, most commonly located on the scalp or face. During infancy and childhood, NSJ usually remains asymptomatic and stable. However, hormonal influences at puberty often trigger histological and morphological changes, including the potential for secondary neoplastic transformation.<sup>1</sup>

Secondary neoplasms arising within NSJ may range from benign adnexal tumors such as trichoblastoma and syringocystadenoma papilliferum to malignant lesions, with basal cell carcinoma being the most frequently reported.<sup>2</sup> These transformations often present as newly developed nodular growths, changes in color or surface texture, or symptoms such as pain or ulceration. Recognizing these changes early is essential, as they may significantly influence clinical management.<sup>3</sup>

The objective of this case report is to describe a rare presentation of NSJ with suspected secondary neoplastic growth in adolescence and to highlight the importance of early recognition and histopathologic evaluation. Emphasis is placed on the clinical, dermoscopic, and microscopic features that suggest transformation, and how these findings impact decisions regarding surgical intervention.

This case underscores the clinical benefit of regular monitoring of long-standing NSJ lesions and provides insight into when further diagnostic workup is warranted to ensure timely and appropriate treatment.

## II. CASE DESCRIPTION

A 15-year-old female patient presented with a yellow-orange patch accompanied by a bluish-gray nodule and localized hair loss on the vertex of the scalp. The nodule had gradually enlarged and become painful over the past two weeks. Seven years earlier, the patient had first noticed a yellowish patch the size of a corn kernel on the same area. Five years ago, the lesion had slowly increased in size and became more prominent. Three years prior to presentation, a small nodule developed within the yellow patch, which was neither itchy nor painful. Over time, the nodule gradually enlarged. Pain began to be felt at the site of the nodule two weeks before presentation. There was no history of malignancy or chronic systemic disease.

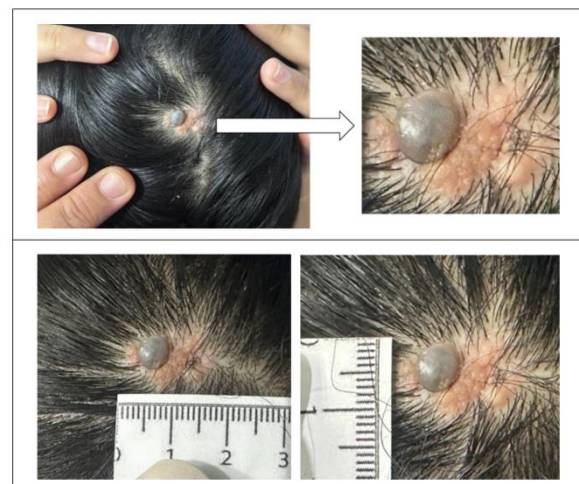
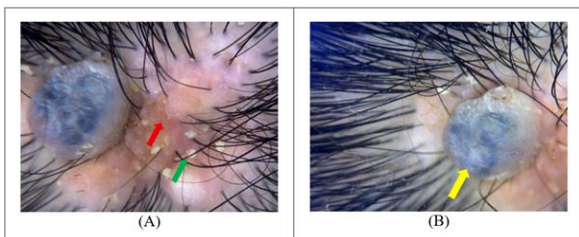
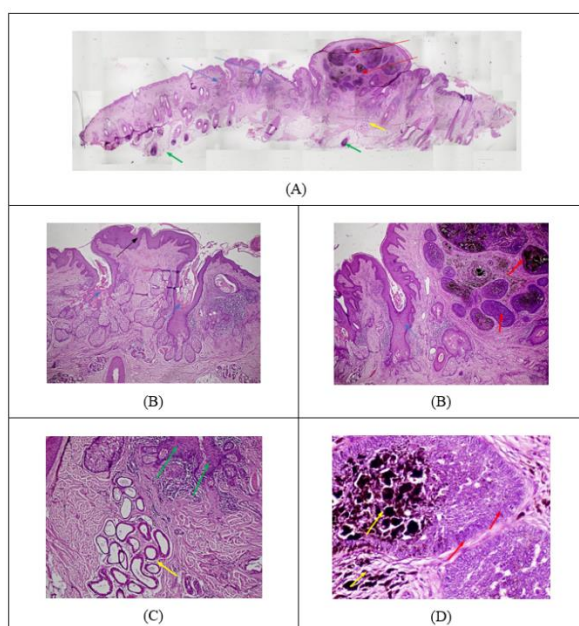


FIGURE 1. CLINICAL PRESENTATION OF THE PATIENT.

Dermatological examination revealed a cerebriform yellow-orange plaque on the vertex of the scalp measuring  $2 \times 1.2 \times 0.1$  cm, a bluish-gray nodule measuring  $0.8 \times 0.7 \times 0.3$  cm, and localized alopecia. On palpation, the nodule was tender, firm, and non-fluctuant. The patient reported a Visual Analog Scale (VAS) pain score of 3. The Dermatology Life Quality Index (DLQI) was 9, indicating a moderate impact on the patient's quality of life. Complete blood laboratory examination showed within normal limit.



**FIGURE 2. DERMOSCPIC EXAMINATION OF THE LESION. (A) THE YELLOW-ORANGE PLAQUE SHOWS YELLOW GLOBULES (GREEN ARROW) AND A HOMOGENEOUS YELLOW-ORANGE BACKGROUND (RED ARROW). (B) THE NODULAR AREA REVEALS A BLUE-GRAY OVOID NEST (YELLOW ARROW).**



**FIGURE 3. HISTOPATHOLOGICAL EXAMINATION. (A) PANORAMIC VIEW OF THE WHOLE SECTION. MAGNIFIED VIEWS AT (B) 4×, (C) 10×, AND (D) 40×. SEBACEOUS GLANDS OPENING INTO THE EPIDERMIS ARE VISIBLE (BLUE ARROW), ALONG WITH ACANTHOSIS (BLACK ARROW), IMMATURE HAIR FOLLICLES (GREEN ARROW), ECTOPIC APOCRINE GLANDS (YELLOW ARROW), AND MELANIN-CONTAINING BASALOID CELLS (RED ARROW).**



**FIGURE 4. CLINICAL PHOTOGRAPH OF THE LESION AFTER TOTAL EXCISION.**

### III. DISCUSSION

The lesion of NSJ typically progresses through three clinical stages. The first stage appears during infancy or early childhood as a yellow to yellow-orange, hairless plaque or nodule.<sup>4</sup> The second stage occurs during puberty and is characterized by sebaceous gland hyperplasia and proliferation of adnexal elements. The lesion may darken and become verrucous. The third stage arises in adulthood, during which the risk of secondary neoplastic transformation increases significantly.<sup>5</sup>

Shandu et al. in 2023 reported that in the patch stage, the epidermis appears normal with numerous hyperplastic sebaceous glands and small primordial hair follicles. In the plaque stage, histopathologic findings include papillomatosis, acanthosis, sebaceous gland hyperplasia, ectopic apocrine glands, and follicular development. In this case, the patient developed a yellowish patch on the scalp at the age of five, which gradually enlarged. Several years later, a bluish-gray nodule appeared at the same site and became painful, indicating progression to the second stage of NSJ.<sup>6</sup>

Nevus sebaceus may give rise to benign or malignant neoplasms. Malignant transformation is rare, with a reported incidence of approximately 2.5%. The most common malignancy arising from NS is basal cell carcinoma (BCC), seen in around 0.8% of 1000 reported cases.<sup>7</sup> Dermoscopic features such as blue-ovoid nests and blue-gray globules are typical of BCC. In this patient, dermoscopy revealed yellow globules and a homogeneous yellow-orange background on the patch, and a blue-gray ovoid nest on the nodule, suggesting basaloid cell proliferation.

Histopathological examination of the total excised specimen revealed epidermal hyperplasia, papillomatosis, acanthosis, sebaceous glands opening directly into the

epidermis, immature hair follicles, ectopic apocrine glands, proliferation of basaloid cells with peripheral palisading, melanin-containing tumor cells, and melanophages within the stroma. These findings were consistent with pigmented trichoblastoma, but the possibility of pigmented BCC could not be ruled out. Immunohistochemical staining using Ber-EP4 can help distinguish BCC from trichoblastoma, as Ber-EP4 typically shows strong and diffuse positivity in BCC, and weak or focal staining in trichoblastoma.<sup>8,9</sup> However, Ber-EP4 testing was not available in this case.

A complete surgical excision was performed to ensure diagnostic accuracy. Limited biopsy specimens may lead to misdiagnosis due to histologic heterogeneity within the lesion. Yeon et al. in 2022 reported two similar cases in which trichoblastoma and syringocystadenoma papilliferum were identified in preoperative punch biopsy, but BCC was not detected until total excision was performed.<sup>10</sup>

The decision to perform excision in cases of NSJ should be based on clinical features and patient age. Prophylactic excision during the first stage remains controversial due to the low malignancy risk. Heather et al., in an 18-year review of 631 patients, reported the youngest case of BCC at age nine, with an overall incidence of 0.8%.<sup>11</sup> Suspicious morphological changes require careful evaluation regardless of age. In the second stage, surgical excision may be justified due to rapid morphological and histological progression. However, the decision must consider the low rate of malignant transformation at this stage and the risk of postoperative complications such as alopecia. Total surgical excision remains the most effective method to achieve accurate diagnosis and reduce diagnostic delay in patients with morphologically evolving lesions.<sup>12</sup>

#### IV. CONCLUSION

Nevus sebaceus of Jadassohn (NSJ) can give rise to secondary neoplasms, including benign and malignant types. Clinical changes such as pigmentation and nodularity should prompt further evaluation regardless of age. In this case, histopathological and dermoscopic findings supported a secondary neoplasm within NSJ, likely pigmented trichoblastoma, with pigmented BCC still a possibility. Complete excision ensured diagnostic accuracy. Early recognition and proper management are key to preventing delayed diagnosis and improving outcomes.

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