Type II Cutaneous Menigioma: A Case Report and Discussion
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Introduction
Meningiomas represent the most common intracranial tumors, while cutaneous menigiomas are rare tumors most commonly occurring on the scalp. Cutaneous menigiomas are thought to arise from the herniation of meningeal lining during embryonic development into the dermis and subcutaneous tissue. They are classified into three subtypes; type I tumors arise congenitally, while type II and III tumors represent multipotent mesenchymal/ectopic arachnoid cells and direct extension of an underlying tumor of the arachnoid lining, respectively.1,2

Case
Here we report a 40-year-old female who presented with an enlarging, itchy, painful mass on the posterior mid-parietal scalp (Fig. 1). This lesion had been slowly enlarging over a 2 year period. The patient denied trauma to this area. Past medical history was significant for hypothyroidism and remote seizures. Surgical, family, and social history were non-contributory. On exam, the lesion was firm and poorly mobile with a normal appearing overlying epidermis. Initial differential diagnosis included an epidermal inclusion cyst, pilonidal cyst and an encapsulated lipoma. The patient elected for surgical removal. Intraoperatively, the lesion was poorly encapsulated, caseous friable, and non-odoriferous. No communication with the galea aponeurotica or cranium was noted. The specimen was sent for pathological diagnosis.

Histology
Whorls of meningothelial cells and few psammoma bodies were noted on 4X magnification in Figure 2. On higher magnification (10X), a psammoma body is depicted well in Figure 3. Positive staining with epithelial membrane antigen (EMA) (Fig. 4) and lack of staining with CD34, desmin, factor Xilla, and pan-keratin (AE-1/AE-3) confirmed the diagnosis of a cutaneous menigioma. Typical histology of menigiomas include meningothelial cells whorling around focal areas of collagen, and psammoma bodies are often noted.4

Discussion
Meningiomas constitute 15-25% of tumors of the central nervous system, making them the most common intracranial/intraspinal neoplasm. They are derived from arachnoid membrane cells. Rarely these neoplasms are seen cutaneously. Differential diagnosis of cutaneous menigiomas may include an epidermal inclusion cyst, acrochordon, lipoma, pilonidal cyst, fibroma, or a vascular lesion. Lopez et al. subclassified cutaneous menigiomas into 3 subsets. Type I lesions occur congenitally and can be distinguished histologically as they tend to have a collagenous stroma. In addition, they tend to be located in the subcutaneous fat, while type II and III lesions typically extend into the dermis. Type II lesions are acquired and represent ectopic arachnoid tissue. Type III tumors are also acquired and arise via direct extension of an intracranial menigioma. Distinguishing between type II and III cutaneous menigiomas cannot be made histologically. This differentiation requires neuroimaging to determine if the cutaneous lesion is an extension of an intracranial menigioma, or by lack of infiltration of the tumor to the galea aponeurotica and/or cranium on excision.

Classic pathological examination of primary intracranial menigiomas reveal psammoma bodies and meningothehial whorls. These findings are not always appreciated in their cutaneous counterparts, and can pose a diagnostic challenge on behalf of the pathologist. Thus, positive staining with vimentin and EMA are considered diagnostic, while lack of desmin, cytokeratin, CD31, CD34, CD68, and smooth muscle antigen rule out other histological differential diagnosis; including hemangiopericytomas, giant cell fibroblastoma, squamous cell carcinoma, hemangioma, and heterotopic neruogliomas.

Prognosis of cutaneous menigiomas are largely determined based on type. Type I lesions have a favorable prognosis if clear surgical margins are achieved intraoperatively. Type II and type III lesions tend to have a poorer prognosis as these lesions may lack classic histological features of congenital menigiomas as mentioned above. In addition, type III lesions may be inoperable if their intracranial direct extension abuts vital structures.

Currently imaging is recommended prior to surgical excision of known cutaneous menigiomas, as intracranial extension requires removal by a neurosurgeon. Although cutaneous menigiomas represent a rare entity, maintaining a low threshold for biopsy of atypical scalp lesions may be warranted prior to surgical removal.

References