Secondary neoplasms associated with nevus sebaceus of Jadassohn: A study of 707 cases

Munir H. Idriss, MD, and Dirk M. Elston, MD
New York, New York

Background: Nevus sebaceus is commonly associated with the development of secondary neoplasms. Data on the frequency of malignant tumors vary considerably in published reports.

Objective: We sought to analyze the distribution of secondary neoplasm in nevus sebaceus.

Methods: A retrospective analysis of all cases of nevus sebaceus diagnosed at the Ackerman Academy of Dermatopathology from 1999 to 2012 was conducted.

Results: A total of 706 patients (707 specimens) were included in the study. Trichoblastoma was the most frequent benign tumor (n = 52, 7.4%) followed by syringocystadenoma papilliferum (n = 33, 5.2%). Malignant tumors were present in 2.5% of the specimens with basal cell carcinoma being the most common (n = 8, 1.1%) followed by squamous cell carcinoma (n = 4, 0.57%). The incidence of secondary neoplasms was statistically related to age and anatomic site ($P < .05$). Almost all malignant tumors were seen in adults.

Limitation: Some of our cases were referred for second opinion and there may be a bias in our data toward unusual secondary neoplasms.

Conclusion: Our study confirms that most of the secondary neoplasms arising in association with nevus sebaceus are benign. As no malignant tumors were seen in children, we believe it is reasonable to delay surgical management until adolescence. (J Am Acad Dermatol 2014;70:332-7.)

Key words: basal cell carcinoma; hamartoma; nevus sebaceus; organoid nevus; skin; trichoblastoma.

Nevus sebaceus is a congenital hamartoma of cutaneous structures including both surface epithelial and adnexal components. It is classified as an organoid nevus, as it is a malformation of most of the normal tissue components of the skin.[1] Nevus sebaceus usually occurs on the head and neck region and is usually clinically apparent at birth. It presents as a well-demarcated skin-colored to yellowish alopecic patch. Proportional enlargement with age is the rule and at puberty the lesion typically becomes more yellowish and ceribriform. The nature of secondary tumors arising within nevus sebaceus and the risk of malignant neoplasm have been matters of controversy. The timing of excision is also a matter of debate, with some advocating early excision and others suggesting that delayed excision is reasonable.[2-6] Others have cited an absence of true malignant neoplasms in some series and questioned the necessity of excision.[7,8]

This retrospective study was designed to assess the nature and distribution of secondary neoplasms associated with nevus sebaceus at a major dermatopathology referral laboratory in the northeastern United States.

METHODS

A database query of specimens submitted to the Ackerman Academy of Dermatopathology, New York, NY, for histologic evaluation from January 1, 1999, through August 15, 2012, was conducted. Search criteria included any accession with the line diagnosis of nevus sebaceus or organoid nevus. In

From the Ackerman Academy of Dermatopathology.
Funding sources: None.
Conflicts of interest: None declared.
Accepted for publication October 2, 2013.
Reprint requests: Munir H. Idriss, MD, Ackerman Academy of Dermatopathology, 145 E 32 St, 10th Floor, New York, NY 10016. E-mail: mhidriss@gmail.com.

Published online November 22, 2013.
0190-9622/$36.00

http://dx.doi.org/10.1016/j.jaad.2013.10.004

addition, database queries were run for trichoblastoma, syringocystadenoma papilliferum, trichilemmoma, basal cell carcinoma (BCC), and apocrine adenoma. Cases exhibiting some features of nevus sebaceous but not unequivocally diagnostic were excluded. Demographic and clinical data along with reported secondary neoplasms were extracted from the pathology reports of query results. All specimens with benign and malignant neoplasms were reviewed to confirm the diagnosis, except for a few consultation cases that were not available for second review. Occurrence of 2 or more secondary neoplasms in a single lesion was considered as separate diagnoses for statistical purposes, whereas multiple biopsy specimens from a lesion or serial excisions were treated as a single lesion. Established criteria, including those published by Ackerman et al,[9] were used to differentiate BCC from trichoblastoma. These include a fibroblast-rich concentric stroma with papillary mesenchymal bodies and lack of stromal retraction.

CAPSULE SUMMARY

- Nevus sebaceus is associated with secondary neoplasms, most of which are benign.
- Our series of 706 patients highlights that malignant neoplasms arise in about 2.5% of lesions, almost exclusively in adults.
- Because of the virtual absence of malignancy in children, surgical intervention can be delayed.

The Statistica 12.0 statistical package (StataCorp, College Station, TX) was used for statistical evaluation. Differences between frequencies were evaluated with 2×2, 2×3, and 2×5 tables method with the χ^2 and Fisher exact tests. A P value less than .05 was considered statistically significant.

RESULTS

The database query yielded a total of 1114 accessions. After cross-referencing of queries and selection of only definitive cases, 707 specimens were included in the analysis. The age of 4 patients was not available. These cases were disregarded for age-specific analysis but included in all other analyses. There were 400 male (56.7%) and 306 female (43.3%) patients. The difference in proportion of neoplasms between male and female patients was not statistically significant (P = .53). Average age of patients was 27.4 years (range 0-95 years). Most of the nevi were located on the scalp (n = 442, 62.5%) followed by the face (n = 173, 24.5%) and trunk (n = 33, 4.7%), respectively. The adult stage was the most common stage biopsied (n = 438, 57.7%) in our series. The demographic characteristics of our series are summarized in Table I.

Histologic features were consonant with the age group of the patient. Childhood cases were characterized by a relatively normal epidermis, alopecia, and immature pilosebaceous units. Adolescent and adult specimens exhibited greater epidermal hyperplasia in addition to mature adnexal structures in the dermis.

We found a total of 159 secondary proliferations in the 707 specimens (22.5%). In 18.9% (n = 132) of the lesions, there was a secondary benign neoplasm. Of the specimens, 1.2% (n = 9) contained warts, cysts, and connective tissue nevi and 2.5% (n = 18) contained malignant neoplasms. The most common neoplasm was trichoblastoma, which was found in 7.4% (n = 52) of our specimens and accounted for about one third of all secondary proliferations (Fig 1). It was followed in frequency by syringocystadenoma papilliferum (Fig 2) seen in 5.2% of lesions (n = 37, 24.7% of neoplasms). Other benign neoplasms observed included 15 cases of apocrine/eccrine adenoma (10% of neoplasms) (Fig 3), 8 cases of trichilemmoma (Fig 4) (5.3% of neoplasms), 3 cases of desmoplastic trichilemmoma (2% of neoplasms), 2 cases of sebaceoma (1.3% of neoplasms) (Fig 2), and 1 case each of solar keratosis, adenomyoepithelioma, and connective tissue nevus. The most common malignant neoplasm in our series was BCC (n = 8, 5.3% of neoplasms) (Fig 5) followed by squamous cell carcinoma (n = 4, 2.7% of neoplasms) (Fig 6) and sebaceous carcinoma (n = 3, 2% of neoplasms). One case each of apocrine carcinoma and microcystic adnexal carcinoma were also observed (Table II).

The frequency of neoplasms with in nevus sebaceus was related to the age group of patients. Only 5 neoplasms were observed in childhood in contrast to 10 in adolescence and 134 in adulthood (Table III). This difference in the proportion of cases having secondary neoplasms between the age groups was statistically significant ($P < .001$). Similarly, the proportion of both benign and malignant neoplasms was found to be highest in the adult age group and the difference between age groups was statistically significant ($P < .001$ and $P = .0417$, respectively). On analysis of individual tumors arising in nevus sebaceus, the difference in incidence between the age groups was only statistically significant in trichoblastomas ($P < .05$) and BCC ($P = .00048$).

In our series, most of the tumors occurred on the scalp (n = 106) accounting for about 70% of all