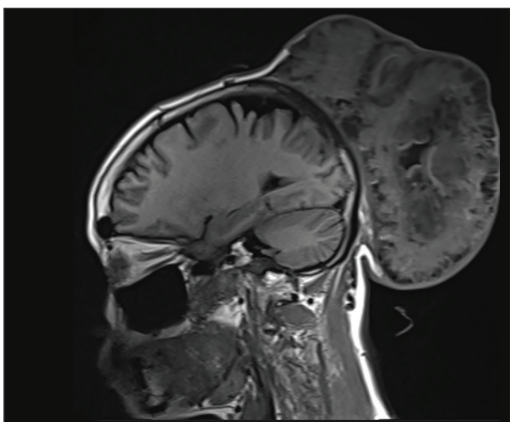


Enlarging Mass on the Scalp

Ava Scott, BS; Samantha K. Ong, MD; Michelle Boettler, MD; Kristin Smith, MD;
Nicole Burkemper, MD; Daniel West, MD

Eligible for 1 MOC SA Credit From the ABD

This Photo Challenge in our print edition is eligible for 1 self-assessment credit for Maintenance of Certification from the American Board of Dermatology (ABD). After completing this activity, diplomates can visit the ABD website (<http://www.abderm.org>) to self-report the credits under the activity title "Cutis Photo Challenge." You may report the credit after each activity is completed or after accumulating multiple credits.



A 61-year-old woman presented to the emergency department with worsening pain and bleeding from a scalp tumor of 16 years' duration. Initially noted as a small nodule on the left parietal scalp on computed tomography of the head, the mass had grown rapidly in recent years and currently measured 22×10×15 cm. At prior consultations with plastic and general surgery, the patient had declined surgical intervention. At the current presentation, biopsies were performed by plastic surgery, and a dermatopathology consultation was ordered. Histopathology revealed atypical keratinocytes, nuclear pleomorphism, lobulating epithelial masses with trichilemmal keratinization, and CD34 positivity. Subsequent computed tomography and positron emission tomography of the head showed occipital skull erosion and bilateral cervical lymphadenopathy, suggesting metastasis.

WHAT'S YOUR DIAGNOSIS?

- cutaneous squamous cell carcinoma
- malignant proliferating trichilemmal tumor
- proliferating trichilemmal tumor
- sebaceous carcinoma
- trichilemmal carcinoma

PLEASE TURN TO **PAGE 153** FOR THE DIAGNOSIS

From the School of Medicine, Saint Louis University, St. Louis, Missouri. Drs. Ong, Boettler, Smith, Burkemper, and West are from the Department of Dermatology. Drs. Ong, Boettler, Smith, Burkemper, and West also are from SLUCare Physician Group & SSM Health, St. Louis.

The authors have no relevant financial disclosures to report.

Correspondence: Ava Scott, BS, 1402 S Grand Blvd, St. Louis, MO, 63104 (ava.scott@health.slu.edu).

Cutis. 2026 May;117(5):142, 153-154. doi:10.12788/cutis.1394

THE DIAGNOSIS:**Malignant Proliferating Trichilemmal Tumor**

Histologic examination revealed atypical keratinocytes, nuclear pleomorphism, and lobulating epithelial masses with trichilemmal keratinization (Figure). The presence of CD34 positivity, a marker of outer follicular root sheath–derived cells, supported the diagnosis of a malignant proliferating trichilemmal tumor (MPTT). Imaging also revealed signs of bone invasion, further supporting a malignant process. Based on these findings, the patient underwent complete excision of the mass with scalp reconstruction, lymph node dissection, and systemic evaluation for metastases. Final pathology confirmed negative surgical margins and no lymph node involvement. Adjuvant radiation was not

required, given the absence of skull invasion or confirmed distant metastasis.

The differential diagnosis for rapidly enlarging scalp tumors can be broad and includes both benign and malignant processes. In this patient, the differential diagnoses included trichilemmal carcinoma, cutaneous squamous cell carcinoma (SCC), sebaceous carcinoma (SC), proliferating trichilemmal tumor (PTT), and MPTT. Due to the notable clinical and histologic overlap among these lesions, definitive diagnosis required histopathologic evaluation in our patient.

Proliferating trichilemmal tumors were first described in 1966 by Wilson-Jones,¹ who used the term

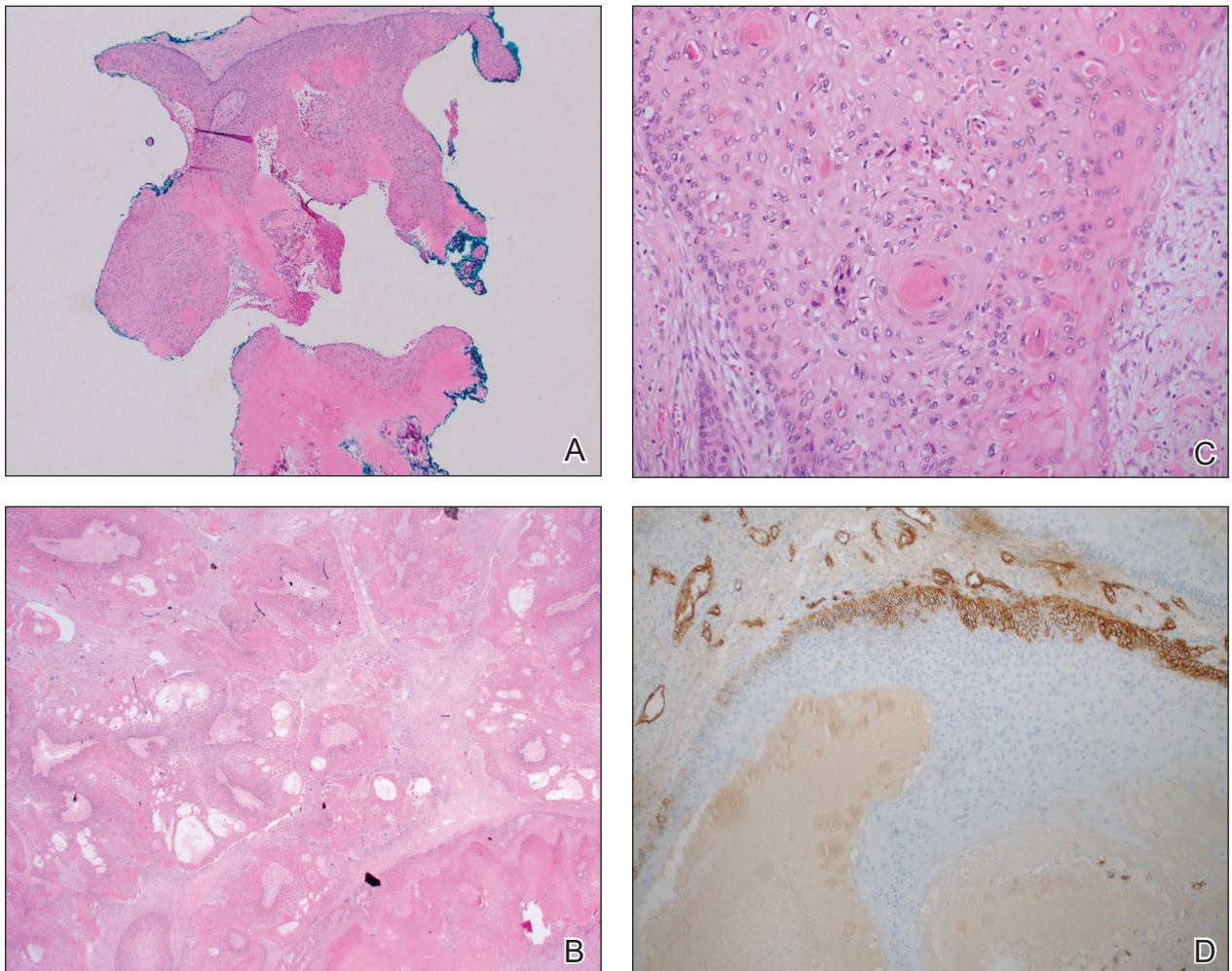


FIGURE. A and B, Histopathology revealed a partially cystic lesion transected at the base as well as lobulated masses of proliferating epithelium with trichilemmal keratinization (no granular layer)(H&E, original magnification $\times 4$ and $\times 10$). C, Higher magnification revealed atypical keratinocytes, apoptotic cells, and nuclear pleomorphism (H&E, original magnification $\times 20$). D, Immunohistochemistry revealed positive CD34 staining, indicating trichilemmal differentiation (original magnification $\times 20$).

proliferating epidermoid cysts, noting their distinct histologic features and resemblance to SCC.² These tumors generally are benign and arise from the isthmus of the outer root sheath of the hair follicle; however, malignant transformation can occur, resulting in a rare entity known as MPTT. This malignant variant was first described in 1983 by Saida et al,³ who emphasized its distinct clinical behavior, including infiltrative growth, high mitotic activity, and potential for local recurrence and metastasis.

A recent literature review identified 60 reported cases of MPTT, with an average patient age of 57 years and a female predominance.⁴ Clinically, MPTTs often manifest as large (>5 cm) lobulated masses located on sun-exposed, hair-bearing areas of the skin, especially the scalp. These lesions may be flesh-colored to pink and often exhibit ulceration, necrosis, or calcification.⁵ Typically, MPTTs follow a biphasic course, beginning with a slow-growing phase followed by a period of rapid growth. Due to their aggressive behavior and resemblance to other cutaneous malignancies, accurate differentiation of MPTT from benign PTTs, cutaneous SCCs, SCs, and trichilemmal carcinomas is critical.

Malignant proliferating trichilemmal tumors demonstrate a substantially higher metastatic potential than either benign PTTs or cutaneous SCCs. While cutaneous SCCs carry a metastasis rate of approximately 1.9% to 2.6%, MPTTs carry a considerably higher rate of approximately 25.0%.⁶ Regional lymphatic spread is the most common route of dissemination, making comprehensive lymph node assessment—both radiographic and clinical—an important component of tumor staging. When lymph node involvement is suspected, surgical dissection may be indicated, along with consideration of adjuvant therapies.

Histopathologically, MPTT is characterized by nuclear atypia, mitotic figures, and lobulated masses of proliferating epithelium showing trichilemmal differentiation and infiltrative growth.⁴ The presence of CD34 positivity, reflecting outer follicular root sheath differentiation, helps distinguish MPTT from cutaneous SCC and SC, which typically lack this marker.^{6,7} Immunohistochemistry is therefore a valuable adjunct in differentiating these lesions.

The mainstay of treatment for MPTT is wide local excision with clear margins. Margins of at least 1 cm generally are recommended. Although Mohs micrographic surgery may be used in anatomically sensitive areas, it typically is not preferred due to the potential for skip lesions in MPTT, which may lead to incomplete excision and recurrence.⁸ In cases with evidence of regional or distant metastasis or when clear margins cannot be achieved confidently, adjunctive treatments such as radiation therapy and systemic chemotherapy may be indicated. Preoperative imaging is used to evaluate for local invasion (skull or bone involvement) and regional lymph node status, which may inform adjuvant therapy postoperatively.

The prognosis for MPTT is variable and depends largely on early recognition, accurate histopathologic diagnosis, complete surgical excision with clear margins, and the presence or absence of metastasis. When the tumor is fully excised with negative margins and no lymph node involvement, the risk for recurrence is substantially reduced; however, MPTT is known for its potential aggressive behavior. Delays in diagnosis or incomplete resection can lead to local recurrence, regional spread, or even distant metastasis. In the literature review discussed previously, the mortality rate of patients with MPTT was 11.7%,⁴ which is notably higher than that of more common cutaneous malignancies such as cutaneous SCC, which is reported at 1.2%.⁹

The clinical course of MPTT remains difficult to predict due to its rarity and the limited availability of large-scale studies. Most published data are derived from isolated case reports or small case series, making standardized treatment guidelines challenging. Given this uncertainty, long-term follow-up is strongly recommended to monitor for recurrence or metastatic progression.²

This case highlights the critical role of clinicopathologic correlation in the evaluation of atypical or rapidly growing scalp lesions. The expertise of dermatologists in recognizing atypical presentations, combined with precise histopathologic analysis, including immunohistochemical staining, is vital to ensuring accurate diagnosis and optimal treatment. Early intervention can improve patient outcomes by reducing the risk for local recurrence and metastatic progression as well as the need for more intensive therapies.

REFERENCES

1. Jones EW. Proliferating epidermoid cysts. *Arch Dermatol*. 1966;94:11-19.
2. Kemaloglu CA, Öztürk M, Aydın B, et al. Malignant proliferating trichilemmal tumor of the scalp: report of 4 cases and a short review of the literature. *Case Reports Plast Surg Hand Surg*. 2022;9:158-164. doi:10.1080/23320885.2022.2077208
3. Saida T, Oohard K, Hori Y, et al. Development of a malignant proliferating trichilemmal cyst in a patient with multiple trichilemmal cysts. *Dermatology*. 1983;166:203-208. doi:10.1159/000249868
4. Abdelhammed MH, Siatecka H, Diwan AH, et al. A rare case of a malignant proliferating trichilemmal tumor: a molecular study harboring potential therapeutic significance and a review of literature. *Dermatopathology (Basel)*. 2024;11:354-363. doi:10.3390/dermatopathology11040038
5. Fronck L, Brahs A, Farsi M, et al. A rare case of trichilemmal carcinoma: histology and management. *J Clin Aesthet Dermatol*. 2021;14:25-30.
6. Osto M, Parry N, Rehman R, et al. Malignant proliferating trichilemmal tumor of the scalp: a systematic review. *Am J Dermatopathol*. 2021;43:851-866. doi:10.1097/DAD.0000000000001991
7. Plaza JA, Mackinnon A, Carrillo L, et al. Role of immunohistochemistry in the diagnosis of sebaceous carcinoma: a clinicopathologic and immunohistochemical study. *Am J Dermatopathol*. 2015;37:809-821. doi:10.1097/DAD.0000000000000255
8. Singh P, Usman A, Motta L, et al. Malignant proliferating trichilemmal tumour. *BMJ Case Rep*. Published online August 17, 2018. doi:10.1136/bcr-2018-224460
9. Ran NA, Granger EE, Brodland DG, et al. Risk factor number and recurrence, metastasis, and disease-related death in cutaneous squamous cell carcinoma. *JAMA Dermatol*. 2025;161:597-604. doi:10.1001/jamadermatol.2025.0128