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A Solitary Lump on the Scalp

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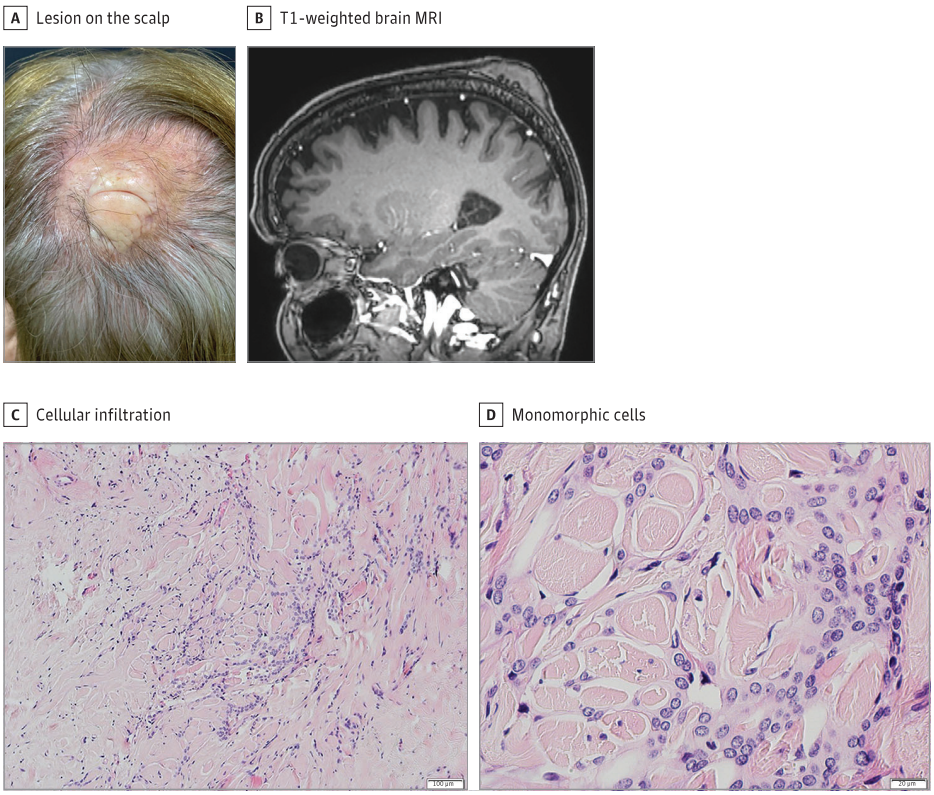

Figure. A, Clinical image of the lesion on the scalp. B, Paramedian sagittal gadolinium-enhanced T1-weighted brain magnetic resonance imaging (MRI). C, Cellular infiltration in the reticular dermis (hematoxylin-eosin, original magnification ×100). D, Monomorphic cells in a syncytial configuration surrounding collagen bundles (hematoxylin-eosin, original magnification ×400).

A woman in her early 70s was referred to the dermatology department for evaluation of a solitary, asymptomatic lump on her scalp. She was born with the lesion but noticed some growth following a minor local trauma a couple of months prior to seeking medical evaluation. The patient denied local inflammation, purulent discharge, and systemic symptoms. Her medical history was unremarkable apart from an allegedly benign breast nodule resection in her teenage years. In addition, her father died of metastatic cutaneous melanoma at 83 years old.

Clinically, there was an ill-defined, infiltrated plaque with a normochromic, exophytic nodule on her scalp vertex measuring 4.2 × 3.8 cm (**Figure**, A). It had a waxy surface with irregular transverse furrows and overlying alopecia. Dermoscopic evaluation findings revealed a homogeneous yellow hue with no visible vascular or pigmented structures and absence of follicular ostia. The lesion was firm and mildly tender to the touch and seemed adherent to the underlying bone. There were no palpable lymph nodes. A brain magnetic resonance imaging was performed for further elucidation (Figure, B), as well as a punch biopsy (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Chondroid syringoma

B. Cutaneous meningeal heterotopia

C. Scalp metastasis

D. Proliferative nodule in a congenital melanocytic nevus

Diagnosis

B. Cutaneous meningeal heterotopia

Discussion

Brain magnetic resonance imaging demonstrated a nonenhancing tumor with sharp margins in the subcutaneous tissue of the scalp with no underlying bone or intracranial involvement (Figure, B). Histopathologic study findings revealed an infiltration by monomorphic cells with ovoid nuclei and eosinophilic cytoplasm amid a fibrous stroma in the lower dermis (Figure, C). Cells did not display discernible cell membranes and tended to form whorls and strands around collagen bundles (Figure, D). There were no adnexal structures overlying the lesion, and the epidermis was spared. Immunostaining was strongly positive for vimentin and epithelial membrane antigen (EMA), focally positive for progesterone receptor, and showed an estimated 1% positivity for Ki-67. Cells were negative for cytokeratins AE1/AE3, S100, CD45, CD68, factor XIII, actin, glial fibrillary acidic protein, and octamer-binding transcription factor 4. The clinicopathologic correlation was compatible with the diagnosis of a cutaneous meningeal heterotopia (CMH).

Extracranial meningothelial tissue can be found in a spectrum of malformations caused by abnormal migration and entrapment of meningothelial tissue outside the central nervous system during embryogenesis.[1] Numerous nomenclatures have been used interchangeably in the medical literature to describe this condition, but currently *cutaneous meningeal heterotopia* is preferred over terms such as *primary cutaneous meningioma* and *meningothelial hamartoma*.[2,3]

Cutaneous meningeal heterotopia is suspected when there is a congenital fibrotic plaque or nodule with overlying alopecia on the scalp[2,3] or lumbosacral region.[2-4] Lesions are usually asymptomatic or mildly tender and do not display significant growth over time.[2,3] Clinically, CMH can resemble aplasia cutis congenita or congenital hamartomas, such as nevus sebaceus of Jadassohn. Therefore, histopathologic study is essential for establishing the diagnosis. In CMH, monomorphic meningothelial cells arranged in strands and pseudovascular structures interspersed with a fibrous collagenous stroma spanning the dermis and subcutaneous tissue can be seen.[1,2] There is usually a marked reduction in overlying pilosebaceous units.[2] Syncytial aggregates with large cuboidal cells in a whorled pattern and encirclement of collagen bundles reminiscent of classic intracranial meningiomas with psammomatous bodies are typical but may be absent.[3,5] Meningothelial cells stain positive for EMA, vimentin, and neuron-specific enolase but negative for S100, CD31, CD34, and factor VIII–related antigen on immunohistochemistry.[2,5]

Primary and metastatic neoplasms on the scalp are distinguished from CMH based on clinical and histopathologic features. Scalp metastases at birth are exceedingly rare and usually show rapid growth and distinct microscopic morphologic features with conspicuous cellular atypia.[3,6] Chondroid syringomas are adnexal neoplasms that present as indistinctive, slow-growing cutaneous nodules. Nevertheless, they show prominent tubular and cystic structures embedded in a basophilic chondromyxoid stroma and a distinctive immunohistochemistry profile.[7] Proliferative nodules arising within congenital melanocytic nevi can be hypopigmented and have an indolent course. However, they are composed of focal cellular areas of large epithelioid melanocytes with variable nuclear pleomorphism within a preexisting congenital nevus.[8]

Surgical resection with exiguous margins is curative for CMH.[5,9] Imaging studies are recommended before invasive procedures to exclude the presence of a residual stalk connecting the lesion to the central nervous system and associated bone defects, which occur in a minority of cases.[3-5] There is no consensus on the ideal timing for the exeresis. However, most reported patients have been treated in their first decade (age range, 3 weeks to 78 years).[1-3,5,10] Prognosis is excellent, with no reported postoperative recurrences. The patient in this case opted not to undergo surgical treatment after being informed of the diagnosis. The lesion remained stable and asymptomatic after a 6-month follow-up.

ARTICLE INFORMATION

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