Cerebriform congenital scalp mass

Kerrie G. Satcher, MD, Reesa L. Monir, MD, Kiran Motaparthi, MD, and Jennifer J. Schoch, MD
Gainesville, Florida

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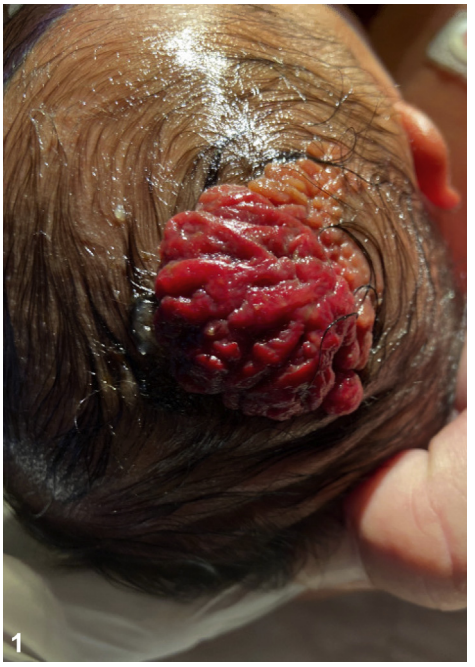

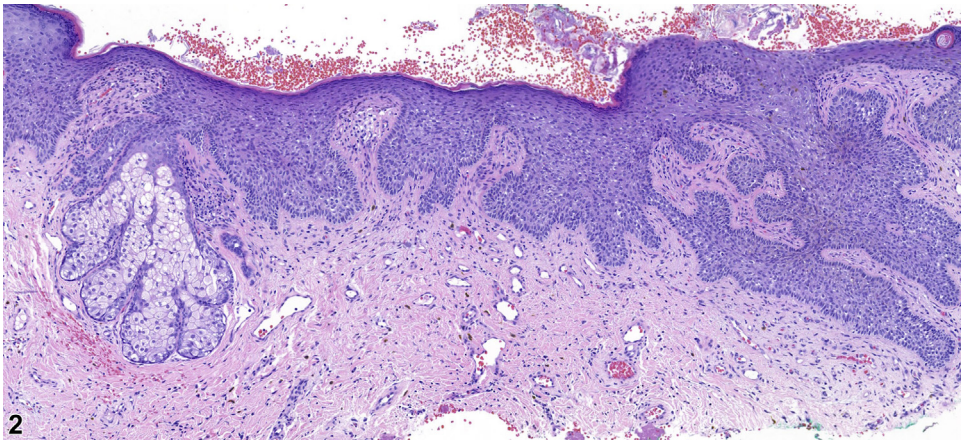

From the Department of Dermatology, University of Florida College of Medicine, Gainesville.

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Correspondence to: Jennifer J. Schoch, MD, Department of Dermatology, University of Florida College of Medicine 4037 NW 86th Terrace, Gainesville, FL 32606. E-mail: jschoch@dermatology.med.ufl.edu.

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An otherwise healthy 3-day old boy was evaluated for a congenital scalp mass. Physical examination revealed a 4-cm exophytic, erythematous, friable, and cerebriform nodule on the right frontoparietal aspect of the scalp (Fig 1). Computed tomography of the head demonstrated a soft-tissue lesion along the right frontoparietal convexity with underlying intact calvarium. Initial biopsy revealed nonspecific histopathologic findings, including a slightly acanthotic epidermis with slit-like dermal vessels. At 2 months of age, an exophytic, yellow-tan cerebriform nodule persisted. Punch biopsy revealed a papillomatous epidermis with abortive follicles, papillary mesenchyme, and ectopic sebaceous lobules (Fig 2).

Question 1: What is the most likely diagnosis?

A. Cutaneous metastasis of neuroblastoma

B. Ectopic brain tissue

C. Encephalocele

D. Keratinocytic epidermal nevus

E. Cerebriform nevus sebaceus

Answers:

A. Cutaneous metastasis of neuroblastoma—Incorrect. These lesions typically present as multiple firm blue-to-purple papules or nodules.

B. Ectopic brain tissue—Incorrect. The histologic findings are not consistent with ectopic brain tissue.

C. Encephalocele—Incorrect. Computed tomography of the head showed an underlying intact calvarium without direct connection to the brain. Additionally, the histologic findings are not consistent with brain tissue.

D. Keratinocytic epidermal nevus—Incorrect. Keratinocytic epidermal nevi typically present as linear or blaschkoid brown plaques with a velvety or verrucous texture. Histology shows epidermal hyperplasia, hyperkeratosis, and papillomatosis.

E. Cerebriform nevus sebaceus—Correct. The cerebriform variant of a nevus sebaceus (NS) is a rare clinical presentation first described in 1998, with less than 15 cases reported. In contrast to the classic NS, which most commonly presents at birth as an alopecic, yellow-tan oval or linear smooth plaque usually on the scalp or face, this variant has a cerebriform appearance, mimicking the sulci and gyri of brain tissue.[1] While histopathologic examination can aid in the diagnosis, findings vary based on the stage.[2] Histopathologic features of early NS include slight papillomatosis, primitive hair follicles, and less prominent sebaceous glands. After puberty, marked acanthosis and papillomatosis of the epidermis can be appreciated along with dilated apocrine glands, prominent sebaceous lobules, and a lack of terminal hair.[3]

Question 2: What is the most common tumor to arise within this lesion?

A. Basal cell carcinoma

B. Syringocystadenoma papilliferum

C. Apocrine cystadenoma

D. Trichoadenoma

E. None, secondary tumors are uncommon

Answers:

A. Basal cell carcinoma—Incorrect. Basal cell carcinomas were initially thought to be common tumors arising within an NS. However, malignant degeneration is rare, with early estimates of prevalence likely including misclassified benign lesions.[3,4]

B. Syringocystadenoma papilliferum—Incorrect. Both syringocystadenoma papilliferum and trichoblastoma may arise within an NS, with studies reporting conflicting data on which is most common.[3,4] However, these secondary tumors develop in a minority of NS, with a recent review citing syringocystadenoma papilliferum in 5.2% and trichoblastomas in 7.4% of such lesions, respectively.[3]

C. Apocrine cystadenoma—Incorrect. This lesion does not commonly arise within an NS.

D. Trichoadenoma—Incorrect. This lesion does not commonly arise within an NS.

E. None, secondary tumors are uncommon—Correct. Neoplasms do not arise within the majority of NS. When secondary tumors do develop within an NS, syringocystadenoma papilliferum and trichoblastoma have been suggested as most common. A retrospective review published in 2016 found that syringocystadenoma papilliferum was the most common tumor to arise within an NS, followed by both trichoblastoma and trichilemmoma.[4] Previously, trichoblastoma was reported as the most common tumor based on a retrospective review in 2014.[3]

Question 3: What are the current management recommendations for most sebaceus nevi?

A. Complete excision in the first 6 months of life

B. Serial treatment with an ablative resurfacing laser

C. Repeat computed tomography of the head at 1 year of life

D. Serial tissue sampling to monitor for malignant degeneration

E. Observation

Answers:

A. Complete excision in the first 6 months of life—Incorrect. Historically, complete excision of an NS was recommended due to concern for secondary malignant epithelial tumors.[5] Elective excision of a large or exophytic NS may be considered for cosmetic concerns, but routine prophylactic excision is no longer recommended. The risks of general anesthesia during the first 6 months of life usually outweigh the benefits of excision early in infancy.

B. Serial treatment with an ablative resurfacing laser—Incorrect. Ablative laser has not been shown to be an effective treatment for NS.

C. Repeat computed tomography of the head at 1 year of life—Incorrect. NS does not extend into the underlying calvarium.[1]

D. Serial tissue sampling to monitor for malignant degeneration—Incorrect. NS is a benign lesion. Although secondary malignant tumors can develop within these lesions, this is rare.[3]

E. Observation—Correct. Given the low risk of malignant secondary tumors, observation is preferred for most NS. In this patient, it would be reasonable to consider excision after 1 year of life based on the size, location on the vertex of the scalp, and exophytic nature of this cerebriform NS.[5]

Abbreviation used:

NS: nevus sebaceus

Conflicts of interest

None disclosed.

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