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Multiple rare neoplasms arising from the nevus sebaceous of the scalp: A case report

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ABSTRACT

Nevus sebaceous of Jadassohn (NSJ) is a cutaneous hamartoma of pilosebaceous origin that differentiates into multiple neoplasms, most commonly trichoblastoma/basal cell carcinoma and syringocystadenoma papilliferum (SCAP). Malignant transformation of NSJ is rare and usually observed in the elderly. However, a diagnosis of more than three separate tumors concurrent to multiple malignancies developing from a single NSJ is extremely rare. We here report a case of a 72-year-old male patient with five distinct tumors arising from NSJ of the scalp, including SCAP, tubular apocrine adenoma, eccrine poroma, apocrine cystadenoma, and cutaneous apocrine carcinoma.

Introduction

Nevus sebaceous of Jadassohn (NSJ), first described by Joseph Jadassohn in 1895, is an organoid nevus of pilosyringosebaceous and adnexal origin that combines sebaceous gland abnormalities with anomalies of the epidermal, follicular, and apocrine glands (1,2).

NSJ typically presents at birth as linear/ovoid plaque most commonly on the face and scalp and progresses through self-differentiation into multiple neoplasms in late adulthood (3-5). However, reports of more than three simultaneous tumors from a single nevus and their malignant transformation are extremely rare (6,7). Here, we describe the features of multiple synchronous benign and malignant neoplasms arising from a long-standing nevus sebaceous of the scalp.

Case Presentation

A 70-year-old male patient was admitted with a complaint of a painful slow-growing ulcer in the right buccal mucosa and swelling in the scalp, which had grown slowly over years to its current size with occasional itching. The patient had a history of chronic tobacco use and betel quid chewing. On physical

examination, a lesion that was irregularly surfaced, ulcerated, and proliferating onto the mandibular alveolar process was detected along with asymptomatic solitary dome-shaped nodular growth measuring approximately 7.5x4.5 cm over the left parietooccipital region of the scalp (Figure 1). The lesion was hyperpigmented and soft to firm. An incisional biopsy of the oral lesion was performed and the histopathological evaluation was consistent with a well-differentiated squamous cell carcinoma (stage IVa). The intra-oral lesion was treated by wide excision with hemimandibulectomy; nerve and vein sparing type of neck dissection and reconstruction of the defect were carried out with pectoralis major myocutaneous flap.

Fine-needle aspiration of the lesion on the scalp was reported as syringocystadenoma papilliferum (SCAP). The lesion on the scalp was surgically excised and the defect was covered with a split-thickness skin graft.

Histopathological examination of the lesion revealed multiple lesions in the primary NSJ, namely SCAP, tubular apocrine adenoma (TAA), eccrine poroma (EP), apocrine cystadenoma (ACA), cutaneous apocrine carcinoma (CAC) and cutaneous apocrine carcinoma, with invasive areas (Figure 2A-G). Immunohistochemical studies ruled out basal cell carcinoma and malignant melanoma. S100 for nerve sheath tumors and malignant melanoma, CK 17 and cytokeratin screening for epithelial origin tumors, and CD 99 for mesenchymal tumors/ soft tissue sarcomas were studied during histopathological evaluations.

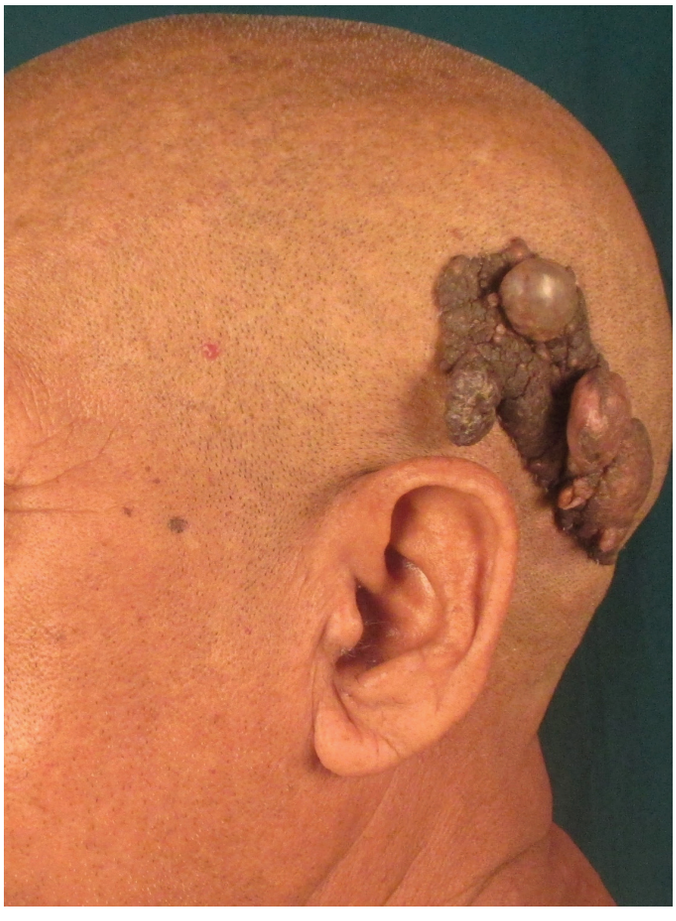

Figure 1. Preoperative lesion on the scalp

Discussion

NSJ follows three phases of its natural history designated by Mehregan and Pinkus (8) as infancy, pubertal stimulation, and post-pubertal histological degeneration; usually appears at birth (0.1-0.3% among newborns) or early childhood as a well-circumscribed, smooth, slightly raised, yellowish-brown alopecic patch that progresses through puberty, assuming a more verrucous texture, possibly due to the expression of various hormonal receptors (4). There have been only a few reported cases in the literature describing the simultaneous development of 5 or more neoplasms in a solitary lesion (2).

SCAPs are the most common hamartoma tumors associated with nevus sebaceous, which originate from the apocrine sweat glands (4). Stavrianeas report an incidence of 5 to 19% of the nevi complicated with SCAP (1). Section from our patient's slides revealed a stratified squamous epithelium with immature sebaceous glands and areas of basaloid hyperplasia along with immature hair structures and dilated ductal infundibulum in the dermis (Figure 2B).

TAA is a minor variant of SCAP consisting of a benign appendage tumor of apocrine origin. TAA and SCAP rarely develop together in the nevus sebaceous, and only a few such cases had been reported in 2004 (9). Sections of our slides showed a well-circumscribed dermal neoplasm comprising lobules of dilated tubules lined by two rows of cuboidal to columnar epithelial cells with eosinophilic cytoplasm and round bland nuclei (Figure 2C).

EP is a benign tumor originating from the epidermal sweat glands and occurs in middle-aged individuals on the sole and hands. However, on the scalp is extremely rare, with 18 cases reported until the year 2012 (10). Sections of our slides showed a downward proliferation of basaloid cells from the epidermis with delicate fibrovascular stroma in the form of anastomosing cords and nests with pigmentation (Figure 2D).

ACA is a rare, benign, solitary adenomatous cystic tumor of the skin. Sections of our slide revealed cystic areas in the dermis lined by columnar cells with small round basal nuclei and abundant eosinophilic cytoplasm arranged in a papillary pattern with a fibrovascular core (Figure 2E).

CAC is a rare malignancy with unknown etiology and only eight cases are reported in 2012 (6). We found many layers of pleomorphic cells with irregular hyperchromatic nuclei, prominent nucleoli, and abundant granular eosinophilic cytoplasm (Figure 2F). There was the presence of atypical mitoses, decapitation secretion, and tumor tissue invasion into the papillary and reticular dermis (Figure 2G).

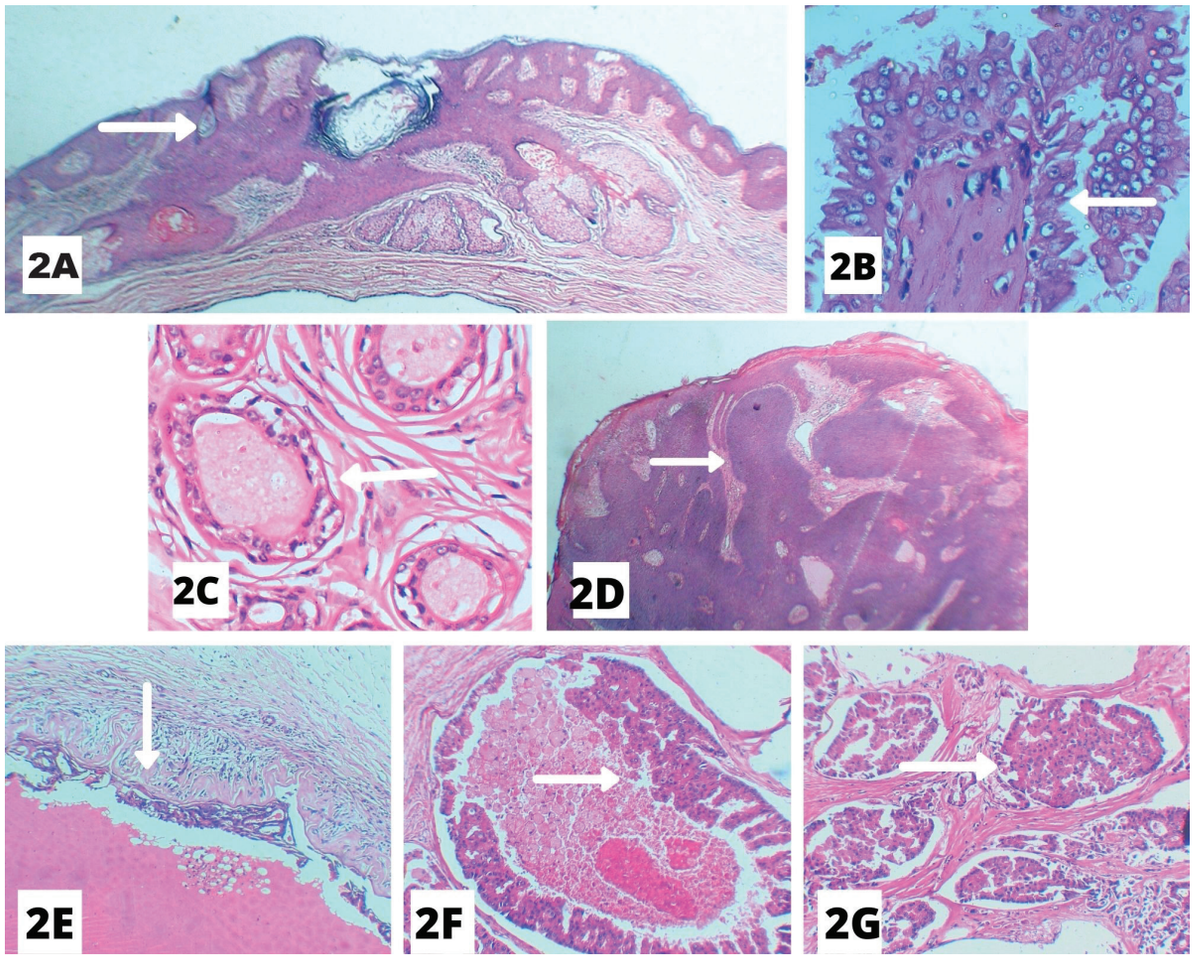

Figure 2. Multiple neoplasms arising from nevus sebaceous of Jadassohn. A: Nevus sebaceous of Jadassohn: [Haematoxylin and eosin (H&E)-stained photomicrograph 30x magnification] of nevus sebaceous of Jadassohn showing immature sebaceous glands, multiple heterotopic apocrine glands, defective hair follicles, and epithelial papillomatosis with hyperkeratosis. B: Syringocystadenoma papilliferum (SCAP): (H&E-stained photomicrograph 100x magnification) of SCAP showing connective tissue composed of glandular and ductal papillary proliferation and dense plasma cell infiltration. C: Tubular apocrine adenoma (TAA): (H&E-stained photomicrograph 100x magnification) of TAA comprised of variably sized intradermal nodules with tubular structures lined by 1-2 layers of cuboidal cells with eosinophilic cytoplasm and round bland nuclei. D: Eccrine poroma: (H&E-stained photomicrograph 30x magnification) of eccrine poroma showing dense basaloid proliferation into the dermis and connective tissue with reactive vessels and chronic inflammatory exudates. Islands of ductal structures were also noted. E: Apocrine cystadenoma (ACA): (H&E-stained photomicrograph 30x magnification) of ACA showing abundant intradermal cystic areas and papillary apocrine metaplasia. The immature glandular structures appear to be lined by a layer of columnar cells with abundant eosinophilic cytoplasm with decapitation secretion noted in multiple areas. F: Cutaneous apocrine carcinoma (CAC): (H&E-stained photomicrograph 100x magnification) of CAC: showing irregular layers of pleomorphic cells with an irregular hyperchromatic nucleus and abundantly granular cytoplasm. Atypical mitoses also were noted. G: CAC: (H&E-stained photomicrograph 100x magnification) with invasive areas and invasion into deeper layers of the dermis

Conclusion

The current case is in agreement with most authors who have suggested that NSJ should be surgically excised, histologically examined, and should be kept on a close follow-up to prevent malignant transformation.

Ethics

Informed Consent: Informed written consent has been obtained from the patient for publication.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: D.S., A.D., N.K., D.U.S., A.A., Concept: D.S., A.D., N.K., D.U.S., Design: D.S., A.D., Data Collection or Processing: D.S., A.A., S.B., Analysis or Interpretation: D.S., A.D., N.K., D.U.S., A.A., Literature Search: D.S., S.B., Writing: D.S., S.B.

Conflict of Interest: No conflict of interest was declared by the authors.

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References

1. Stavrianeas NG, Katoulis AC, Stratigeas NP, Karagianni IN, Paterou-Stavrianea M, Vareltzidis AG. Development of multiple tumors in a sebaceous nevus of Jadassohn. Dermatology. 1997;195:155-158.
2. Gozel S, Donmez M, Akdur NC, Yikilkan H. Development of six tumors in a sebaceus nevus of jadassohn: report of a case. Korean J Pathol. 2013;47:569-574.
3. Manonukul J, Omeapinyan P, Vongjirad A. Mucoepidermoid (adenosquamous) carcinoma, trichoblastoma, trichilemmoma, sebaceous adenoma, tumor of follicular infundibulum and syringocystadenoma papilliferum arising within 2 persistent lesions of nevus sebaceous: report of a case. Am J Dermatopathol. 2009;31:658-663.
4. Idriss MH, Elston DM. Secondary neoplasms associated with nevus sebaceous of Jadassohn: a study of 707 cases. J Am Acad Dermatol. 2014;70:332337.
5. Kantrow SM, Ivan D, Williams MD, Prieto VG, Lazar AJ. Metastasizing adenocarcinoma and multiple neoplastic proliferations arising in a nevus sebaceus. Am J Dermatopathol. 2007;29:462-466.
6. Paudel U, Jha A, Pokhrel DB, Gurung D, Parajuli S, Pant A. Apocrine carcinoma developing in a naevus sebaceous of scalp. Kathmandu Univ Med J (KUMJ). 2012;10:103-105.
7. Namiki T, Miura K, Ueno M, Arima Y, Nishizawa A, Yokozeki H. Four Different Tumors Arising in a Nevus Sebaceous. Case Rep Dermatol. 2016;8:75-79.
8. Mehregan AH, Pinkus H. Life history of organoid nevi. Special reference to nevus sebaceus of Jadassohn. Arch Dermatol. 1965;91:574-588.
9. Ahn BK, Park YK, Kim YC. A case of tubular apocrine adenoma with Syringocystadenoma papilliferum arising in nevus sebaceus. J Dermatol. 2004;31:508-510.
10. Park E, Lee DS, Eom KS. Eccrine poroma on the scalp: a case report with MR findings. J Korean Neurosurg Soc. 2015;1:53-54.