Unusual case of temporal dermoid cyst presenting as oculomotor nerve palsy

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Dermoid cysts are choristomas resulting from the inclusion of ectodermal tissue during closure of the neural tube and occur along the epithelial lines of fusion. Frontotemporal dermoids are the most common type and generally present as an asymptomatic mass. We present an unusual case of frontotemporal dermoid presenting as sudden onset oculomotor nerve palsy in young male patient and describe the neurosurgical approach in its management.

Key words: Cyst, dermoid, oculomotor palsy, proptosis

Dermoid cysts are choristomas resulting from the inclusion of ectodermal tissue during closure of the neural tube and occur along the epithelial lines of fusion. Within the head and neck region, dermoid cysts occur most typically in the orbitofacial region and are classified as frontotemporal, nasoglabellar and orbital. Frontotemporal dermoids are the most common type and typically present as an asymptomatic mass. We present a rare case of frontotemporal dermoid presenting as sudden onset oculomotor nerve palsy in young male patient. We highlight this case for its rarity, characteristic progression of tumor and complexity of surgical management.

Case Report

A 40-year-old Asian-Indian male presented to our clinic with the complaints of sudden onset diplopia of 5 days duration associated with headache and vomiting. He was a known diabetic and was on oral hypoglycemic agents. On examination, his best-corrected visual acuity was OU 6/6, N6. On further examination, it was evident that he had a left pupil sparing oculomotor nerve palsy and loss of corneal sensations along with a firm mass palpable in the left temporal fossa. On questioning, patient gave history of presence of "painless progressively increasing bump" on his left temple for 35 years [Fig. 1]. Magnetic resonance imaging (MRI) brain revealed a heterogeneously enhancing mixed intensity dumb-bell shaped lesion measuring 5.64 × 3.12 cm, with fat-fluid level noticed in the left orbit, along the superolateral aspect with the medial end extending up to the superior orbital fissure and anterior cavernous sinus. The lateral end of the cyst was extending into the infratemporal fossa [Fig. 2]. In the anterior cavernous sinus, the lesion was

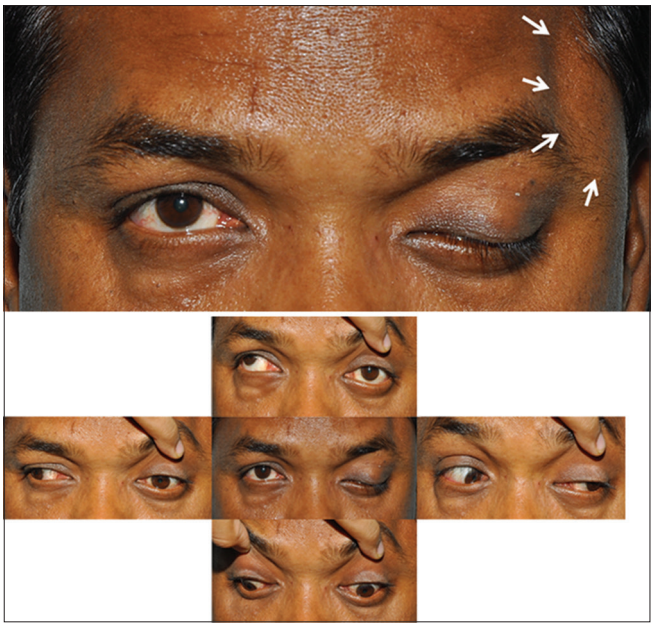

Figure 1: Top- external color photograph of the patient showing left ptosis with a temporal "bump" (arrows). Bottom- gaze photographs demonstrating oculomotor nerve palsy

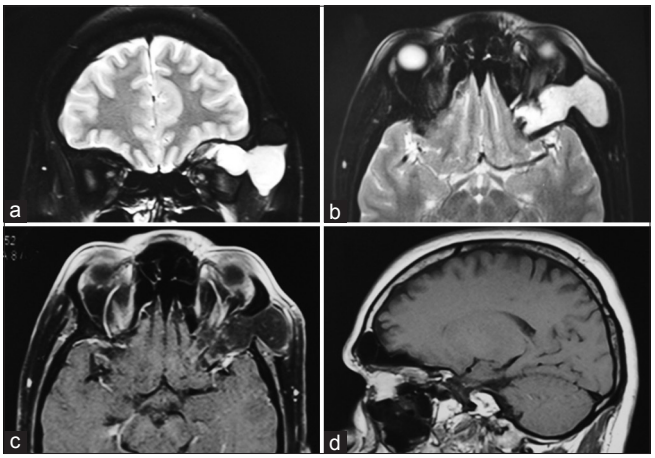

Figure 2: Magnetic resonance imaging showing heterogeneously enhancing mixed intensity dumb-bell shaped lesion with fat-fluid level (a) enhancement postcontrast (b) noticed in the left orbit along the superolateral aspect with medial end extending up to the superior orbital fissure and anterior cavernous sinus (c), extension of the lesion into the anterior cavernous sinus can be seen on sagittal section (d)

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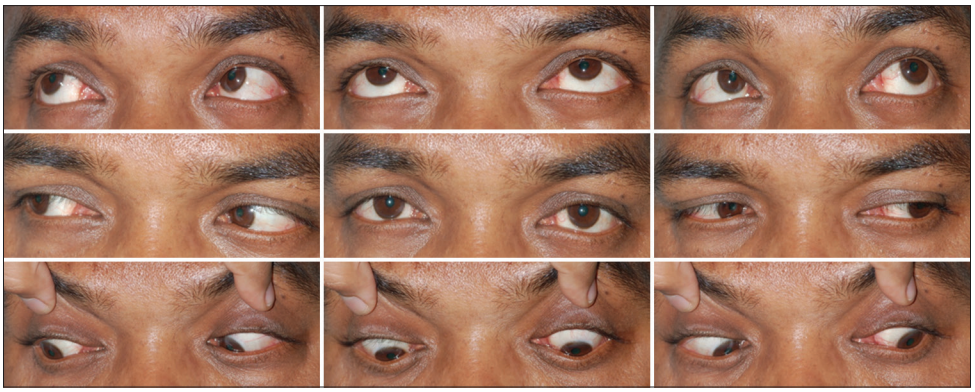

Figure 3: Two months postoperative photograph of the patient showing complete resolution of ptosis, orthophoria in primary gaze and recovery of ocular movements in all gazes

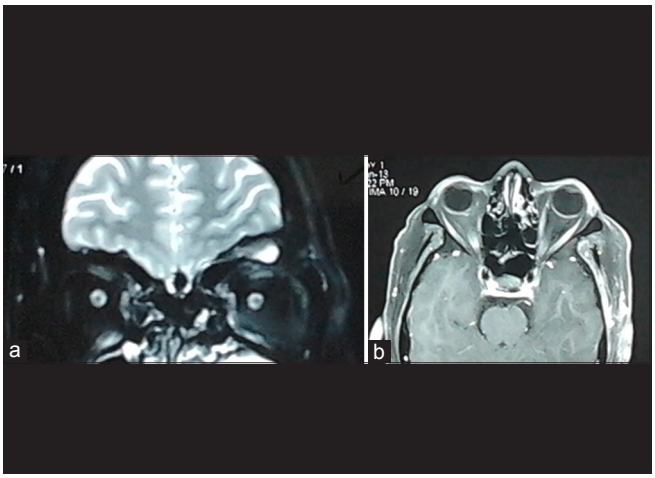

Figure 4: Two months postoperative magnetic resonance imaging photograph showing postoperative changes and granulation tissue with complete excision of the dermoid. There is no evidence of residual lesion

compressing the oculomotor and the ophthalmic division of the trigeminal nerve. On the basis of the findings, a clinico-radiological diagnosis of frontotemporal intraosseous dermoid cyst was made.

Patient underwent total excision of the dermoid cyst using a left infratemporal extradural approach. A formal temporal craniotomy was not performed; instead the lesion was approached through the left temporal bony defect due to erosion caused by the tumor itself. After excising the superficial part of the lesion with capsule, the microscope was used to further navigate and drill the bone to excise the lesion in toto. Under the microscope, left sphenoid bone was drilled up to the superior orbital fissure. The dermoid was extending to the left superior orbital fissure and reaching the left anterior cavernous sinus region where it was compressing the nerves in the lateral wall of the sinus. The dermoid cyst was soft yellowish with semi-solid cheese-like consistency with a thin-walled capsule. The capsule was radically excised and also scrapped off along with the periosteal layer of the bone. In the immediate postoperative period, his ophthalmoplegia and ptosis started improving.

Two months postoperatively, patient recovered well with marked improvement in diplopia and ptosis [Fig. 3] Follow-up MRI study showed postoperative changes with granulation tissue at the lateral wall of the cavernous sinus wall, without evidence of any residual mass [Fig 4].

Comment

Frontotemporal dermoids are the commonest group in the orbitofacial region accounting for 64% of cases in one of the largest series.[1] They usually present as discrete slow growing masses, located superficially with no deep extension and can be managed by direct excision biopsy via an upper eyelid incision.[2] Deep orbital dermoids may occasionally extend through the lateral orbital wall to secondarily involve the temporal fossa, sometimes in a "dumb-bell" or "collar-stud"pattern. In these cases, the intraorbital component of the dermoid usually predominates relative to the extraorbital component, and ophthalmic manifestations such as proptosis or ocular motility disturbances or both typically are present. Intracranial dermoid cysts typically increase in volume slowly over time and tend to grow in intracranial subarachnoid spaces that offer minimal resistance, such as cisterns, sulci or fissures.[3] The growth rate of the dermoid cyst significantly affects the clinical presentation. Primary cavernous region dermoids present with oculomotor nerve palsy early in the course of the disease.[4]

However, our patient had the temporal swelling for many years preceding the onset of oculomotor paresis and is therefore a case of primary temporal dermoid extending to the orbit, superior orbital fissure and then anterior cavernous sinus; with MRI revealing the characteristic dumb-bell shaped tumor entering the orbit through a small bony window [Fig. 2]. In our patient, the oculomotor nerve and the ophthalmic division of the trigeminal nerve were involved and symptoms were present only for 5 days prior to presentation suggestive of progressive growth and enlargement of a primary temporal dermoid. To the best of our knowledge this unique presentation of primary temporal dermoid has not been described so far in the literature and reiterates the continuous slow growing nature of the tumor.

Magnetic resonance imaging permits the recognition of these lesions with their typical appearance as hyperintense signals on T1-weighted MRI and hypointense to mixed signals on T2-weighted MRI. Absence of peripheral edema due to slow growth with signal characteristics on T1-weighted and

T2-weigted sequences helps in the differential diagnosis of dermoids from cystic glioma, arachnoid cysts or other lesions.[5]

Excision of temporal dermoids can be complicated by factors such as thin capsular wall, liquefied cyst contents, large size, rupture or leakage, adhesions to surrounding tissues, and variable involvement of bone with potential extension to the intraorbital or intracranial compartments. Where the cyst is attached to the bone, careful subperiosteal dissection is prudent as intraoperative rupture is common at these sites.[3] If a portion of the dermoid is found to extend through a small bony aperture with pronounced expansion within the intraorbital or intracranial cavity, like in our patient, it is necessary to facilitate exposure by enlarging the bony opening to allow continuing dissection from the original approach. This was performed in our patient. Some surgeons prefer to open the second compartment by a formal lateral orbitotomy or craniotomy.

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