

Metastatic Meningioma of the Scalp

Dhaval G. Bhanusali, MD; Candrice Heath, MD; Deniz Gur, MD; Sheryl Miller, MD

PRACTICE POINTS

- Meningiomas are common intracranial neoplasms that generally present with a benign course.
- Rarely, meningiomas exhibit malignant potential and can present as extracranial cutaneous neoplasms.
- Patients presenting with nodules on the scalp warrant a thorough medical history and consideration beyond common cysts and lipomas.

Meningiomas generally present as slow-growing, expanding intracranial lesions and are the most common benign intracranial tumor in adults. Rarely, meningiomas can exhibit malignant potential and present as extracranial soft-tissue masses through extension or as primary extracranial cutaneous neoplasms. Although they are uncommonly encountered by dermatologists, it is important to include meningioma in the differential diagnosis for scalp neoplasms. We present a rare case of a 68-year-old woman with scalp metastasis of meningioma 11 years after initial resection of the primary tumor.

Cutis. 2018;101:386-389.

Meningiomas generally present as slow-growing, expanding intracranial lesions and are the most common benign intracranial tumor in adults.¹ Rarely, meningioma exhibits malignant potential and presents as an extracranial soft-tissue mass through extension or as a primary extracranial cutaneous neoplasm. The differential diagnosis of scalp neoplasms must be broadened to include uncommon tumors such as meningioma. We present a rare case of a 68-year-old woman with scalp metastasis of meningioma 11 years after initial resection of the primary tumor.

Case Report

A 68-year-old woman presented for evaluation of an asymptomatic nodule on the left parietal scalp of 2 years'

duration. She denied any headaches, difficulty with balance, vision changes, or changes in mentation. Her medical history was remarkable for a benign meningioma removed from the right parietal scalp 11 years prior without radiation therapy, as well as type 2 diabetes mellitus and arthritis. The patient's son died from a brain tumor, but the exact tumor type and age at the time of death were unknown. Her current medications included metformin, insulin glargine, aspirin, and a daily multivitamin. She denied any allergies or history of smoking.

Physical examination of the scalp revealed 4 fixed, nontender, flesh-colored nodules: 2 on the left parietal scalp measuring 3.0 cm and 0.8 cm, respectively (Figure 1A); a 0.4-cm nodule on the right posterior occipital scalp; and a 1.6-cm sausage-shaped nodule on the right temple (Figure 1B). No positive lymph nodes were appreciated, and no additional lesions were noted. No additional atypical lesions were noted on full cutaneous examination.

A diagnostic 6-mm punch biopsy of the largest nodule was performed. Intraoperatively, there was no apparent cyst wall, but coiled, loose, stringlike, pink-yellow tissue was removed from the base of the wound before closing with sutures.

The primary histologic finding was cells within fibrous tissue containing delicate round-oval nuclei, inconspicuous nucleoli, and lightly eosinophilic cytoplasm with an indistinct border (Figure 2). Immunohistochemical studies for S100 protein were focal and limited to the cytoplasm of a subset of neoplastic cells (Figure 3). Tumor cells stained positive for epithelial membrane antigen (EMA) and were focally positive for progesterone receptor (Figure 4). Tumor cells were negative for CD31 and CD34. Based on the clinical and histologic findings, a diagnosis of metastatic meningioma of the scalp was made.

Magnetic resonance imaging and positron emission tomography of the head, neck, and chest demonstrated 3 residual subcutaneous nodules on the scalp and an indeterminate subcentimeter nodule in the right lung.

From the Department of Dermatology, Icahn School of Medicine at Mount Sinai, New York, New York.

The authors report no conflict of interest.

Correspondence: Dhaval G. Bhanusali, MD, Icahn School of Medicine at Mount Sinai, 5 E 98th St, 5th floor, New York, NY 10029 (drbhanusali@bhanusali.md.com).

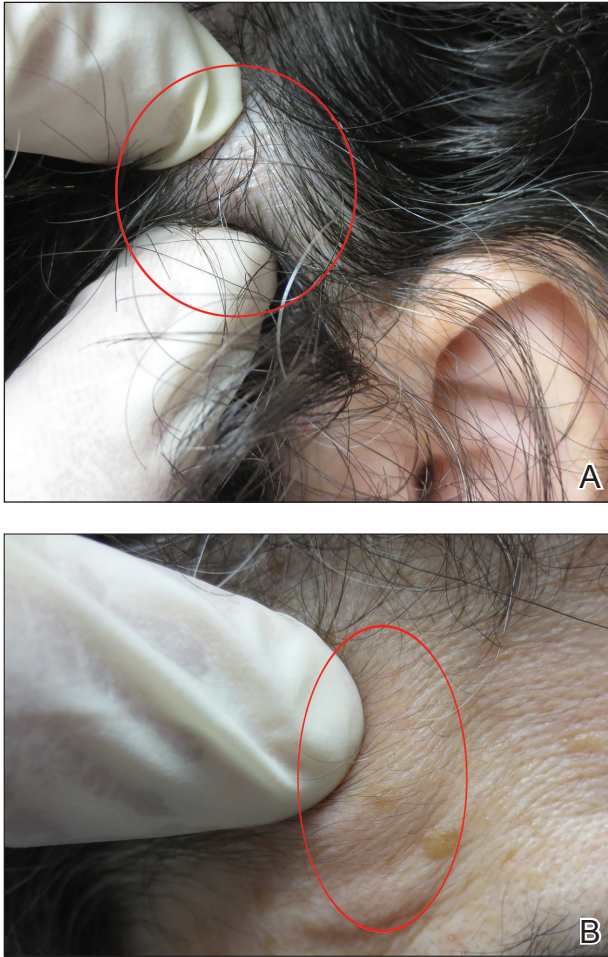


FIGURE 1. Metastatic meningioma presenting as a firm nodule on the left parietal scalp (A) and a 1.6-cm, sausage-shaped nodule on the right temple (B).

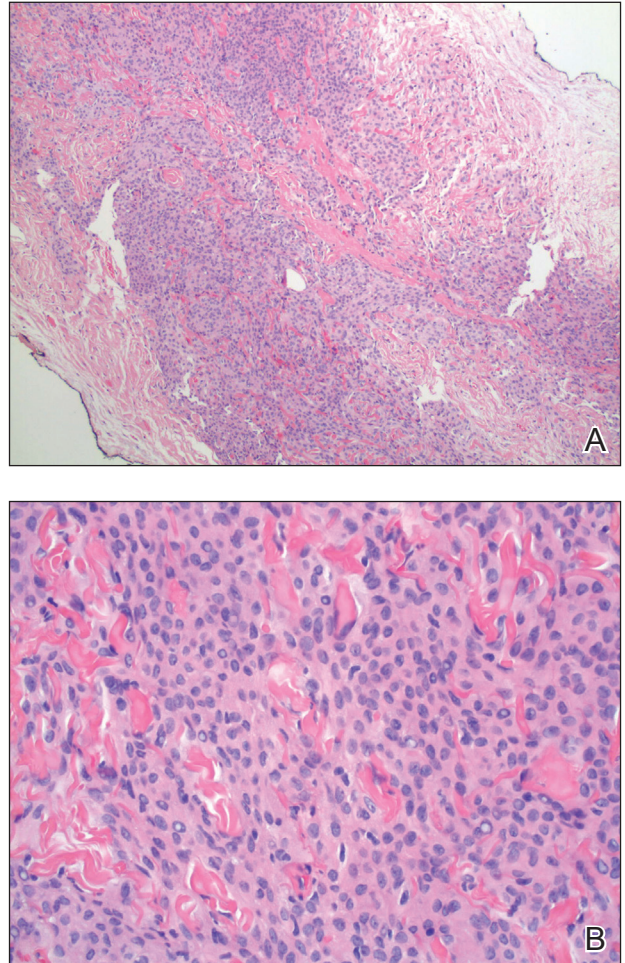


FIGURE 2. Tumor cells forming whorl-like structures in a metastatic meningioma (A)(H&E, original magnification $\times 4$). The tumor cells had round-oval nuclei, inconspicuous nucleoli, lightly eosinophilic cytoplasm, and indistinct cytoplasmic borders (B)(H&E, original magnification $\times 40$).

The 0.4-cm nodule on the right posterior occipital scalp was removed without complication, and no radiation therapy was administered. The rest of the lesions were monitored. She remained under the close observation of a neurosurgeon and underwent repeat imaging of the scalp nodules and lungs, initially at 3 months and then routinely at the patient's comfort. The patient currently denies any neurologic symptoms.

Comment

Meningiomas are derived from meningotheial cells found in the leptomeninges and in the choroid plexus of the ventricles of the brain.² They are common intracranial neoplasms that generally are associated with a benign course and present during the fourth to sixth decades of life. Meningiomas constitute 13% to 30% of intracranial neoplasms and usually are female predominant (3:1).^{3,4} Rarely, malignant transformation can lead to local and distant metastasis to the lungs,^{5,6}

liver,⁷ and skeletal system.⁸ In cases of metastatic spread, there is an increased incidence in males versus females.⁹⁻¹¹

Risk Factors—Although many meningiomas are sporadic, numerous risk factors have been associated with the disease development. One study showed a link between exposure to ionizing radiation and subsequent development of meningioma.¹² Another study found a population link between a higher incidence of meningioma and nuclear exposure in Hiroshima, Japan, after the atomic bomb blast in 1980.¹³ There is an increased incidence of meningioma in patients exposed to radiography from frequent dental imaging, particularly when older machines with higher levels of radiation exposure are used.¹⁴

Another study demonstrated a correlation between meningioma and hormonal factors (eg, estrogen for hormone therapy) and exacerbation of symptoms during pregnancy.¹⁵ There also is an increased incidence of meningioma in breast cancer patients.⁴ Genetic

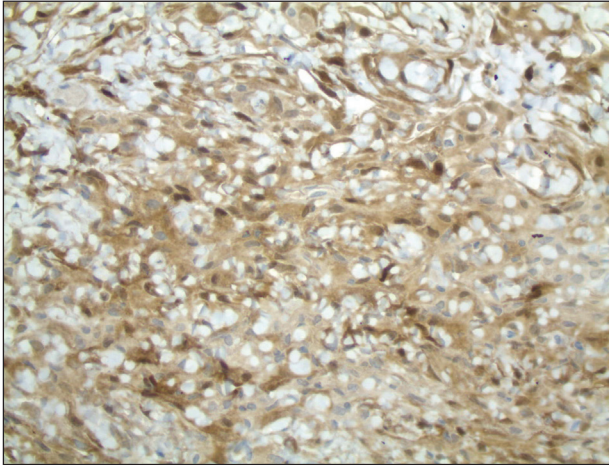


FIGURE 3. Areas of focal S100 protein positivity in a metastatic meningioma on the scalp (original magnification $\times 40$).

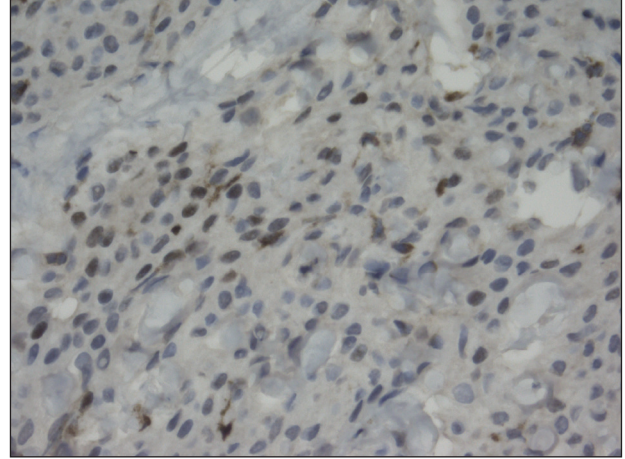


FIGURE 4. Tumor cells in a metastatic meningioma on the scalp were focally positive for progesterone receptor (original magnification $\times 40$).

alterations also have been implicated in the development of meningioma. It was found that 50% of patients with a mutation in the neurofibromatosis 2 gene (which codes for the merlin protein) had associated meningiomas.^{16,17} Scalp nodules in patients with neurofibromatosis type 2 increases suspicion of a scalp meningioma and necessitates biopsy.

Clinical Presentation—Cutaneous meningiomas typically present as firm, subcutaneous nodules. Scalp nodules ranging from alopecia^{18,19} to hypertrichosis²⁰ have been reported. These neoplasms can be painless or painful, depending on mass effect and location.

Classification—The primary clinical classification system of metastatic meningioma was first described in 1974.²¹ Type 1 meningioma refers to congenital lesions that tend to cluster closer to the midline. Type 2 refers to ectopic soft-tissue lesions that extend to the skin from likely remnants of arachnoid cells. These lesions are more likely to be found around the eyes, ears, nose, and mouth. Type 3 meningiomas extend from intracranial tumors that secondarily involve the skin through proliferation through bone or anatomic defects. Type 3 is the result of direct extension and the location of the cutaneous presentation depends on the location of the intracranial lesion.^{4,22,23}

Pathology—Meningiomas exhibit a range of morphologic appearances on histopathology. In almost all meningiomas, tumor cells are concentrically wrapped in tight whorls with round-oval nuclei and delicate chromatin, central clearing, and pale pseudonuclear inclusions. Lamellate calcifications known as psammoma bodies are a common finding. Immunohistochemical studies show that most meningiomas are positive for EMA, vimentin, and progesterone receptor. S100 protein expression, if present, usually is focal.

Differential Diagnosis—Asymptomatic nodules of the scalp may present a diagnostic challenge to physicians.

Most common scalp lesions tend to be cystic or lipomatous. In children, a broad differential diagnosis should be considered, including dermoid and epidermoid tumors, dermal sinus tumors, hemangiomas, metastasis of another tumor, aplasia cutis congenita, pilomatricoma, and lipoma. In adults, the differential should focus on epidermoid cysts, lipomas, metastasis of other tumors, osteomas, arteriovenous fistulae, and heterotopic brain tissue. Often, microscopic examination is necessary, along with additional immunohistochemical staining (eg, EMA, vimentin).

Treatment—Treatment options for meningioma include observation, surgical resection, radiotherapy, and systemic therapy, as well as a combination of these modalities. The choice of therapy depends on such variables as patient age; performance status; comorbidities; presence or absence of symptoms (including focal neurologic deficits); and tumor location, size, and grade. It is important to note that there is limited knowledge looking at the results of various treatment modalities, and no consensus approach has been established.

Conclusion

Our patient's medical history was remarkable for an intracranial meningioma 11 years prior to the current presentation, and she was found to have biopsy-proven metastatic meningioma without recurrence of the initial tumor. Patients presenting with a scalp nodule warrant a thorough medical history and consideration beyond common cysts and lipomas.

REFERENCES

1. Mackay B, Bruner JM, Luna MA. Malignant meningioma of the scalp. *Ultrastruc Pathol.* 1994;18:235-240.
2. Whittle IR, Smith C, Navoo P, et al. Meningiomas. *Lancet.* 2004; 363:1535-1543.
3. Bauman G, Fisher B, Schild S, et al. Meningioma, ependymoma, and other adult brain tumors. In: Gunderson LL, Tepper JE, eds. *Clinical*

- Radiation Oncology*. Philadelphia, PA: Elsevier Churchill Livingstone; 2007:539-566.
4. Claus EB, Bondy ML, Schildkraut JM, et al. Epidemiology of intracranial meningioma. *Neurosurgery*. 2005;57:1088-1095.
 5. Tworek JA, Mikhail AA, Blaivas M. Meningioma: local recurrence and pulmonary metastasis diagnosed by fine needle aspiration. *Acta Cytol*. 1997;41:946-947.
 6. Shin MS, Holman WL, Herrera GA, et al. Extensive pulmonary metastasis of an intracranial meningioma with repeated recurrence: radiographic and pathologic features. *South Med J*. 1996;89:313-318.
 7. Ferguson JM, Flinn J. Intracranial meningioma with hepatic metastases and hypoglycaemia treated by selective hepatic arterial chemoembolization. *Australas Radiol*. 1995;39:97-99.
 8. Palmer JD, Cook PL, Ellison DW. Extracranial osseous metastases from intracranial meningioma. *Br J Neurosurg*. 1994;8:215-218.
 9. Glasauer FE, Yuan RH. Intracranial tumours with extracranial metastases. case report and review of the literature. *J Neurosurg*. 1963;20:474-493.
 10. Shuangshoti S, Hongsaprabhas C, Netsky MG. Metastasizing meningioma. *Cancer*. 1970;26:832-841.
 11. Ohta M, Iwaki T, Kitamoto T, et al. MIB-1 staining index and scoring of histological features in meningioma. *Cancer*. 1994;74:3176-3189.
 12. Wrensch M, Minn Y, Chew T, et al. Epidemiology of primary brain tumors: current concepts and review of the literature. *Neuro Oncol*. 2002;4:278-299.
 13. Shintani T, Hayakawa N, Hoshi M, et al. High incidence of meningioma among Hiroshima atomic bomb survivors. *J Rad Res*. 1999;40:49-57.
 14. Claus EB, Calvocoressi L, Bondy ML, et al. Dental x-rays and risk of meningioma. *Cancer*. 2012;118:4530-4537.
 15. Blitshteyn S, Crook JE, Jaeckle KA. Is there an association between meningioma and hormone replacement therapy? *J Clin Oncol*. 2008;26:279-282.
 16. Fontaine B, Rouleau GA, Seizinger BR, et al. Molecular genetics of neurofibromatosis 2 and related tumors (acoustic neuromas and meningioma). *Ann N Y Acad Sci*. 1991;615:338-343.
 17. Rabin BM, Meyer JR, Berlin JW, et al. Radiation-induced changes of the central nervous system and head and neck. *Radiographics*. 1996;16:1055-1072.
 18. Tanaka S, Okazaki M, Egusa G, et al. A case of pheochromocytoma associated with meningioma. *J Intern Med*. 1991;229:371-373.
 19. Zeikus P, Robinson-Bostom L, Stopa E. Primary cutaneous meningioma in association with a sinus pericranii. *J Am Acad Dermatol*. 2006; 54(2 suppl):S49-S50.
 20. Junaid TA, Nkposong EO, Kolawole TM. Cutaneous meningiomas and an ovarian fibroma in a three-year-old girl. *J Pathol*. 1972; 108:165-167.
 21. Lopez DA, Silvers DN, Helwig EB. Cutaneous meningioma—a clinicopathologic study. *Cancer*. 1974;34:728-744.
 22. Shuangshoti S, Boonjunwetwat D, Kaoroptham S. Association of primary intraspinal meningiomas and subcutaneous meningioma of the cervical region: case report and review of literature. *Surg Neurol*. 1992;38:129-134.
 23. Miedema JR, Zedek D. Cutaneous meningioma. *Arch Pathol*. 2012;136:208-211.