Case Report

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Giant Orbital Encephalocele with Proptosis Due to Sphenoid Wing Dysplasia Associated With Neurofibromatosis 1: Soft Tissue Repair

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Abstract

A 35 year female with progressive proptosis of right eye since childhood with diminution of vision presented with a swelling extending into the upper eyelid with a pulsatile proptosis. Globe deviated inferionasally with a visual acuity of finger counting at 1 meter. 3D-NCCT/MRI- Complete aplasia of the right sphenoid wing with an intraorbital temporal lobe meningoencephalocele. Dura and orbital tissue were inseparable due to adhesions. Redundant dura was plicated and sutured to the surrounding dura & falxcerebri (> 270° periorbital circumference) with no additional mesh construct. Post op improvement in vision to 6/9 with minimal enophthalmos was seen.

Keywords: Sphenoid Wing Dysplasia; Orbital Encephalocele; Neurofibromatosis-1; Encephalocele; Proptosis.

Introduction

Neurofibromatosis1 (NF1) is an autosomal dominant genetic disorder with an incidence of approximately 1 in 2600 to 3000 individuals. Sphenoid wing dysplasia/aplasia is a rare congenital anomaly occurring in 3-11% of NF1. They are mostly asymptomatic and incidentally detected on screening investigations of the brain, but rarely can they present with orbital meningoencephalocele. It can mimic a plexiform neurofibroma/lymphangioma and can result in catastrophe if not suspected. Orbital encephalocele presents with relentlessly progressive pulsatile proptosis. Visual deterioration occurs as a late event and is the cause for presentation in developing countries. The intraorbital content is a form of traction encephalocele of the temporal lobe into the meningocele. Principles of surgical management would include truncation of the redundant dura and reconstruction of the orbit. Care should be exhibited while separating the dura from periorbital tissue otherwise would result in dire consequences of permanent visual loss, strabismus. Repair with bone and mesh can be dangerous as there are no bony anatomical landmarks in an aplastic sphenoid wing for the placement of screws. Important soft tissue structures, major vessels at the base of the skull would mean that the constructs may not completely obliterate the communication between intracranial and intraorbital compartment. These features led to our assumption of simple soft tissue repair. We thus proposed that a reduction & plication of the redundant dura with anchorage to the surrounding dura maintains an adequate orbital volume. Bony or mesh constructs will be benevolent if there are sufficient bone landmarks for placing the screws.

Case Report

A 35-year-old female presented with progressive enlargement of the right upper eyelid, progressive proptosis since childhood and diminution of vision in the right eye of 1-year duration. The patient was operated upon 15 years back by a right temporal approach at a district hospital but showed no improvement (details not available). Due to financial constraints and with no symptoms of visual deterioration, she had neglected to seek further

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medical help. No prior history of trauma or family history of neurofibromatosis was present. On examination, there was a diffuse expansile/pulsatile swelling extending into the upper eyelid of 8cms (width) x 6cms (height) x 4cms (anterior/posterior) **(Figure 1a & 1b)**, with an impulse on cough and was brilliantly trans-illuminant **(Figure 1c)**. The eyeball deviated inferionasally with minimal movement of upto 20° in all the planes. A visual acuity of finger counting at 1 meter was noted. On lid retraction, the visualized globe had prolapsed till the level of the tip of the nose with evident attachments of the recti muscles **(Figure 1d)**. No evidence of exposure keratoconjunctivitis. A 5cm linear healed surgical scar running over right zygomatic arch was noted with a defect of the lateral orbital rim (iatrogenic) through which an expansile pulsation was felt. Two neurofibromas in the left thigh and multiple inguinal/axillary freckles &café au lait spots were visualized in the trunk.

Computed tomography (CT) scan with 3D reconstruction revealed right sphenoid wing aplasia with resultant encephalocele into the right orbit. Right orbit also appears enlarged in comparison to left orbit **(Figure 2a; 2b & 2c)**. MRI brain showed meningocele is arising from the right temporal fossa dura with temporal lobe encephalocele. The perisylvian and chiasmatic cisterns are also enlarged. The redundant dura is thick and appears fibrotic; encephalocele has a pencil tip tapering; suggestive of thick arachnoid bands connecting the anterior temporal lobe pole to redundant intraorbitaldura – "traction encephalocele" **(Figure 2d)**. Right globe appears to be pushed inferiomedially, and the entire optic nerve is pushed medially and severely stretched by the intraorbitalmeningocele**(Figure 2e & 2f)**. No plexiform neurofibroma, optic nerve glioma & schwanomma, gliosis of temporal lobe noted.

Right frontotemporal craniotomy approach. Dura could not be separated from the periorbita all around the orbit. On opening the dura and CSF drainage, the prolapsed orbital contents regressed. Intradurally, thick arachnoid adhesions were seen extending from the tip of the right temporal lobe to the inner aspect of the redundant intraorbitaldura**(Figure 3a)**. Adhesiolysis was done, and the temporal lobe tip appeared healthy(no gliosis). Middle cerebral artery, cavernous sinus, optic nerve & ophthalmic artery were visualized and preserved **(Figure 3b)**. The amount of bone purchase for reconstruction of the orbit using mesh was minimal and due to the abnormal orientation of the vital soft tissue in the skull base; mesh fixation was abandoned. Dura- Dura repair was performed; the redundant dura was reduced intracranially and plicated**(Figure 3c &3d)**. The edges were anchored to the surrounded rigid dura of frontal, temporal, ethmoid region and falx cerebri with 3-0 mersilk suture for > 270° area of the periorbital region leaving areas inferiomedially around the major vessels and the optic nerve **(Figure: 3e)**. This plication of dura resulted in sufficient debulking of orbital content and prevented excessive enophthalmos. An Omaya reservoir was placed into subarachnoid space. Valsalva maneuver after watertight dural closure showed no evidence of intraorbital prolapse. Dura closed primarily, and bone was deposited back.

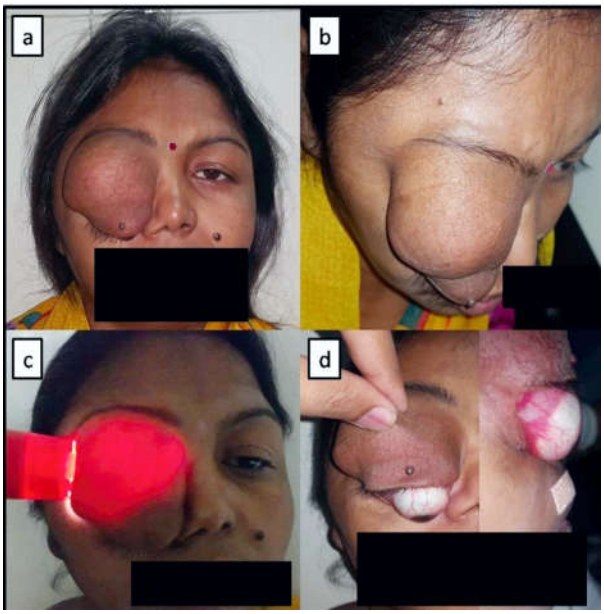

Fig: 1 (a &b). 8x6x4cms diffuse swelling with inferiomedially proptosis of the right eye, **(c).**Brilliantly transilluminant, **(d).** On lid retraction, recti muscle insertions are visible.

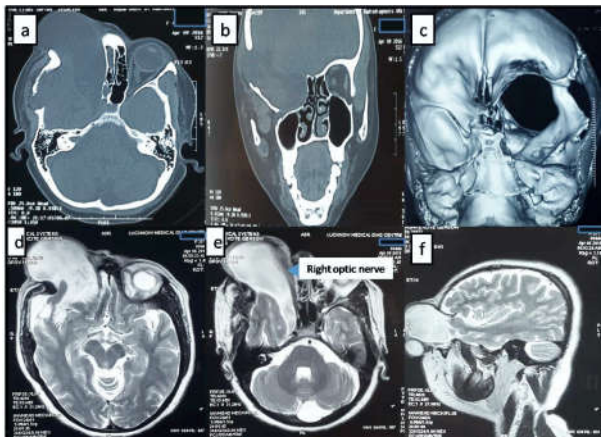

Fig: 2 (a; b & c): NCCT with 3DCT – Complete aplasia of the right sphenoid wing with a right lateral orbital defect (Iatrogenic) with enlarged right orbital foramen, **(d; e & f):** MRI T2W1 shows giant orbital meningoencephalocoele. Dilated perisylvian cisterns and chiasmatic cisterns with inferiomedial displacement of right eye to the level of tip of nose with stretched right optic nerve.

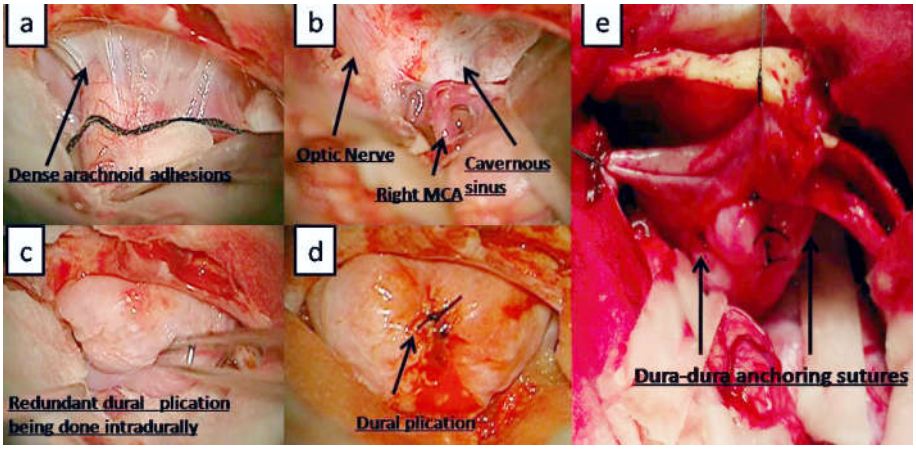

Fig: 3 (a & b). Dense arachnoid bands between the dura and temporal lobe. All the key structures visible with no evident bone, **(c & d).** Redundant dura being pulled inside the temporal fossa for plication. **(e).** Dura-dura anchoring sutures.

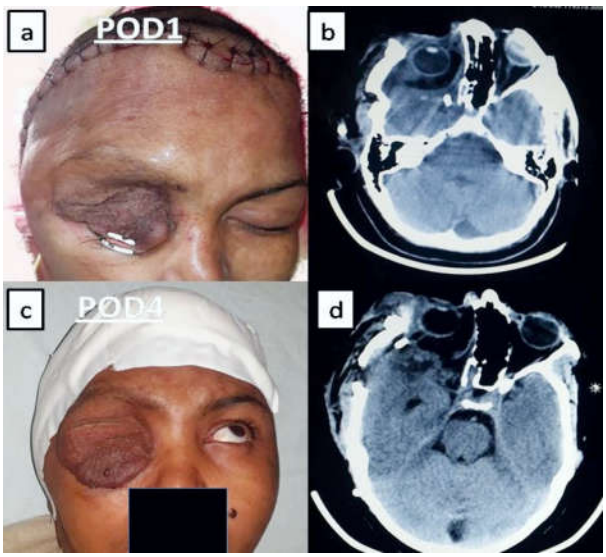

Fig: 4 (a-d). Immediate post op resolution of proptosis with evident mild enophthalmos on NCCT

Post operatively patient showed minimal enopthalmos and external ophthalmopareses on day 1 **(Figure 4a & 4b)**, which improved by day 4 **(Figure 4c & 4d)**. Temporary tarsorrhaphy reversal was done on day 3. Most importantly, there was visual acuity improvement to 6/18. Follow-up at three months, showed no recurrence of symptoms. Further reconstructive orbital surgery for correction of orbital floor defect by plastic surgery is awaited as the right eye globe appears to be depressed inferiorly due to a defect in orbital floor.

Discussion

NF1 is a rare autosomal dominant familial disorder with an incidence of 1 in 3,500 individuals [4].Various skeletal anomalies are known to occur with the most common feature being scoliosis. Sphenoorbital dysplasia occurs in 4- 11% of NF1 cases and complete unilateral sphenoid wing aplasia which are considered pathognomonic of NF1 has been documented in few anecdotal reports only [4]. Jacquemin et al. however found that a sphenoid bone disorder is a secondary dysplasia following the effect of contiguous plexiform neurofibromas on the development of skull or orbit [8,9]. In most of the case reports, incidentally detected dysplasia in an otherwise asymptomatic patient is the most common form of presentation. Rarely, in less than 5% of cases, it can present as a pulsatile exophthalmos with proptosis causing significant craniofacial deformity [3]. It is worthwhile to do a lateral tarsorrhaphy and avoid corneal and optic nerve injuries. While many authors term the intraorbital content as transorbitalencephalocele (11), brain herniation [12], cranio-orbital-temporal neurofibromatosis [6], the common entity in all these cases is intraorbital meningocele of temporal dura with encephalocele of the temporal lobe. The driving force for such pathogenesis is yet to be postulated. Several possible mechanisms are; pathological absence of the sphenoid wing; an associated intracranial tumor with raised ICP; constant and everlasting CSF pulsation with enlarged CSF cisterns resulting in meningocele; dense arachnoid adhesions leading to a type of "traction herniation of the temporal lobe". A case report of severe enophthalmos associated following resection of an astrocytoma associated with this pathology suggests the need for a bony or metallic construct to prevent dissipation of intracranial and intraorbital pressures [7]. Rangarajan V et al. reported a case of posterior herniation in NF1 treated by lumboperitoneal shunt alone with complete resolution [12]. Thick intraorbitaldura and adhesions

to periorbita and orbital soft tissue may impair a plane of dissection and can be hazardous [5,10]. Unless this plane is established, extradural reconstruction of the orbit is dangerous.

Resection of gliotic temporal lobe tip may be needed, but can also be avoided [2]. Reconstruction of the orbit is important for both cosmetic and functional reasons and had been demonstrated by Dandy in 1929 [2].

Reconstruction of the orbit and the sphenoid has traditionally been done using autologous rib/ fibular/ calvarial bone grafts, but these have inherent problems of bone resorption, infection, and failure. Titanium plates (radial/ conical)in conjunction with bone grafts have been used [1]. There are no case reports of soft tissue repair by plication and anchorage. A definitive repair of the orbit by bone graft or titanium mesh graft is warranted as they give the superior cosmetic outcome. Nevertheless, a long duration follow-up of such cases is required.

Conclusion

Sphenoid wing dysplasia may be congenital or tumor-induced dysplasia of the sphenoid bone; hence, always look for intracranial tumors.Lateral tarsorraphy preserves corneal and avoids perioperative optic nerve injury.

Mesh reconstruction with or without bony strut is the ideal and safe surgical option, but in the event of difficulty, a dural plication with dura-dura anchoring method may suffice. Arachnoid band release prevents encephalocoele.

Patient's Consent

Patient and Patient's husband has given consent to submit the case report which includes detail of the case and clinical photographs of the face and head region of the patient following which the identity of the patient is revealed.

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