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Nevus Sebaceus of Jadassohn of the Scalp Clinically Mimicking Syringocystadenoma Papilliferum: A Case Report

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KEYWORDS

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ABSTRACT:

Nevus sebaceus of Jadassohn is a congenital organoid hamartoma of the pilosebaceous unit that most commonly presents as an alopecic plaque over the scalp and may become thicker or more verrucous with age, occasionally raising concern for secondary adnexal neoplasms. We report the case of a 23-year-old woman who presented to the Department of Dermatology, Venereology and Leprosy, Sree Balaji Medical College and Hospital, with a skin-coloured scalp lesion present since birth and showing gradual increase in size over the preceding 1 year. She had no history of pain, pruritus, discharge, bleeding, trauma, or topical application. Cutaneous examination revealed a solitary, well-defined, non-tender 3 × 3 cm plaque with a mildly cerebriform surface over the left parietal scalp, without ulceration or surrounding inflammation. In view of the recent enlargement and plaque morphology, syringocystadenoma papilliferum arising in a congenital adnexal hamartoma was considered clinically. Histopathological examination, however, confirmed the diagnosis of nevus sebaceus. As there were no clinical or histological features suggestive of secondary neoplasia, the patient was reassured and advised regular follow-up, with instructions to report any further increase in size, nodularity, bleeding, ulceration, or other surface change. This case highlights the importance of considering nevus sebaceus in the differential diagnosis of longstanding congenital alopecic scalp plaques with recent morphological change and underscores the key role of histopathology in establishing the diagnosis and excluding associated neoplasms.

Introduction

Nevus sebaceus of Jadassohn is a congenital organoid hamartoma of the skin composed of abnormal epidermal, follicular, sebaceous, and often apocrine elements, and it is usually identified at birth or in early infancy as a circumscribed alopecic plaque on the scalp or face.(1) Its reported prevalence is approximately 0.3% in newborns, and the scalp is the commonest site of involvement. In a clinicopathologic study of 596 cases, Cribier et al. found that 49.8% of lesions were located on the scalp, underscoring the strong predilection of this entity for hair-bearing areas of the head.(2) The clinical appearance of nevus sebaceus changes with age in a manner that reflects its histologic maturation. Alessi and Sala, in their clinicopathologic study of 130 specimens, showed that all structures derived from the surface ectoderm were affected to varying degrees and that the lesion evolved from a relatively flat childhood plaque to a more verrucous and papillomatous lesion after puberty.(3) Kamyab-Hesari et al., in a 168-case clinicopathologic study, further emphasized that nevus sebaceus is not a static lesion but one that undergoes age-related architectural change, with progressive epidermal hyperplasia, sebaceous prominence, and adnexal alteration accounting for its increasingly raised, verrucous, or cerebriform morphology in adolescence and adulthood.(4) These clinicopathologic features explain why a longstanding congenital alopecic plaque that begins to enlarge later in life may still represent benign maturation rather than overt neoplastic transformation.

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At the molecular level, nevus sebaceus is now recognized as a mosaic RAS-pathway disorder. Sun et al. demonstrated postzygotic activating HRAS and KRAS mutations in lesional tissue from nevus sebaceus and nevus sebaceus syndrome, establishing a genetic basis for its congenital, localized, and mosaic distribution.(5) This molecular understanding is important because it links nevus sebaceus to abnormal adnexal differentiation and helps explain its potential to give rise to secondary follicular, sebaceous, and apocrine neoplasms over time. Although most lesions remain benign, secondary tumours are a major reason nevus sebaceus continues to attract clinical attention. In Cribier et al., benign tumours occurred in 13.6% of cases, whereas basal cell carcinoma was identified in only 0.8%, with a mean age of 39.3 years; importantly, no malignant tumours were observed in children.(2) Syringocystadenoma papilliferum and trichoblastoma were the most frequent benign neoplasms in that series, and Jaqueti et al. likewise identified trichoblastoma as the commonest neoplasm arising in nevus sebaceus.(6) Dermoscopy can aid recognition of lesion stage and secondary change: Kelati et al., in 13 cases, reported yellowish or brown globules aggregated on a yellow background in early lesions, while whitish-yellow lobular and grayish papillary patterns were associated with more verrucous plaques;(7) more recently, Sandhu et al. noted that exophytic papillary projections with vessels, erosions, crusting, and scale were suggestive of associated syringocystadenoma papilliferum.(8) Against this background, the present case documented in the Department of Dermatology, Venereology and Leprosy, Sree Balaji Medical College and Hospital, was noteworthy because a clinically evolving scalp plaque raised concern for secondary adnexal change but was ultimately confirmed histopathologically as nevus sebaceus.

Case Report

A 23-year-old woman presented with a skin-coloured lesion over the scalp that had been present since birth and had increased in size over the preceding 1 year. According to the patient, a localized patch of hair loss had been noted at birth over the left parietal scalp, and the lesion had remained asymptomatic and stable in size throughout childhood and adolescence before it gradually enlarged, which was in keeping with the typical evolution of nevus sebaceus from a congenital alopecic plaque to a more prominent plaque in later years. There was no history of pain, pruritus, discharge, bleeding, preceding trauma, or application of topical medications, and systemic examination was unremarkable. Cutaneous examination revealed a solitary, well-defined, non-tender 3 × 3 cm plaque with a mildly cerebriform surface over the left parietal region of the scalp, without ulceration or surrounding inflammation. In view of the gradual increase in size and plaque morphology, syringocystadenoma papilliferum arising in a congenital adnexal hamartoma was considered clinically in the differential diagnosis. Histopathological examination, however, was diagnostic of nevus sebaceus. As no features suggestive of secondary neoplasia were present, the patient was reassured and advised regular follow-up, with instructions to report promptly if there was any further increase in size, development of nodularity, bleeding, ulceration, or other surface change, as secondary tumours in nevus sebaceus are uncommon and are more often benign than malignant.

Discussion

The present case was most consistent with nevus sebaceus of Jadassohn, a congenital organoid hamartoma of the pilosebaceous unit that affects approximately 0.3% of newborns and most often involves the scalp, face, or neck. In infancy and early childhood, these lesions typically appear as well-circumscribed, hairless, smooth yellow-orange patches or plaques, and localized alopecia over the scalp is a classic clue.(9) The history in our patient—a congenital alopecic scalp lesion that remained stable for many years and then became more apparent in early adulthood—therefore closely paralleled the natural history described for nevus sebaceus. The evolution of this lesion in our patient also fit the recognized stage-wise progression of nevus sebaceus. Early lesions are relatively flat and clinically subtle, whereas around puberty and thereafter they tend to thicken and become more verrucous, mamillated, or nodular because sebaceous elements enlarge, malformed follicular structures become more conspicuous, and ectopic apocrine differentiation becomes more evident.(10) Troupi et al. describes three developmental stages: a childhood stage with flat alopecic plaques, a pubertal stage with thickening and verrucous change, and a later stage in which secondary neoplasms may arise.(11)

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Kerwin & Menter similarly notes that childhood change is unusual, but that expected pubertal thickening may make the lesion appear bumpy, warty, or scaly.(12) Although our patient presented at 23 years of age rather than during adolescence, the recent gradual enlargement and mildly cerebriform surface were still in keeping with the expected later morphologic maturation of nevus sebaceus rather than, by themselves, evidence of malignancy.

Current molecular data have substantially clarified the pathogenesis of nevus sebaceus. Groesser et al. studied 65 sebaceous nevi and identified postzygotic activating HRAS mutations in 62 lesions (95%) and KRAS mutations in the remaining 3 lesions (5%), establishing nevus sebaceus as a mosaic RAS-pathway disorder.(13) Happle subsequently emphasized that HRAS c.37G>C was the hotspot event, occurring in 91% of lesions, which supports the concept that these lesions represent localized developmental anomalies caused by somatic mutation rather than inherited disease.(14) This molecular framework explains why nevus sebaceus is usually present at birth, remains sharply localized, and may later serve as a background for other adnexal proliferations that share overlapping folliculosebaceous-apocrine differentiation. In the present case, the solitary scalp lesion and normal systemic examination also argued against a syndromic nevus sebaceus phenotype such as Schimmelpenning-Feuerstein-Mims syndrome, in which large or disseminated lesions are accompanied by neurologic, ocular, or skeletal abnormalities.(15)

Histopathology remained decisive in this patient because the clinical differential diagnosis included syringocystadenoma papilliferum. In a clinicopathologic analysis of 21 cases, Simi et al. found abortive or immature hair follicles in 100% of lesions, absence of normal terminal hair follicles within the lesion in 100%, epidermal acanthosis, papillomatosis, and hyperkeratosis in 86%, dilated apocrine glands in 19%, and eccrine hyperplasia in 14%.(16) They also emphasized that the absence of normal terminal follicles within the lesion is a particularly valuable low-power diagnostic clue. Gu et al. similarly describes immature and abnormally formed pilosebaceous units in early lesions, followed by sebaceous glands located unusually high in the dermis and increased malformed ducts as the lesion matures.(17) Thus, a histologic diagnosis of nevus sebaceus in our patient would have been supported by the expected constellation of follicular immaturity, sebaceous prominence, and epidermal papillomatous change, even though the surface morphology had raised concern for an associated adnexal neoplasm.

The clinical suspicion of syringocystadenoma papilliferum was nevertheless reasonable. Basu et al., in a case series and literature review, described syringocystadenoma papilliferum as a rare benign adnexal tumor that is seen commonly in infants and children (50%) and less commonly around puberty or adolescence (15%–30%); they also noted that the head and neck account for 75% of cases and that the lesion may present as a smooth hairless plaque or undergo nodular or verrucous transformation.(18) Additionally, Agrawal et al. reported that approximately one-third of syringocystadenoma papilliferum lesions arise within a nevus sebaceus.(19) Histologically, syringocystadenoma papilliferum is characterized by duct-like invaginations communicating with the surface, papillary projections, and a two-layered epithelium, while the stromal core typically shows a dense plasma-cell infiltrate; decapitation secretion in the luminal cells favours apocrine differentiation. Because our patient's biopsy was diagnostic of nevus sebaceus without those papillary glandular features, the clinically suspected syringocystadenoma papilliferum was excluded.

An important point in discussing this case is the true risk of secondary tumour development in nevus sebaceus, because this has historically driven aggressive management. Namiki et al. quoted malignant transformation rates of 5% to 20%,(20) but Morimura et al. have reported markedly lower rates of 0% to 2.5%,(21) and this discrepancy has been attributed in part to historic misclassification of trichoblastoma as basal cell carcinoma. Jiang et al. states that the actual incidence of basal cell carcinoma arising in nevus sebaceus is less than 1% and identifies trichoblastoma as the most frequent secondary tumor, followed by syringocystadenoma papilliferum.(22) A 2024 systematic review and meta-analysis by Pang et al. reported an overall secondary-neoplasm rate of 12.8%, comprising 10.3% benign tumors and 2.4% malignant tumors.(23) These data are clinically relevant here because the patient had a lesion that enlarged but showed no ulceration, bleeding, pain, or histologic evidence of neoplasia; in that context, enlargement

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alone should not be overinterpreted as malignant transformation.

Regarding management, the current recommendations acknowledge that treatment remains controversial, with approaches ranging from observation to elective excision, depending on cosmetic burden, patient preference, diagnostic certainty, and the presence of suspicious clinical change. Lopez & Lam notes that the treatment of nevus sebaceus is controversial and that options range from observation to early excision in childhood.(1) Castagna et al. advises biopsy when unexpected surface change occurs and highlights crusting or a new focal change as indications for reassessment.(24) Accordingly, the advice given to our patient—to report any further increase in size, nodularity, ulceration, bleeding, or other surface change—was appropriate, because it balanced the very low absolute risk of malignancy against the need for surveillance of a lesion known to evolve over time and occasionally to develop secondary adnexal tumours in adulthood. Overall, this case illustrated a classic but diagnostically instructive presentation of nevus sebaceus on the scalp in a young adult. The congenital onset, localized alopecia, delayed enlargement, and plaque morphology were characteristic, while the mildly cerebriform surface appropriately prompted consideration of syringocystadenoma papilliferum, one of the better-recognized secondary neoplasms associated with nevus sebaceus. The case also reinforced a practical dermatopathologic principle: when a longstanding scalp plaque with alopecia shows recent textural change, histopathology is essential not only to confirm nevus sebaceus but also to exclude associated adnexal tumours that may mimic it clinically. In that respect, this patient's benign histology and absence of alarming clinical features justified surveillance rather than immediate intervention, while still underscoring the need for continued follow-up in adulthood.

Conclusion

This case highlighted a classic presentation of nevus sebaceus of the scalp with recent increase in size in early adulthood, clinically mimicking syringocystadenoma papilliferum. It underscored the importance of histopathological examination in confirming the diagnosis and excluding secondary adnexal neoplasms in longstanding congenital scalp plaques showing morphological change. In the absence of suspicious clinical or histological features, careful counselling and regular follow-up remained an appropriate management approach.

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