Cutaneous Meningioma: A Cytomorphological Diagnosis

Abstract

Cutaneous meningiomas are rare tumors. These are ectopic meningothelial cells located in the dermis and subcutis, and are usually seen on the scalp. Here, we report the case of a 40-year-old woman who presented with a slowly growing asymptomatic mass over the scalp in the right parieto-occipital region. The lesion was firm, adherent to underlying structures, and covered by normal appearing skin. The cytomorphological features along with histological and immunohistochemical studies showing positivity for epithelial membrane antigen, Vimentin, and S100 helped in making a definitive diagnosis of cutaneous meningioma. No evidence of intracranial meningioma was noted in contrast-enhanced computed tomography of the brain.

Keywords: *Adnexal tumors, cutaneous lesion, cytopathology, immunohistochemistry, meningioma*

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Introduction

Meningiomas are the most common neural tumors of adults constituting 15–20% of them, and commonly arise in the intracranial and intraspinal regions. They are derived from arachnoid cells of meninges. Rarely, they can present extracranially,[1] where they are accessible to fine needle aspiration cytology (FNAC) and pose a diagnostic dilemma. Here, we report a case of cutaneous meningioma of parieto-occipital region to emphasize the importance of keeping it as one of the differential diagnosis in the pathological evaluation of scalp swelling.

Case Report

A 40-year-old woman presented with a slowly growing asymptomatic mass over the scalp in the right parieto-occipital region. Clinical history revealed that there was no trauma or fracture in the past. There was no history of headache or convulsion. The patient did not have weight loss or loss of appetite. Her menstrual cycles were normal. The mass had been progressively increasing in size over the last 4 years. On examination, the swelling measured approximately 2 cm × 2 cm, was firm, slightly fixed, nontender, and covered by normal skin [Figure 1a]. It was non-fluctuant. There was no discharge or punctum associated with the swelling. There was no evidence of cervical lymphadenopathy on palpation. Further systemic and bilateral breasts examination revealed no abnormality in any of the systems.

X-ray of the skull showed bony calvaria with no evidence of any bony abnormality or fracture. Laboratory investigations revealed normal hematological parameters such as complete blood counts and peripheral smear examination. Biochemical tests such as thyroid, kidney, and liver function tests, as well as lipid profile of the patient were normal.

FNAC was performed and blood mixed cellular aspirate was obtained. Papanicolaou and Giemsa stains were applied. Smears prepared showed high cellularity comprising of clusters, sheets, whorls, and singly scattered epithelial cells. These cells showed mild pleomorphism and were round to oval with eccentric nuclei, fine granular chromatin, and moderate amount of cytoplasm [Figure 2a]. Few of the cells showed intranuclear inclusions [Figure 2a]. Many tight whorls of such cells were also noted [Figure 2a, inset]. No necrosis or any mitotic figures were seen, and the background showed blood. Finally, an impression of cutaneous meningioma was made based on the cytomorphological features. The patient

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was advised radiological investigations and neurosurgical intervention.

CECT brain showed evidence of soft tissue enhancing swelling over the right parieto-occipital region [Figure 1b]. Bony calvaria showed underlying erosion in the corresponding region, which was not detectable on X-ray of the skull. Underlying brain parenchyma, bilateral gangliocapsular region, posterior fossa, and falx midline all appeared normal. Based on the radiological findings, surgical excision of the mass was planned and pre-anesthetic checkup was advised. The tumor was surgically excised and sent for histopathological examination. The mass was completely excised with the intact capsule, and there was no intraoperative bleeding or any other complication.

Grossly, a single circular fibrofatty firm soft tissue mass measuring 3.5 × 3 × 0.3 cm was received. Multiple sections showed fibroadipose and fibrocollagenous tissue with interspersed whorls, nests, and syncytial sheets of meningothelial cells [Figure 2b and 3a]. An occasional focus of microcalcification was also noted [Figure 2b, inset]. Immunohistochemical examination revealed positivity for vimentin, epithelial membrane antigen (EMA), and S-100 [Figure 3b-d].

Based on the radiological, cytomorphological, and histological features in conjunction with immunohistochemical examination, a diagnosis of cutaneous meningioma (cutaneous meningothelial heterotopia) was made.

Discussion

Ectopic meningiomas are rare tumors derived from meningothelial cells located in the dermis or the subcutaneous tissue. These are most commonly found in the scalp and occur in both congenital and acquired forms.[2] They may be (a) an extension through foramina or defect, (b) may arise from embryonic arachnoid cells/multipotent mesenchymal cells anywhere in the body, and (c) may represent metastasis from primary intracranial/supraspinal meningioma. Extracranial and extraspinal meningiomas are rare and should be included as a differential diagnosis while examining FNAC from tumor, especially from the head and neck regions.

Clinically, these lesions present as firm, subcutaneous swellings, usually non-tender. These are of three types, as described below.[3] Type I tumors are congenital, whereas the other two are acquired later in life. Acquired lesions are cytologically similar to those of the congenital type, except for being more lobulated and cellular, and having less collagen and deeper extension into the dermis.

Intracranial meningiomas are common whereas extracranial ones are rare. The extracranial location of meningioma can occur by direct extension of an intracranial meningioma through bone foramina after any trauma or defect, or it can occur as cutaneous metastasis without any underlying bony

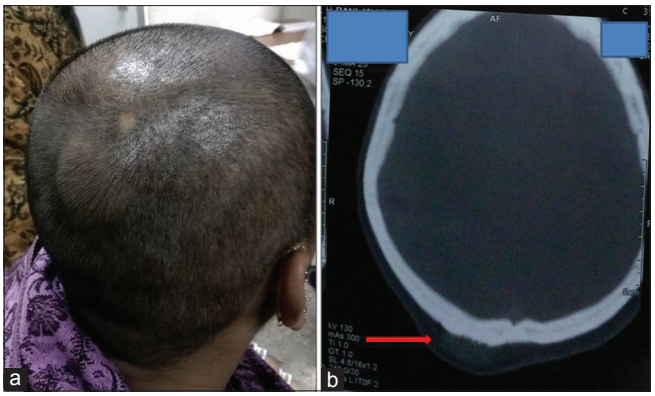

Figure 1: (a) A slowly growing asymptomatic mass over scalp on the right parieto-occipital region. (b) Contrast-enhanced computed tomography of the head showing bony erosion in adjacent area and no such intracranial lesion

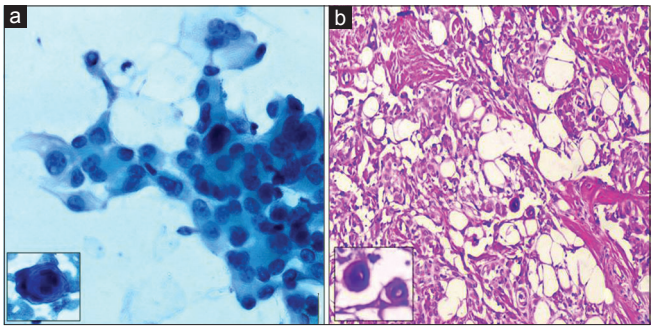

Figure 2: (a) Cells showing mild pleomorphism and round to oval eccentric nuclei, fine granular chromatin, and moderate amount of cytoplasm; few cells also show intranuclear inclusions (Pap, ×200); (Inset shows a whorl of meningothelial cells). (b) Sections showing fibroadipose and fibrocollagenous tissue with interspersed whorls, nests, and syncytial sheets of meningothelial cells (H and E, ×100); (Inset shows foci of calcification)

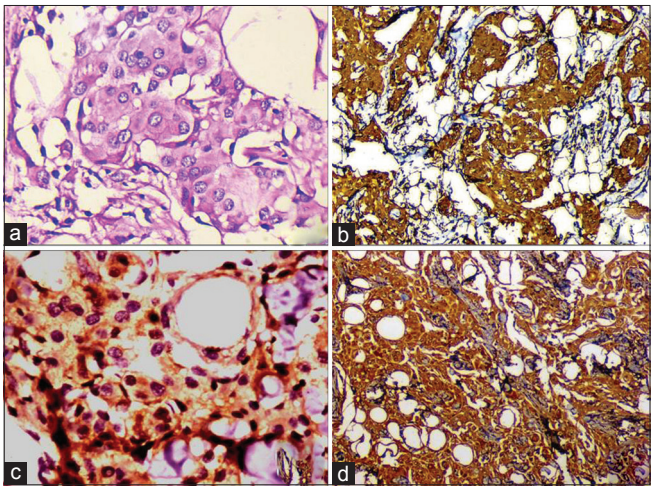

Figure 3: (a) Photomicrograph showing monomorphic cells mild pleomorphism with round to oval eccentric nuclei, fine granular chromatin, and moderate amount of cytoplasm (H and E, ×200). Meningothelial cells strongly immunoreactive for vimentin (b), EMA (c), and S100 (d)

defect or it can also be a primary ectopic meningioma, occurring de novo.[4,5]

Lopetz *et al*., in 1974, developed a widely used classification based on clinicopathological criteria. They divided cutaneous meningiomas into three types as follows.[3]

1. Congenital type, at birth, occur on the scalp and paravertebral region, developing from ectopic arachnoid trapped in the dermis and subcutaneous tissue. These may represent rudimentary meningoceles that lost connection to the central nervous system. Type I cutaneous meningioma is usually confined to the subcutaneous tissue
2. Ectopic soft tissue meningioma that extends to the skin by contiguity, occurs around the eyes, ears, nose, and mouth. No associated meningiomas of neuraxis are noted
3. Tumor extending into the dermis and subcutaneous tissue from meningioma that involves neuroaxis are much more common in adults. They may arise through bony defect, arising because of trauma or any other surgical reason.

Waghmare *et al*. reported a similar case of cutaneous meningioma; the patient had a bony defect which led to the migration of meningothelial tissue upto the scalp, resulting in extracranial meningioma.[4]

Important cytomorphological features in favor of meningioma are tightly cohesive clusters of spindle cells, whorls, lobular syncytial cellular fragments, intranuclear inclusions, nuclear grooves, and psammomatous calcification.

On the basis of cytological and histomorphological findings, most epithelial tumors can be easily distinguished by the presence of keratinization and sebaceous or sweat gland differentiation. Because meningiomas are rare tumors associated with vague clinical features, they may have to be differentiated from lesions such as appendageal cell tumors, epithelioid/spindle cell sarcoma, peripheral nerve sheath tumors, squamous cell carcinoma, hemangioma, giant cell fibroblastoma, hemagiopericytoma, or other types of heterotopic neuroglial lesions.[2-5] FNAC is a simple and effective procedure and can be highly valuable in diagnosis of cutaneous meningioma, as done in the present case.

Results are often inconclusive when based solely on clinic-radiological findings, hence, cytology and histology are crucial for a definitive diagnosis. Mostly noted are sheets, lobules, and nests of oval to polygonal meningothelial cells, with whorled arrangement along with collagen and psamomma bodies, which help in clinching the diagnosis. Immunohistochemically, these lesions are immunoreactive for EMA and Vimentin and immunonegative for cytokeratins, CD34 and CD31, CD68, Desmin, SMA.[5] These markers help to exclude epithelial, melanocytic, vascular, or myogenic differentiation.

However, sometimes, immunohistochemistry may overlap giving inconclusive results, in such cases, ultrastructural findings help to confirm the diagnosis. Ultrastructurally, these tumors resemble that of intracranial meningiomas.[6,7] Nochomovitz *et al*. described these lesions as interdigitating processes, desmosomes, and hemidesmosomes, resulting in a jigsaw pattern, whereas Miyamoto *et al*. in their respective study demonstrated the presence of swollen, stellate meningothelial cells with a moderate number of microfilaments, straight, and interdigitating cytomembranes, and desmosomes-like junctions.[4-7]

Genetic predisposition has not yet been established for these lesions; however, Tron *et al*. and Miyamoto *et al*. highlighted familial presentation of type I cutaneous meningiomas with autosomal dominant mode of inheritance in their studies.[7,8]

Definitive treatment is complete surgical excision. However, if the lesion is small and asymptomatic, simple surgical excision can be considered.[2,5] In our case, the lesion was completely excised and sent for histopathological confirmation. Some studies found that cutaneous meningioma Type I has better prognosis. Type III lesions can be difficult to operate leading to poorer prognosis. Presence of any deep-seated neoplasm should be properly evaluated because it may require different mode of intervention. Role of vascular endothelial growth factor inhibitors and platelet derived growth factor inhibitor is being intensively studied in the management of these lesions.[9-11] However, the case presented here does not fit accurately into any of the specific types mentioned in the classification. This may be considered as type II lesion arising because of head injury resulting in the extension of the meningeal tissues through the bony defect mentioned in the CECT report of the patient.

Conclusion

The present study emphasizes the importance of cytology in conjunction with histopathology in making a diagnosis of cutaneous meningioma and to keep it as one of the important differential diagnosis while evaluating soft tissue mass in the head and neck region. Radiological studies are important to detect any other association such as an underlying bony defect, any connection to the neuraxis, or presence of an intracranial meningioma, which help the surgeons in planning a proper intervention.

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Conflicts of interest

There are no conflicts of interest.

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