Cutaneous meningioma

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Cutaneous meningioma is a rare tumor that most commonly occurs on the scalp and occurs in both congenital and acquired forms. It involves a wide clinical differential diagnosis, but diagnosis is based on characteristic histologic and cytologic findings. Congenital lesions can often present years after birth and even in adult patients. Acquired lesions occur in adulthood. We review histologic, cytologic, and electron microscopic findings and explore how these are used to separate this entity from other entities in the differential diagnosis. While ultrastructural and cytologic findings are similar to those of more common intracranial meningiomas, these tumors exhibit a range of histologic differences. A lack of awareness of this entity precludes correct diagnosis. (Arch Pathol Lab Med. 2012;136:208–211; doi: 10.5858/arpa.2010-0505-RS)

Meningioma is one of the most common neural tumors of adults and is the most common extracerebral intracranial tumor, generally presenting as a slowly expanding intracranial lesion. These tumors are typified by a classic meningeal location, which helps to facilitate diagnosis. Most of these tumors surround the brain or ventricles and are benign. Tumors exhibit a wide range of clinical, gross, and histologic findings. A particularly fascinating presentation is that of the cutaneous meningioma. The first known reported case of this lesion was in 1904, while its first description in the English literature was in 1956. It is a rare finding that is now more widely recognized. Variations of cutaneous meningioma have been described as cutaneous heterotopic meningeal nodule and cutaneous meningeal hamartoma or meningothelial hamartoma. The largest and much cited report by Lopez et al in 1974 provides the basis for the classification system; however, most of the literature regarding these tumors is in the form of case reports, reflecting their relative rarity.

DEVELOPMENT AND CLASSIFICATION

The widely used classification system for cutaneous meningioma was developed by Lopez et al. The authors collected and studied 25 cases of cutaneous meningioma. They divided these tumors into 3 types based on different etiologies that, in turn, shape the characteristics of each tumor type.

The first type (type I) is the congenital type that is present at birth and generally occurs on the scalp and paravertebral regions. Type I tumors develop from ectopic arachnoid cells (meningothelial cells) that become trapped in the dermis and subcutis during development. This results from the failure of neural tube closure to completely trap all neural elements, which leads to ectopic neural tissue by a mechanism similar to that of heterotopic glial nodules.

Because of this mechanism, rudimentary meningoceles and acocelic meningeal hamartomas most likely represent 2 ends of a spectrum, and many authors simply prefer the term cutaneous meningioma. This developmental mechanism has been supported by the occasional finding of a sinus track connecting the central nervous system and the lesion upon surgical exploration, as well as the often midline location of the lesion. It is worth pointing out, however, that this “meningocele mechanism,” while generally accepted, has not been without controversy. In fact, for some congenital lesions, some authors support a mechanism in which meningeal cells are displaced along nerves during embryogenesis (similar to the mechanism for type II lesions)

The second type (type II) consists of ectopic soft tissue meningiomas that extend to the skin by contiguity. These tend to occur around the eyes, ears, nose, and mouth. An important finding in this type of cutaneous meningioma is the lack of corresponding meningioma of the neuroaxis. Lopez et al hypothesized that these tumors were formed by remnants of arachnoid cells, which extend along cranial nerves.

Type III cutaneous meningiomas are tumors that have extended into the dermis or subcutis from a meningioma that involves the neuroaxis (ie, a primary intracranial meningioma). Type III lesions are primary meningeal tumors that secondarily involve the skin by direct extension of underlying tumors through bone, traumatic defects, or surgical defects. These tumors are, therefore, much more common in adults.

CLINICAL FEATURES

Lesions present as firm, subcutaneous nodules ranging in color from pallid to slightly dark. Lesions have been described as alopecic, with overlying tufts of hair or even hypertrichosis. While generally painless, in some cases, pain or tenderness has been described. One lesion was described as pedunculated. Type I tumors are congenital, whereas types II and III develop later in life. In an article published in 1992, the authors collected age and site characteristics associated with 92 primary cutaneous meningiomas and found that these tumors...
tended to present in younger patients with an average age of 34 and a female to male ratio of 4:5. Age characteristics are potentially clouded by the fact that many lesions probably were present since birth but only received medical attention later, at times after a period marked by growth. This is in contrast to intracranial meningiomas for which the incidence tends to increase with age.

Type II and III cutaneous meningiomas occur in adults as de novo lesions. In our literary review, 7 patients who were specifically diagnosed with type II lesions had a mean age of 48 years (range, 27–61 years). Of 4 patients specifically diagnosed as having type III lesions, the mean age was 54 years, with a range of 20 to 68 years.

Type II lesions occur around sensory organs of the head and along the course of cranial and spinal nerves. Type III lesions can occur on the face, temple, and scalp as slow-growing subcutaneous masses. Tumors in special locations may also present with symptoms specific to these locations. For example, periorbital meningiomas may present with proptosis, periordal edema, and lid induration. Paranasal meningiomas may present with sinusitis or rhinitis. These noncongenital tumors develop in areas of trauma.

GROSS PATHOLOGY

Primary lesions present grossly as a solitary, firm, gray/white subcutaneous nodule ranging in size from a few millimeters to several centimeters. If the tumor exhibits a deep connection, there may be remnant fluid discharge. Lesions demonstrating both alopecia and hypertrichosis have been described. Depending on the depth of the tumor, there may be involvement of underlying soft tissue, fascia, or bone. Rudimentary cystic cavities were seen in 3 patients in the series of Lopez and colleagues. An intimate association with nerves is observed with some lesions.

Type II or III lesions with erosion through bone have been reported. These acquired lesions often extend more superficially than their primary counterparts.

HISTOPATHOLOGY

In regard to histologic findings, the type I lesion reported by Lopez et al deserves special mention, as several variants have been described and the authors themselves identified histologic variants in their study. Of type I lesions, 6 had the more traditional histologic findings of meningothelial cells, often in nests and with psammoma bodies, as well as structures resembling psammoma bodies but lacking calcification, which they termed collagen bodies. Seven more lesions, termed type I variants or acoelic meningeal hamartomas, differed from typical type I lesions by being less circumscribed and by having scattered foci of hyperplastic meningothelial cells and psammoma bodies, often in streaks. Three final lesions falling under the type I classification were referred to as rudimentary meningocytes because of the presence of a rudimentary cystic cavity or stalk with nests and strands of meningothelial cells. This spectrum of histologic findings is logical if one accepts the proposed genesis of type I lesions. As Lopez and colleagues proposed, all of these lesions fall within a spectrum and the differences in nomenclature reflect this concept. Because of this variation in architectural features, some emphasize the need to make the diagnosis based predominately on cytologic features. Hematoxylin-eosin staining of a type I primary lesion with hamartomatous features (admixed collagen, corded and streaked architecture) is shown in the Figure, A through C.

Acquired lesions (types II and III) tend to have similar cytologic detail as congenital lesions but usually have less collagen, more lobulated and cellular, and extend higher into the dermis. The number of inflammatory cells is also more variable. Well-developed collagen bodies are more typical of type I lesions.

CYTOPATHOLOGY

Kalfa et al reported 2 cases associated with a diagnosis of cutaneous meningioma via fine-needle aspiration. In both cases, the patients’ lesions smeared easily, and microscopic examination revealed characteristics typical of meningioma, including whorls and psammoma-body formation. Both tumors were positive for epithelial membrane antigen (EMA) and vimentin. Both patients lacked a primary intracranial tumor and therefore would have been classified as having either type I or type II lesions. Another group also found that the cytologic findings for the primary cutaneous meningioma were similar to findings characteristic of central nervous system meningioma, with bland nuclei, cellular whorls, and psammoma bodies. These authors advocated the use of cytologic smears if making the initial diagnosis on frozen section, especially for difficult cases.

A third group described the fine-needle aspiration findings as a secondary cutaneous meningioma on the scalp of a 45-year-old man with a lesion of 3 months’ duration (type III cutaneous meningioma). Fine-needle aspiration in this case revealed spindled cells in clusters and in a concentric arrangement, again producing characteristic whorls. Cells showed pale nuclei with finely granular, evenly distributed chromatin, and occasional intranuclear cytoplasmic invaginations. Diagnosis in this case was made on cytology.

ELECTRON MICROSCOPY

Ultrastructural features of cutaneous meningioma are similar to those of intracranial meningioma. One study evaluated a cutaneous meningioma by electron microscopy in a 32-year-old man with a congenital lesion. The authors described this tumor as consisting of intimately associated cells that contained complicated interdigitating processes, desmosomes, and hemidesmosomes, thus creating a jigsaw pattern. Another study described the findings from 2 cases. The first showed swollen and stellate meningothelial cells with round and oval nuclei, cytoplasm with a moderate number of microfilaments, straight and interdigitating cytomembranes, and desmosome-like junctions between cells. The authors also described “collagen bodies,” collagen bundles encompassed by meningothelial cells. The second case showed meningothelial cells with a moderately irregular shape and cytoplasm densely packed with microfilaments, arranged in a stepping-stone pattern in the collagenous matrix. Small electron-dense substances were present in the cytoplasm. These typical findings of meningothelial cells may be helpful in ruling out other types of lesions, particularly vascular endothelial proliferations, when electron microscopic analysis is available.

DIFFERENTIAL DIAGNOSIS AND IMMUNOHISTOCHEMISTRY

The clinical differential diagnosis is broad and includes a nevus sebaceous, cyst, fibroma, glioma, hemangioma, lipoma, scar, verrucous hamartoma, and alopecia.
areata, among others. This wide clinical differential diagnosis is to be expected, given the low incidence of this lesion.

Most epithelial tumors can be easily and readily distinguished by the presence of keratinization or follicular, sebaceous, or sweat gland differentiation. Otherwise, the histologic differential diagnosis includes squamous cell carcinoma, hemangioma, giant cell fibroblastoma, hemangiopericytoma, or other types of heterotopic neuroglial lesions, and even these are often easily ruled out. When the diagnosis of cutaneous meningioma enters the differential, careful microscopic examination with the judicious use of selected immunohistochemical stains, especially EMA (Figure, D), can confirm the diagnosis. Positivity for EMA and vimentin are widely regarded as supporting the diagnosis of meningioma, while lack of staining for cytokeratins has been used to rule out epithelial lesions.

In some cases, the cuboidal epithelioid cells of the ectopic meningothelial hamartoma may be intimately associated with vessels and have prominent, freely anastomosing vascular channels. In these cases the diagnosis of well-differentiated angiosarcoma is ruled out by expression of vimentin and EMA and lack of staining for CD31 or CD34, as well as absence of marked nuclear atypia, mitotic activity, and intracytoplasmic lumen formation. Other vascular lesions, such as epithelioid hemangiendothelioma, spindle cell hemangiendothelioma, and intravascular papillary endothelial hyperplasia, are excluded in a similar fashion.

ASSOCIATIONS AND GENETICS

There is no known predisposition in children born prematurely. In one study, one patient was described with both cutaneous meningioma and pheochromocytoma, although this may have been coincidental as it has not been reported since to our knowledge. Unusual presentations of cutaneous meningioma have been reported for a muscle markers smooth muscle actin and desmin can be used to rule out vascular, histiocytic, or myogenic tumors, respectively. In some cases, the cuboidal epithelioid cells of the ectopic meningothelial hamartoma may be intimately associated with vessels and have prominent, freely anastomosing vascular channels. In these cases the diagnosis of well-differentiated angiosarcoma is ruled out by expression of vimentin and EMA and lack of staining for CD31 or CD34, as well as absence of marked nuclear atypia, mitotic activity, and intracytoplasmic lumen formation. Other vascular lesions, such as epithelioid hemangiendothelioma, spindle cell hemangiendothelioma, and intravascular papillary endothelial hyperplasia, are excluded in a similar fashion.
set of siblings with occipital tumors in nearly identical locations, for a case associated with sinus pericranii (a congenital vascular anomaly), and for a case involving a 3-year-old girl with an ovarian fibroma. In another case, a fetus demonstrated rapid third-trimester growth, and the authors concluded that this was most likely due to the influence on the tumor by endogenous steroid hormones, similar to other meningiomas. However, these authors did not test for the presence of receptors on the described lesion. In another report, a 33-year-old woman described a painful growth during weaning. Sometimes, a history of trauma is noted before the development of a scalp lesion, which has been attributed to the presence of displaced meningial tissue at the time of trauma.

It is commonly recognized that patients with neurofibromatosis may present with meningioma, and reports of cutaneous meningioma in the setting of neurofibromatosis means that clinical vigilance is warranted when encountering these lesions.

Ouazzani et al recommend ruling out breast cancer upon encounter of this lesion because of the association of this disease with noncutaneous meningioma. Again, this exclusion may be warranted in the appropriate clinical context.

Otherwise, data regarding specific genetic defects in cutaneous meningioma are lacking, and no cases have pointed out common specific genetic associations. However, clues to the possibility of a genetic component exist in some cases, as reported in a study by Tron et al that highlights a familial presentation of type I cutaneous meningiomas with autosomal dominant transmission. These lesions were found on the scalp of 11 of 24 family members, spanning 4 generations. One other report described occipital nodules consistent with cutaneous meningioma that had been present since birth in a pair of teenage Japanese siblings. Interestingly, another report describes the case of a young Japanese girl diagnosed with meningial hamartoma of the scalp who also had a brother with a similar lesion.

**PROGNOSIS, TREATMENT, AND FUTURE DIRECTIONS**

The mainstay of treatment is complete surgical excision, and prognosis is dependent upon lesion type. Type I lesions are associated with a very good prognosis and can be managed surgically to obtain clear margins. To our knowledge, recurrence has not been documented. On the other hand, the prognosis for type II/III lesions is worse. Type III lesions located in surgically troublesome locations may not be amenable to surgical intervention. In these cases, the prognosis associated with the tumor depends on the degree to which the tumor can be excised, its rate of growth, and its ability to compress or interfere with vital structures. Of particular importance is the need to rule out the presence of a more deeply seated neoplasm, as this lesion may also need to be treated. Radiologic exploration and preoperative precautions should be taken into consideration before surgical therapy. Doing so will minimize unnecessary error and complication.

Targeted therapies are being investigated for the treatment of meningiomas that are not amenable to surgery (particularly type III lesions). These include vascular endothelial growth factor and platelet-derived growth factor inhibitors. These modalities remain investigational.

**References**