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Case Report

Primary Type I Cutaneous Meningioma of the Scalp: Cytohistological and Immunohistochemical Features of a Rare Neoplasm

Abstract

Primary cutaneous meningioma of scalp is a rare lesion and often clinically misdiagnosed. As clinical features are nonspecific, the diagnosis is often perplexing in this exceptional site. We report a case of a 7-year-old girl presenting with a nodule over occipital region of scalp since birth. No bony defect was noted on X-ray. On fine needle aspiration cytology, smears were cellular consisting mostly cohesive clusters of cells having pale pink granular cytoplasm, round to oval nuclei with unassuming nucleoli along with psammomatous calcification suggestive of cutaneous meningioma. Histology and immunohistochemistry with epithelial membrane antigen (EMA) confirmed the diagnosis. Histological sections revealed a proliferation of a spindle-shaped cell in the dermis, arranged in whorls or individually amid collagen fibers and psammoma bodies. Immunohistochemical study revealed positivity for EMA. A diagnosis of Type I cutaneous meningioma was finally rendered based on characteristic clinical, intraoperative, morphological, and immunohistochemical observations.

Keywords: *Fine needle aspiration cytology, histopathology, immunohistochemistry, primary cutaneous meningioma, scalp*

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Introduction

Meningiomas are a common neoplasm of the central nervous system where the tumor is derived from the arachnoidal cells. Such neoplasm accounts for 15% of intracranial and 25% of intraspinal tumors.[1] Meningiomas are rarely extracranial (<2% of all meningioma) and encountered in the head neck region, skin, lung, and mediastinum.[2,3] Cutaneous meningiomas are extremely uncommon. Although such lesion is generally present from birth, they are usually diagnosed in childhood or later.[4] There is no specific clinical feature and symptoms are related to the involved anatomic site, however, alopecia and hair collar sign may be present. Consequently, the diagnosis is puzzling in these exceptional locations and includes pathological differential diagnosis such as paraganglioma, carcinoma, melanoma, schwannoma, and olfactory neuroblastoma.[5] Clinical, cytological, histological and immunohistological findings of one such rare case is presently being depicted.

Case Report

A 7-year-old girl presented with a firm, painless swelling with localized alopecia on the occipital region of the scalp since birth. Skull X-ray and all the laboratory investigations were normal [Figure 1]. Due to financial constraints magnetic resonance imaging (MRI) examination could not be done. Fine needle aspiration cytology of the scalp swelling was done using a 24-gauge needle and 10-ml syringe to obtain scanty fluid aspirate. The smears were stained with Papanicolaou and Leishman-Giemsa stains. Cytologically, smears were highly cellular, consisting predominantly of cohesive clusters, some seen as small tight whorls and scattered single cells [Figure 2]. These cells had pale pink granular cytoplasm, indistinct cytoplasmic borders, round to oval nuclei and inconspicuous nucleoli. Psammomatous calcification and cells arranged around eosinophilic globules were noted. The mass was excised under general anesthesia. It had well-defined margins and was present subgaleally without involving periosteum. Gross examination revealed an ovoid brownish piece of tissue measuring 1 cm × 1 cm × 0.8 cm. Cut surface was

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grayish white and solid. On histopathological examination, sections show dermal proliferation of spindle-shaped cell, some arranged in whorls and other individually among collagen fibers. Psammoma bodies were noted [Figure 3]. A histological diagnosis of cutaneous meningioma was made. Immunohistochemical analysis of the tissue was done and was found positive for epithelial membrane antigen (EMA) [Figure 4]. Therefore, the final diagnosis is Type I cutaneous meningioma, based on clinical, cytological, histopathological, and immunohistological features.

Discussion

Primary cutaneous meningioma is an exceptionally rare but well-distinguished entity. The first reported case of such neoplasm was in 1904,[6] though its first description was found in an article in the year 1956.[7] The rarity of these tumors may be appreciated by the fact that most of the documentation regarding such lesion is in the form of case reports. The classification system was introduced in the year 1974 by Lopez *et al*., in a large report.[4]

Lopez *et al*.[4] divided cutaneous meningioma into three types based on different etiologies. Type I is the congenital type that is generally present at birth and occur most often on the scalp and paravertebral regions. These tumors arise from ectopic arachnoid cells that become trapped in the dermis and subcutis during development. Considering the clinical and histopathological features, the present case may be classified in this category. Type II includes ectopic soft tissue meningiomas extending to the skin by contiguity and occurs around the eyes, ears, nose, and mouth.[4] They occur later in life. There is no corresponding meningioma of the neuroaxis. These tumors arise from remnants of arachnoid cells, which extend along cranial nerves. Type III lesions are primary meningeal tumors that secondarily involve the skin by direct extension of these tumors and are more common in adults.[4]

Cytologic features for the primary cutaneous meningioma include bland nuclei, cellular whorls, and psammoma bodies which are similar to findings characteristic of central nervous system meningioma.[8,9] In another cytological report of a Type III cutaneous meningioma, smears revealed spindled cells in clusters and a concentric arrangement,

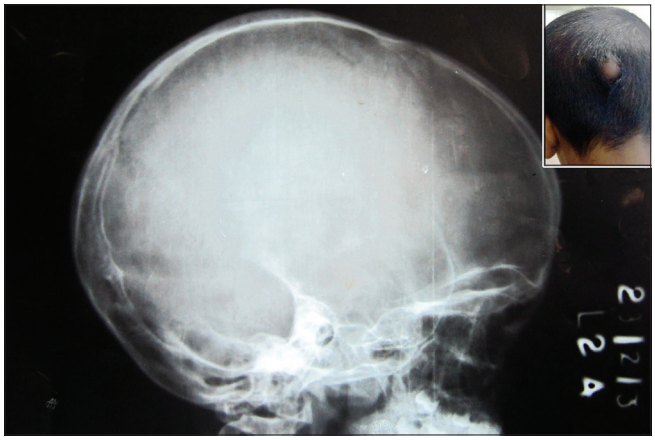

Figure 1: X-ray skull was normal showing no bony lesion or lytic lesion. Inset (upper left) demonstrates the occipital swelling of the case

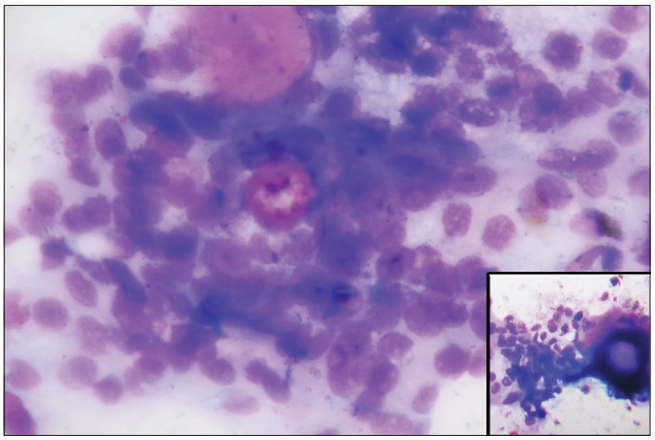

Figure 2: Cytological smear showing cohesive clusters and scattered single meningothelial cells having indistinct cytoplasmic borders, round to oval nuclei and inconspicuous nucleoli. Inset (lower left) exhibits a psammomatous calcification. (Leishman-Giemsa, ×400)

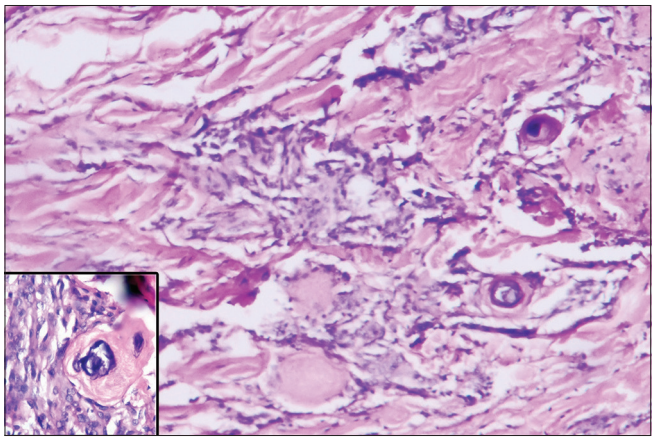

Figure 3: Section showing dermal proliferation of spindle-shaped meningothelial cell in whorls and psammoma bodies (H and E, ×100)

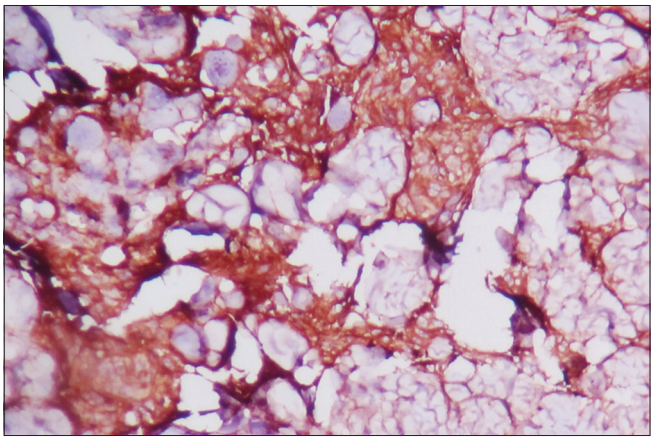

Figure 4: Section showing immunostaining positivity for epithelial membrane antigen in meningothelial cells (×100)

generating characteristic whorls. Cells showed pale nuclei with finely granular, evenly distributed chromatin, and occasional intranuclear cytoplasmic invaginations.[10]

Clinically, cutaneous meningiomas resemble lipoma, cysts, skin tag, nevi, vascular lesions, and fibroma. Histologically, this tumor must be differentiated from squamous cell carcinoma, hemangioma, giant cell fibroblastoma, hemangiopericytoma, or other types of heterotopic neuroglial lesions.[11] Vigilant microscopic examination with appropriate immunohistochemical stains is helpful to clinch the diagnosis. Cutaneous meningioma express vimentin, EMA, and S-100 protein, just like intracranial meningioma. EMA and vimentin positivity are considered supportive evidence for the diagnosis of meningioma[12] while cytokeratin, CD34, and desmin are to be used for ruling out histological differentials.

Neurofibromatosis Type I and malformations of fingers and toes may be associated with cutaneous meningioma[13] warranting clinical vigilance.

A close histopathological differential of Type I cutaneous meningioma is atretic meningocele. It has the presence of rudimentary cystic cavity or stalk with strands and nest of meningiothelial cells. They are usually present in the midline in parietal and the occipital region as palpable skull defect along with swelling. X-ray skull generally shows midline bone defect and computed tomography scan helps to confirm the diagnosis. MRI brain is helpful in diagnosing associated anomalies and is recommended for the symptomatic patient. Intraoperative distinguishing features are generally the presence of calvarial defect and fibrous strands extending intracranially.[14]

Though primary cutaneous meningioma is rare, the tumor has a better prognosis than secondary tumors arising from metastasis from a primary intracranial tumor. The preferred management is complete surgical excision.[11] Prognosis is related to the type of lesion. Type I lesions have a very good prognosis without any recurrence when a clear surgical margins can be obtained. However, prognosis for Type II/III lesions is poor.[4] Type III lesions may not be surgically resectable if situated in a troublesome locations. Prognosis in such situation depends on the extent the tumor can be excised, its rate of growth, and its magnitude to compress or interfere with vital structures. Presence of a more deeply seated tumor should be ruled out by radiologic investigations.[11]

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Conflicts of interest

There are no conflicts of interest.

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