

CUTANEOUS MENINGIOMA: CLINICOPATHOLOGIC AND IMMUNOHISTO-CHEMICAL FEATURES OF TWO CASES WITH A BRIEF REVIEW OF THE LITERATURE

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INTRODUCTION

Meningiomas are fairly common neoplasms of the central nervous system in adults, presenting in intracranial or spinal sites. They are rare in childhood. These tumors are thought to arise from specialized cells of the pia-arachnoid matter. Meningiomas are rarely encountered outside the central nervous system. Extracranial and extraspinal meningiomas can be divided into four categories: (1) direct extension of a CNS meningioma outside the skull, often through bone or bony foramina; (2) metastatic meningiomas; (3) extracranial meningiomas arising from arachnoid cells in cranioperipheral nerve sheaths and (4) primary ectopic meningioma. Each of these categories of ectopic meningiomas can involve the skin. Similar tumors have been described in the soft tissues of the orbit, the conjunctiva, the caruncle, ear, head and neck and much more uncommonly at sites more remote from the CNS.

Cutaneous meningiomas are extremely uncommon. The clinical, histological and immunohistochemical features of two cases of primary cutaneous meningiomas are reported in this article.

PATIENTS & METHODS

CASE REPORTS:

Case 1.

A 24-year-old man was admitted to Hamad General Hospital on the 6th of July 1985 with the chief complaint of a lump on his back since childhood, that he wanted removed before enrolling in the army. Physical examination revealed a subcutaneous mass,

3 cm in diameter, in the upper posterior thoracic area at the level of T4. It was firm and painless on palpation and adherent to the deep planes but not to the overlying skin, which appeared normal. CT-scan of the thoracic spine showed dysraphism of upper dorsal and lower cervical vertebrae with a small meningocele at the D4. All other laboratory and radiologic investigations carried out pre-operatively were normal. A firm whitish mass was excised under general anesthesia. It had no obvious connection to the underlying bone nor to the surrounding soft tissue. The patient recovered uneventfully and was discharged on the 10th of July 1985.

Case 2.

A 2-year-old girl presented to the Day Care Unit of Hamad General Hospital (on 20th July 1990) because of a painless, mobile, non-compressible mass palpated on the parietal region of the scalp. Skull X-ray and all the laboratory investigations performed pre-operatively were normal. The mass was excised under general anesthesia. It had no connection with intracranial structures.

METHODS:

Formalin-fixed paraffin-embedded four micron sections were stained with hematoxylin and eosin, Gordon and Sweet stain for reticulin, Periodic Acid Schiff with and without diastase digestion, and Von Kossa for calcium. Immunohistochemical stainings were performed by peroxidase anti-peroxidase technique (PAP) for epithelial membrane antigen (EMA), cytokeratin, vimentin, desmin, neurone specific enolase (NSE), carcinoembryonic antigen (CEA) and glial fibrillary acidic protein (GFAP). All antibodies were purchased from Dako Patt Denmark.

RESULTS:

Pathological Findings:

The specimen in case 1 consisted of an ovoid mass measuring 3.0 x 2.0 x 1.8 cm with a ragged outer surface. Cut section was grey, firm with a yellow central soft area. Sections showed a poorly circumscribed unencapsulated lesion involving dermis and subcutaneous fibroadipose tissue. The lesion consisted of a collagenous stroma with multiple

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Fig 1:
Primary
cutaneous
meningioma
(case 1),
Note
psammoma
bodies in the
centre. H&E

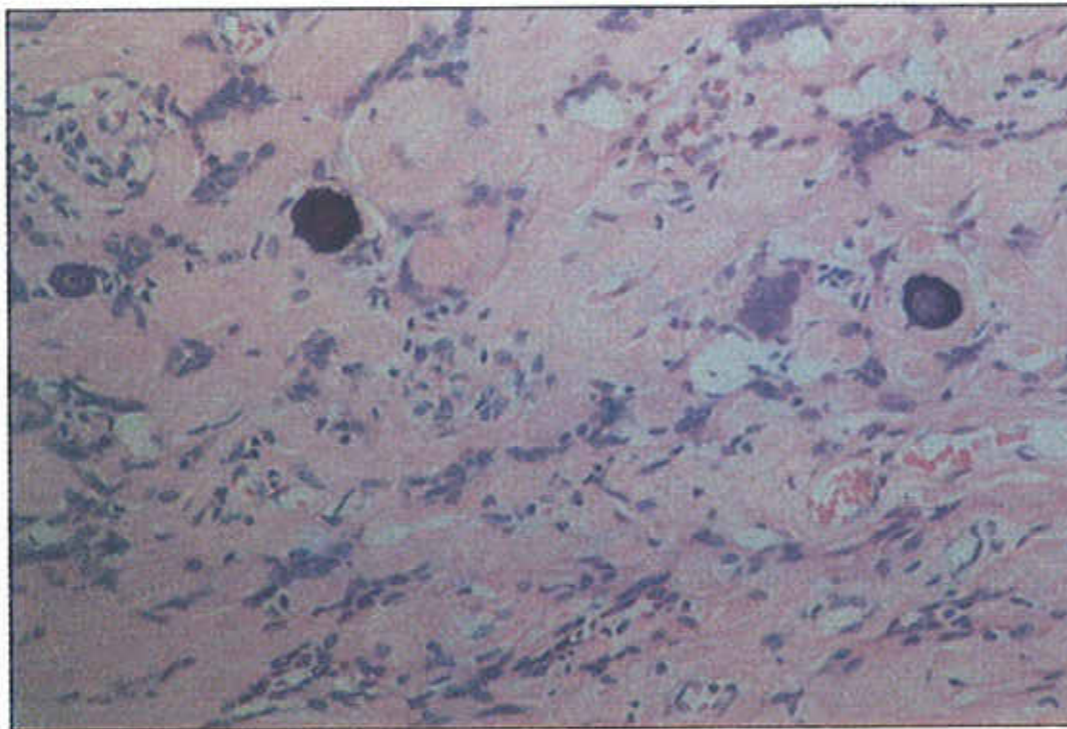


Fig 2:
Primary
cutaneous
meningioma
(case 1),
Foci of
meningothelial
proliferations
in a
collagenous
background

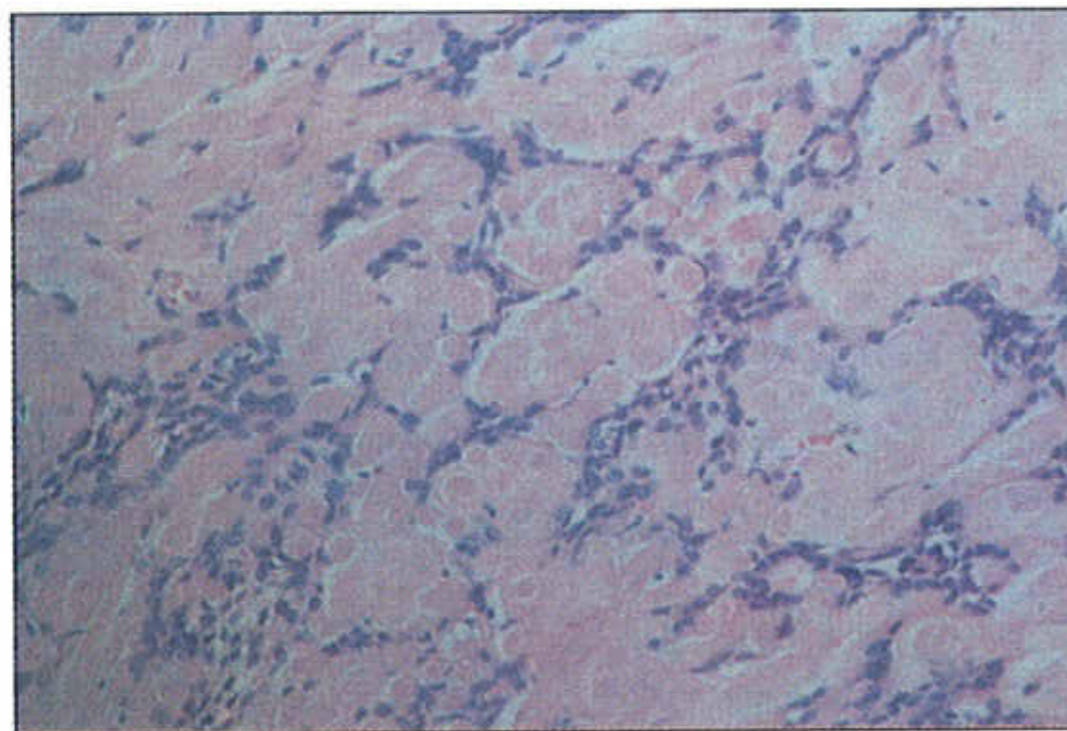


Fig 3:
Primary
cutaneous
meningioma
(case 2), An
ovoid lesion in
the deep
dermis with
multiple
lymphatic
spaces in the
centre. H&E.

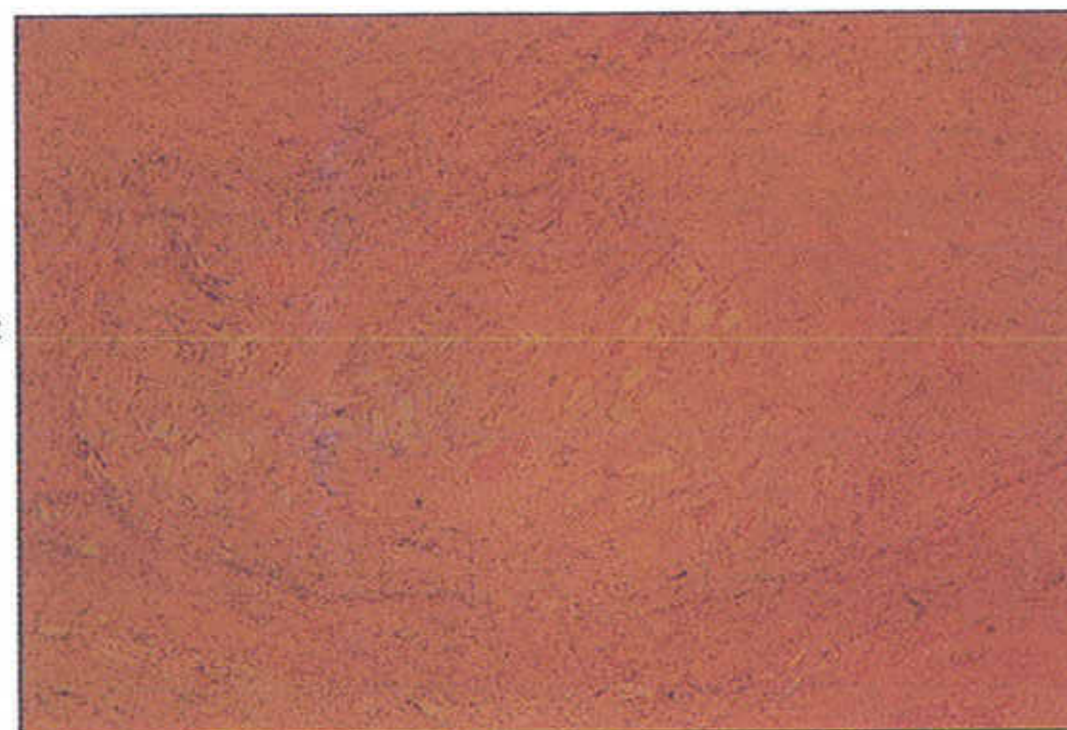
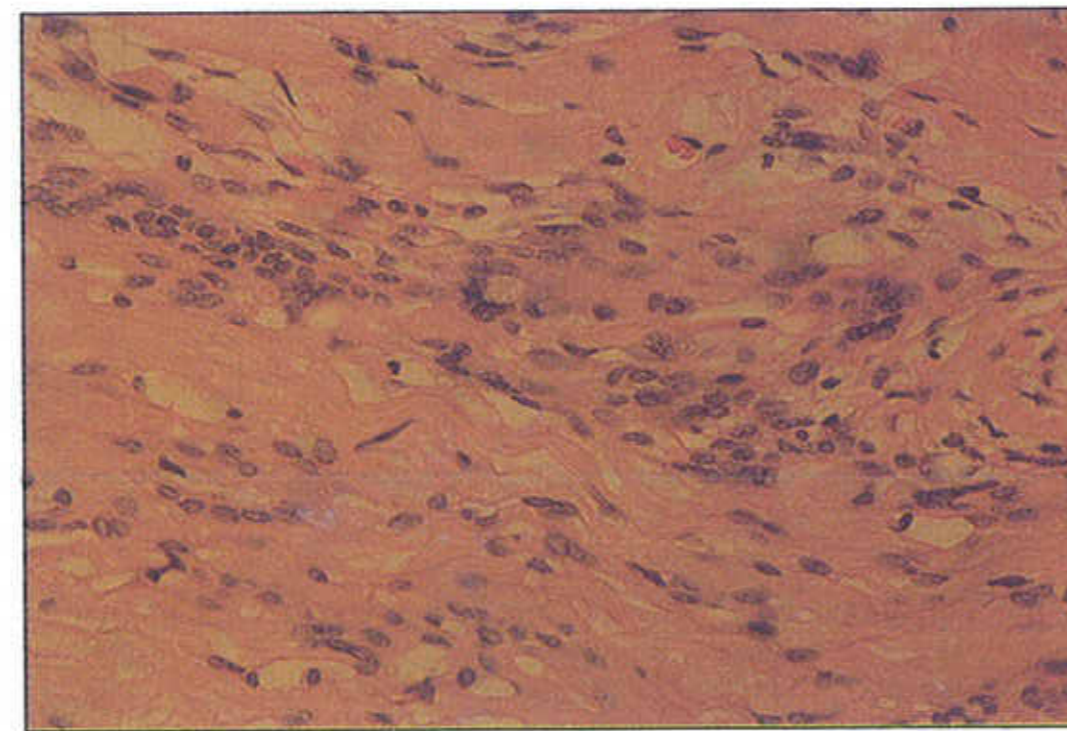


Fig 4:
Primary
cutaneous
meningioma
(case 2),
Zones of
meningothelial
proliferations.
H & E



lamellated, calcified psammoma bodies (Fig 1) and nests of predominantly round meningothelial proliferations (Fig 2). Multiple small, often slit-like, spaces were present, some containing groups of meningothelial cells. Multiple rounded collagenous bodies were noticed in the stroma.

The specimen in case 2 consisted of an ellipse of hair-bearing skin 1.5 x 0.8 x 0.8 cm. Microscopically the lesion had involved all dermal layers and extended to the subcutis. However, the main body of the lesion had occupied deep dermal layers. This was characterized by a poorly circumscribed unencapsulated ovoid nodule with a core consisting mainly of collagenous stroma and multiple lymphatic spaces (Fig 3). The lesion was more cellular in the periphery with nests of round, oval and spindle cells, arranged in whorled patterns in multiple foci (Fig 4). Many of these whorled patterns had been produced by meningothelial cells wrapped around dermal structures such as nerves, vessels and arrector pili muscles. Round and spindle cells were also seen closely associated with skin adnexa especially hair follicles and eccrine sweat gland ducts. A few psammoma bodies were present in the periphery of the lesion (Fig 5). Small rounded collagenous bodies were frequent (Fig 6), some surrounded by whorls of meningothelial cells.

Immunohistochemical Findings:

Spindle cells and polygonal cells in both cases were strongly positive for vimentin (Fig 7) and EMA (Fig 8). Focal staining was present for cytokeratin and S100 protein. Stainings for GFAP, NSE, desmin and CEA were all negative in the tumor cells.

DISCUSSION:

Primary (ectopic) cutaneous meningioma is a rare but well documented entity. These lesions are generally present from birth, but may be detected during childhood and early adulthood⁽¹⁾. They occur most often on the scalp, forehead or paravertebral areas^(2,3,4), and may mark a further underlying abnormality of closure of the neural tube. This class of cutaneous meningioma has an excellent prognosis in contrast to direct extension, or metastasis into the skin, of an intracranial meningioma which occurs in adults and carries a

poor prognosis. The situation is somewhat similar to the equally rare but more widely recognized occurrence of glial tissue well documented in the nose and in a variety of sites, predominantly in the head and face region^(5,6). It is worth noting that the combination of heterotopic glial and meningeal elements has been described recently in a single case report of a subcutaneous nodule in the buttock of a 1-day-old female neonate⁽⁷⁾.

Cutaneous meningiomas have some similarities and overlap with meningoceles. Sibley and Cooper (1988) have concluded that lesions referred to as primary cutaneous meningioma fall into two categories: ⁽¹⁾ those that microscopically resemble conventional meningioma of the central nervous system, and ⁽²⁾ those that have microscopic features consistent with a developmental anomaly or hamartoma⁽⁸⁾. Morrogi et al (1991) suggest that rudimentary meningocele of the skin and classic meningocele represent closely related developmental malformations⁽⁹⁾.

Immunohistochemical studies and electron microscopic observations have highlighted extensive immunophenotypic and ultrastructural similarities between cutaneous meningiomas, intracranial meningiomas and arachnoid cells^(3,4,10). Intracranial meningiomas stain positively for vimentin, cytokeratin, S-100 protein, EMA, and desmoplakin, and ultrastructurally, interdigitating cell processes and true desmosomes are hallmarks in their diagnosis. In cutaneous meningioma as with intracranial meningioma, there is widespread expression of both vimentin and EMA as well as cytokeratins and S100 protein.

Electron microscopic studies also have confirmed ultrastructural similarities to intracranial meningiomas such as long, interdigitating processes, pseudo-mesaxons containing collagen fibrils and foci of external lamina, cytoplasmic intermediate filaments and complex, tangled cell processes joined by punctate desmosomes focally⁽¹⁰⁾.

An interesting concept to consider is the relationship between perineurioma and cutaneous meningioma and their principal cell of origin i.e. perineurial cells and pia arachnoid cells.

Fig 5:
Primary cutaneous meningioma (case 2), Note a psammoma body in the periphery of the lesion. H & E

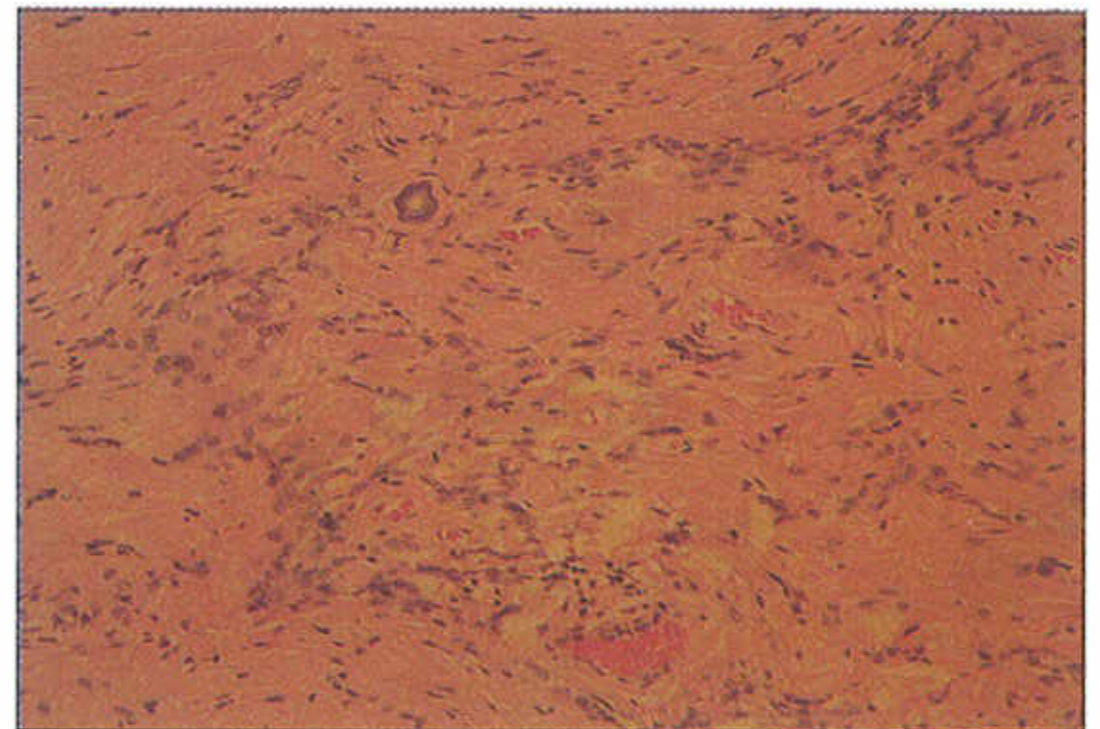


Fig 6:
Primary cutaneous meningioma (case 2), Note multiple hyaline collagenous bodies. H&E

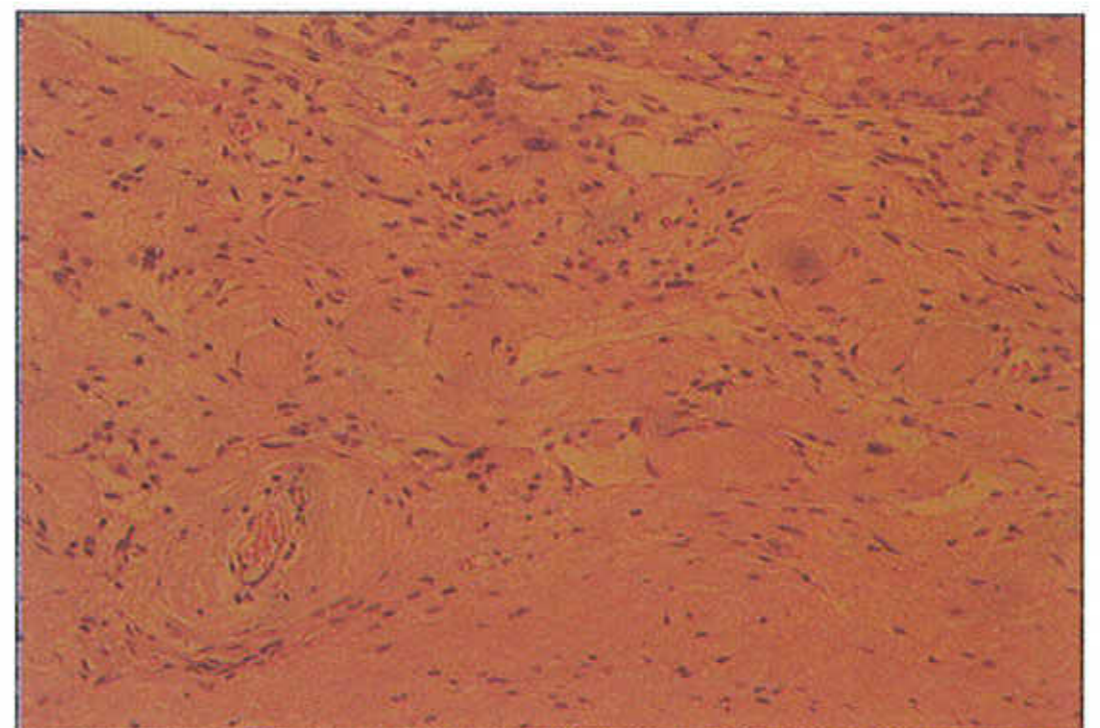


Fig 7:
Primary cutaneous meningioma (case 2), Tumor cells show positive immunostaining for Vimentin (PAP)

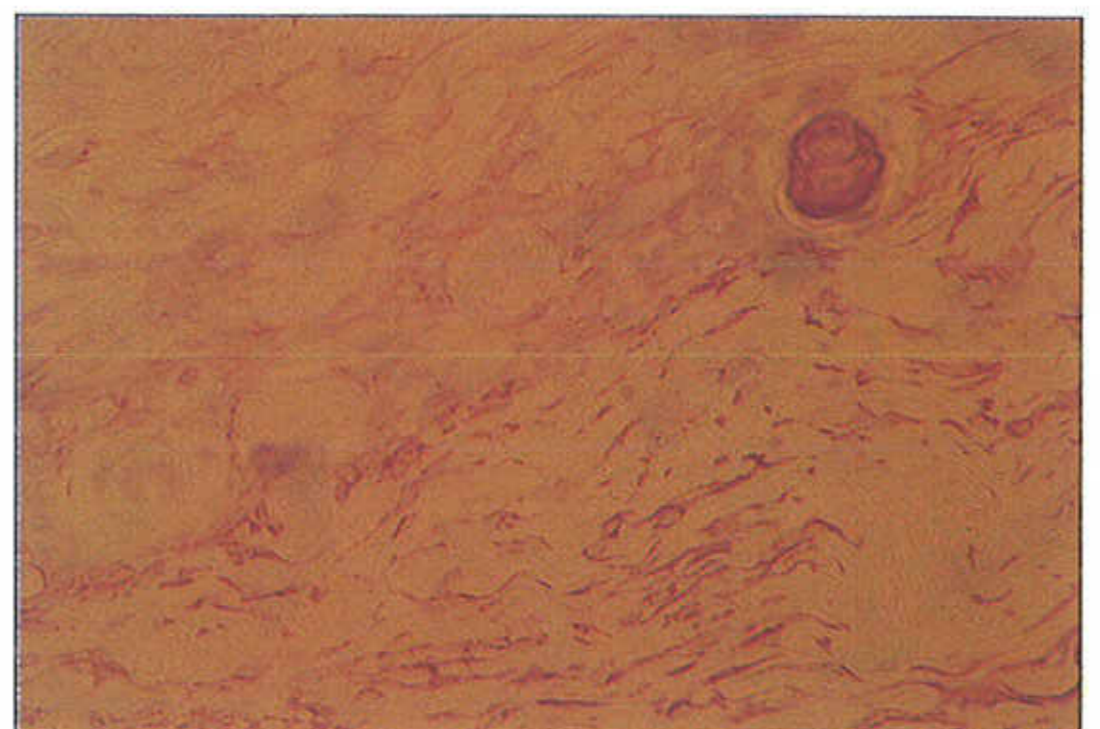
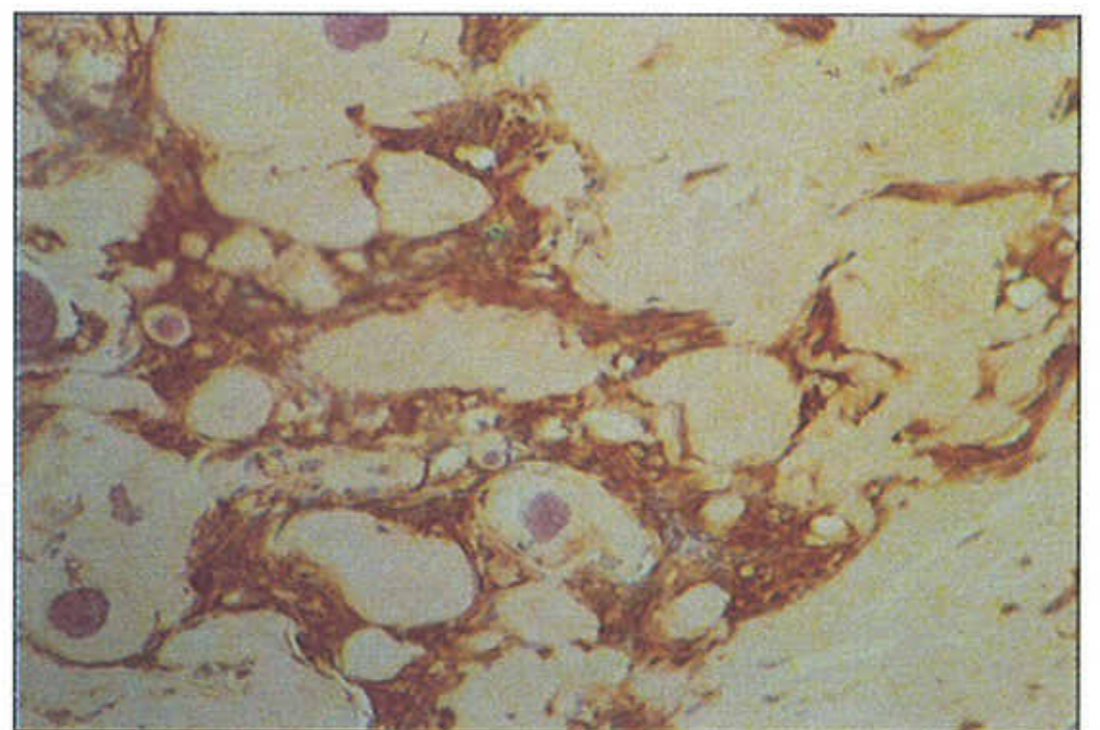


Fig 8:
Primary cutaneous meningioma (case 1), Tumor cells demonstrate strong staining for epithelial membrane antigen (PAP).



Perineurioma (storiform perineurial fibroma) first described in 1978 by Lazarus and Trombetta⁽¹¹⁾ is an uncommon but distinctive soft tissue tumour arising in the dermis, subcutis, or deep soft tissue. It is a morphologically distinguishable neoplasm believed to be closely related to cutaneous meningiomas. Perineuriomas are also positive for EMA and vimentin, but are negative for S-100 protein, neurofilament, CD34, desmoplakin, desmin and smooth muscle actin⁽¹²⁾. Perineurial cells of peripheral nerves have been demonstrated to become continuous with the pia arachnoid membrane of the central nervous system at the intervertebral foramina⁽¹³⁾. Cellular morphology and enzyme histochemistry is identical for perineurial and pia arachnoid cells and both cell types are immunopositive for vimentin and EMA. Meningiomas demonstrate similar features to the arachnoidal cap cells. These findings suggest that cutaneous meningiomas probably originate from

arachnoidal cell nests misplanted into the skin during embryogenesis and occur around sensory organs, cranial and peripheral nerves.

Immunohistochemical staining of epithelial membrane antigen must be accepted as a most useful marker of meningeal cells. Immunophenotypic profiles of cutaneous meningiomas together with light microscopic features such as infiltrative border of the lesion, involvement of subcutaneous tissue and the epithelial appearance of the nests of the cells may lead the unwary into a falsely based misdiagnosis of carcinoma. Presence of psammoma bodies in a tumour, especially in the neck region may lead to a futile search for thyroid carcinomas. The single most important factor leading to their accurate identification is the realization that meningeal lesions do actually occur in the skin.

ABSTRACT

Cutaneous meningiomas are rare lesions of the scalp and back which are generally present at birth but may be detected during childhood or early adulthood. Two cases are described: in the paravertebral (D4) region of a 24-year old man and in the parietal region of a 2-year-old girl. Diagnostic histological features includes psammoma bodies, small collagenous bodies and varying degrees of meningiothelial proliferation. Cutaneous meningiomas

show extensive ultrastructural and immunophenotypic similarities to intracranial meningiomas including widespread expression of vimentin and epithelial membrane antigen. Cutaneous meningiomas probably originate from arachnoid cell nests misplaced into the skin during embryogenesis and occur around sensory organs, cranial and peripheral nerves.

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