Type 1 Primary Cutaneous Meningioma with Atretic Meningocele

To Editor,

Meningiomas by far are the most common intracranial neoplasm described. While meningiomas are primarily described as intracranial neoplasms, rarer variants have been reported with extracranial extension. It is thought to be less likely via erosion of the bone from the primary intracranial mass. Benign extracranial extension due to growth of arachnoid cell rests entrapped in the calvarium during embryogenesis is thought to be the primary mechanism. Cutaneous forms of meningiomas are also more commonly seen following trauma, and tend to grow more rapidly following surgical intervention or radiotherapy.[1,2] Clinical diagnosis is often difficult for these tumors- presenting as a subcutaneous nodule and misdiagnosed as sebaceous cysts or lipomas.

A 30-year-old male patient presented with complaints of a chronic swelling at the back of his head. Though not associated with any neurological symptoms, he had noticed the mass to be progressively increasing in size. There is no history of trauma to the head, and his family remember the swelling to be present since birth.

The lesion was partially resected at the primary care with no significant intra or post-op complications. The specimen was subsequently sent for histopathology- which showed paucicellular dense fibrous tissue with psammoma bodies. Suspecting a neoplasm, the patient was referred to us for further management.

Upon presentation to our clinic, neurological examination was unremarkable. A 4.5 × 2.5 cm spherical swelling was noted at the site of the original swelling, with overlying focal alopecia. No other skin changes were noted over the swelling. The specimen from the surgery was sent for review at the local Cancer Center and the patient was scheduled for imaging.

A 3T MRI of the brain with contrast was done, which showed widening of the outer table of the bone in the midline parietal area consistent with the area of the swelling, measuring 4.76 × 3 × 1.2 cm [Figure 1]. Areas of post-surgical changes were seen in the subcutaneous plane, and a discrete tract was visualized through the bone to the dura at the level of the superior sagittal sinus. No intracranial lesion was seen. The lesion along with the tract was contrast-enhancing with a short T1 signal.

We followed up with a CT with contrast with focus to the area of the swelling. This showed a 5 × 1 mm tract within the posterior-parietal segment of the superior sagittal suture with the caudal end of the tract extending to the dura of the superior sagittal sinus and continuous with it. This was clearly visualized on the virtual 3D reconstruction of the skull showing the calvarial defect [Figure 2].

Our suspicion was confirmed with the histopathology report, showing EMA, ER, and Vimentin positivity [Figure 3].

Meningiomas are the most common intracranial extra-axial neoplasms. They have a propensity to invade adjacent structures, which often causes recurrence following surgery. Cutaneous meningiomas have been classified into three subtypes by Lopez *et al*.[3]

Proposed mechanisms include:- (1) heterotopic rests of arachnoid tissue entrapped in between soft tissue layers during embryonal development and subsequent growth, (2) vascular turbulence (due to transient cerebral hypertension during development) leading to detachment and embolization of arachnoid islets, (3) blunt trauma to the head- causing displacement of meningeal tissue superficially and subsequent growth, (4) centrifugal migration of arachnoid cells to the skin along the cranial and spinal nerves, and (5) persistent atretic encephalocele with intracranial extension.[4-7]

Type-I meningiomas are primarily cutaneous- found near the scalp and paravertebral region, thought to arise by the "meningocoele mechanism." They are usually congenital, benign, and asymptomatic.

Type-II lesions are acquired, developing from abnormal migration of arachnoid cells along cranial and spinal nerves during embryogenesis. This is consistent with the sites they are commonly seen- along the sinonasal tract, orbit, and the oral cavity.

Type-III lesions are direct extensions of intracranial meningiomas to the dermal or subcutaneous plane. This may be via bone erosion or secondary defects created by trauma or surgery. They are usually seen on the face, temple, or scalp and are slow-growing.

Type-II and III lesions tend to be symptomatic and carry a worse prognosis. Extracranial meningiomas up to 45cm in size, weighing 3.5 kg after resection have been reported.[2]

Immunohistochemistry and imaging are the cornerstones to the diagnosis of a cutaneous meningioma, which must be recognized and differentiated from other scalp swellings. MRI remains the mainstay imaging modality to delineate the margins of the tumor.

Tissue diagnosis can be obtained via fine needle aspiration from the swelling and positivity for EMA, ER, and vimentin in a background of densely packed fibrous tissue confirms the diagnosis. Cytokeratin, actin, S-100, CD31, and CD 34 negativity excludes tumors of epithelial, melanocytic, vascular, and myogenic lineage. There an association of cutaneous meningiomas with breast cancer, neurofibromatosis, and pheochromocytoma. An autosomal dominant mode of inheritance

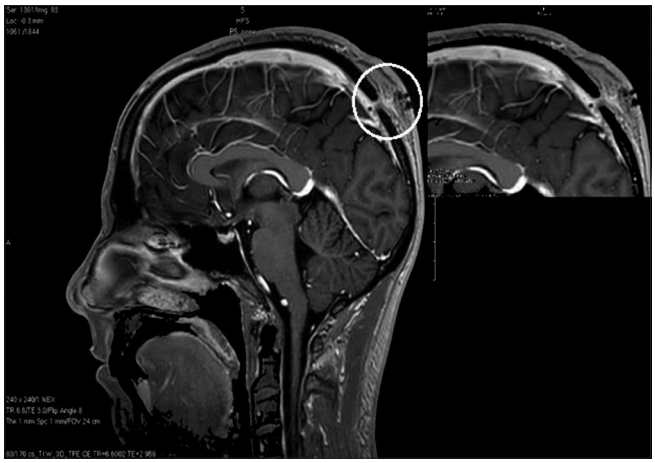

Figure 1: T1 Sagittal section with contrast. The film demonstrates the intracranial communication (of the resected mass) along with the calvarial defect, the tract enhancing with contrast

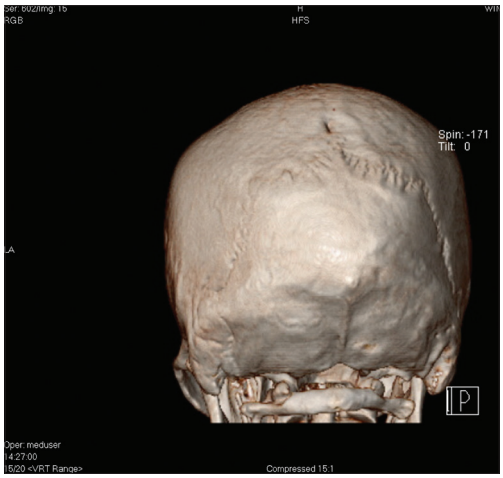

Figure 2: 3D reconstruction of the skull using CT images. The midline calvarial defect is seen in the posterior parietal region as a depression indicating the site of the communicating tract

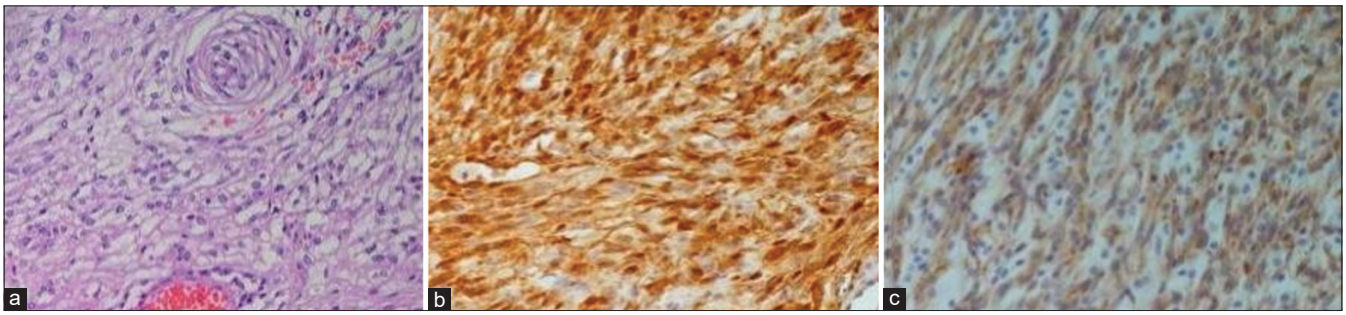

Figure 3: (a) Low power histopathological examination of tissue obtained from the lesion. Paucicellular tissue with psammoma bodies are often seen on histopathology. (b) Specimen stains positive for S-100. (c) IHC positivity for EMA

has also been proposed. They are thought to grow in the presence of high levels of endogenous steroids, including estrogen.[8-10]

Treatment includes surgical resection of the tumor. For type I lesions, the tract must be identified and removed with a rim of surrounding bone along with the pathologic meningeal tissue. Intracranial lesions should always be suspected especially if the meningioma is located near the orbit, nose, or the oral cavity. Type I lesions carry a very good prognosis and recurrence following complete resection has not been reported.[8]

Deep-seated type II and III lesions may not be surgically accessible for which radiation therapy remains the mainstay. VEGF and PDGF inhibitor-based therapies are still under study. Type II and III lesions are associated with a poorer prognosis. Meningiomas, even though primarily described as intracranial, extra-axial tumors can also be seen extracranially as cutaneous lesions. Cutaneous meningiomas are divided into three types based on location and pathogenesis. The reported case is of a type I cutaneous meningioma. This is confirmed by positive histopathology and evidence of persistent intracranial communication through a calvarial defect. Surgical resection is the gold standard of treatment, and the intracranial communication along with the surrounding bone should be identified and removed with the tumor to prevent recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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