CASE SERIES

SPHENOID WING DYSPLASIA: REPORT OF 3 CASES

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Sphenoidal Dysplasia is the absence of complete or a part of sphenoid bone, most commonly the greater wing of sphenoid. It can occur as an isolated deformity or in Neurofibromatosis-1 (NF1). Features of NF1 include café au lait spots, inguinal or axillary freckling, neurofibromas, optic gliomas, scoliosis and tibial deformity. Our study is retrospective case series of 3 cases of Sphenoid wing dysplasia. There was 1 case of isolated bone defect, 1 case of NF-1 and 1 case of operated Craniofacial Fibrous Dysplasia involving the sphenoid wing. There were 2 primary operated cases while 1 was operated secondarily. There was resolution of pulsatile exophthalmos in patient with sphenoid and temporal bone defect. Patient with facial deformity NF1 was debulked to the satisfaction of the patient, the patient however declined surgery to correct the sphenoid bone deformity. The 3rd patient was a re-do surgery patient in which the previous implant material was removed and the CSF rhinorrhoea, the patient did not consent to the correction of vertical orbital dystopia. Sphenoid wing dysplasia is a complex deformity requiring multi-specialty care and treatment planning. With meticulous planning and surgery, good results can be achieved as shown in our case series.

Keywords: Neurofibromatosis; Sphenoid Bone; Craniofacial Fibrous Dysplasia; Craniotomy

INTRODUCTION

Sphenoidal dysplasia can occur alone or as one of the features of Neurofibromatosis 1(NF1), the incidence of NF1 is 1 in 2500 to 33000 live births.1,2 Sphenoid wing dysplasia is present in 5–7% of NF1 characterized by unilateral agenesis of greater wing of the sphenoid3 NF1 is characterized by at least 2 or more of the following features; café-au-lait-spots, neurofibromas, freckling in the axillary or inguinal regions, optic glioma, lisch nodules (hamartomas of the iris), a distinctive osseous lesion such as sphenoid dysplasia or tibial pseudo-arthritis and a first degree relative with NF-1.4 Jackson et al4 have classified orbital-temporal neurofibromatosis into 3 groups; Group 1 has orbital soft-tissue involvement only with a seeing eye; Group 2 has orbital soft-tissue and significant bone involvement with a seeing eye; and Group 3 has orbital soft-tissue and significant bone involvement with a blind or absent eye. The specific pathogenesis of the condition remains controversial, suggested mechanisms include abnormal development of the mesoderm or acquired via secondary processes.5,6 The earliest reported procedure was done by Dandy.7 He did a transcranial bone graft for sphenoid dysplasia It was not until 1980s that cases were being reported in the literature. Surgical procedures described in these case reports had a wide spectrum.8-14 Graft materials that have been used are calvarium, split rib, iliac crest and outer cortex of mandible in decreasing order of use. The bone grafts can be fixed with plates and screws, screws alone or wires and bone dust (which is collected during craniotomy) mixed with fibrin glue.15 Each graft material has its advantages and limitations. Resorption of bone grafts is one of the key limitations in the use of autologous bone material.16-18 Titanium mesh is easy to manipulate into desired place and shape, however its removal if at all required, is difficult as there is tissue growth into the holes of mesh.18,19 To prevent the growth of fibrous tissue through the holes of titanium plate, use of titanium- reinforced porous polyethylene sheets3 has been proposed. Titanium and bone graft composite reconstruction of posterior sphenoid wing has also been reported to yield satisfactory long-term result.3

CASE 1

A 7 years old male child presented to our outpatient department with the chief complaint of eye deformity which was pulsatile. Systemic medical examination and family history were unremarkable. On clinical examination there was a soft area in the right temporal region with inconspicuous pulsations. The CT scan showed a non-confluent temporal bone defect and a 3×2.7 cm defect in the orbital roof.
According to the parents of the child, soon after birth the child had bulging of the right eye which became progressively worse with age. Ipsilateral pulsatile exophthalmos was also noted with normal extraocular movements of the affected eye. Exophthalmous was 28 mm as measured on Hertel’s exophthalmometer. MRI brain revealed herniation of the cerebral tissue into the orbit which was pushing the eye ball anteriorly, there was no intracranial mass. The lack of bony barrier was transmitting the pulsations to the eye. Patient was prepared for surgery according to institutional practice. Baseline investigations were done. Two units of red cell concentrate (RCC) were arranged before surgery and were available on demand. The funduscopic examination was normal. Neurological consultation was done which showed no significant finding. The case was discussed in a multidisciplinary team consisting of Craniofacial Surgeon, Neurosurgeon, Oral and Maxillofacial Surgeon, Anaesthetist and Radiologist. Preanaesthesia assessment was done by anaesthesia department and the patient was deemed fit for surgery in ASA-1 grading. Cuffed orotracheal intubation was carried out by the anaesthesiologist. CVP line was inserted into right subclavian vein for fluid infusion and blood transfusion. Access was gained through bicoronal flap with zigzag incision with elevation of temporalis muscle. The frontal and lateral orbital rims were removed enblock after securing screw holes for accurate post-surgery bone alignment. The frontal bone panel was removed by craniotomy. The frontal panel was placed in sterile saline for the duration of the surgery. The neurosurgeon separated the dura from the frontal, sphenoid and temporal bone. An anomalous venous channel was encountered during the dissection between the cerebral tissue and the orbital contents, leading to troublesome bleed. The bleeding was controlled with no major blood loss. The defect was visualised after careful separation of dura mater from the bone and orbital contents with gentle dissection. There was a minor dural tear which was repaired. The defect was measured with a template which was used as the reference for bone graft. Calvarial bone graft was harvested from the inner table of the preserved frontal panel and outer table was placed back into its original position afterwards. The bone graft was secured in place with 5–6 mm mini-screws and mini-plates laterally. Care was taken to avoid trauma to the dura and orbital contents during graft placement. All the sharp edges of the graft were burred/smoothed by a powered bur. Adequate space was spared for optic nerve medially. The inner table from the frontal bone was also used to fill the temporal bone defect and was fixed with 1.5mm plates and screws, the remaining bone gaps were filled with bone dust.

The antibiotic regime consisted of amikacin 15 mg/kg/day every 8 hours and vancomycin 10 mg/kg every 6 hours daily intravenous for 05 days with use of opioids as analgesics. Two non-suction drains were placed over the coronal aspect and they were removed once the drainage was less than 20 ml per 24 hours. The patient was kept in a propped up 30° head elevation in intensive care unit for the first 2 days after which he was shifted to surgical ward. Postoperatively funduscopic examination and general eye examination was carried out. No neurological/ophthalmological deficit was detected. There was an episode of bleeding (suspected venous bleed) on the 6th postoperative day, the patient was shifted to intensive care unit and was observed for 2 days. He developed redness and swelling of upper and lower eyelids on right side which did not increase on serial examination and assessment. No decrease in visual acuity was detected on serial ophthalmological examinations. The patient was found hemodynamically stable and the swelling and ecchymosis remained non progressive and settled without intervention. After surgery, the patient had immediate cessation of pulsations in the eye and was satisfied with the result. The patient came back after 1 year, there was no pulsation in the eye. The patient was advised postoperative CT scan which he declined.

CASE 2

A 35 years old male patient presented to us with the chief complaint of facial disfigurement and requested an improvement in facial appearance. On clinical examination he had grossly enlarged right side of face with pulsatile exophthalmos. The patient had plexiform neurofibromatosis of face. The ophthalmological examination revealed lisch nodules (hamartomas of iris) of right eye, he had reduced visual acuity with only perception to light. Apart from a history of fits in the past, patient didn’t have any other neurological issue. He had his last attack of seizure more than 5 years back. Contrast enhanced CT scan revealed extensive subcutaneous facial mass and an expanded orbit. The aim of the surgery was reduction of facial bulk and facial lift so as to make him acceptable in society. The patient was counselled regarding need to improve the orbital volume and shape, but he refused and wanted facial debulking surgery only. Preoperative work up and case discussion was done according to institutional practice. Four units of red cell concentrate were arranged for surgery as plexiform neurofibromas have a propensity to bleed profusely. Since the patient didn’t consent to any treatment for pulsatile exophthalmos, the transcranial approach was not applied for access during surgery. Standard Face lift incision was used. Skin excisions were done for debulking the face. Generous use of eletrodiathermy was done for haemostasis as...
there was troublesome bleed and patient was transfused 2 units RCC during the operation. Reduction of upper and lower eye lids was performed using respective eyelid reduction incisions. Fascia lata was harvested and used as a fascial sling for elevation of angle of mouth with insertion to temporal bone with 5-6mm screws. Elevation of lower eye lid was also done with fascia lata sling fixed to the temporal and nasal bone with 5mm screws. A negative suction drain was used to prevent hematoma formation. Tarsorrhaphy was done for 2 weeks. A custom-made compression bandage was used in the post operative period to reduce oedema and prevent hematoma formation. The post operative recovery was uneventful. Patient was shifted to intensive care unit for 2 days after which he was shifted to ward. The follow up continued up to 1 year and remained satisfactory.

**CASE 3**

A 19-year-old male patient reported to our outpatient clinic with the chief complaint of watery discharge from the nose and a bulging left eye. The patient had vertical orbital dystopia. On detailed history it was revealed that patient had a surgery 6 years ago for a prominent left eye. According to patient’s father, surgery for an unknown benign pathology was carried out. The surgical procedure did not improve the chief complaint of the patient. Another surgery was done in March 2019 in which titanium mesh was placed on the skull (Figure-1) and an unknown implant material was placed in the orbital roof by the neurosurgeon (Figure-2,3). Following surgery, the patient complained of a persistent discharge from the nose. On clinical examination there was cerebrospinal fluid rhinorrhea and non-pulsatile proptosis of the left eye. CT Scan images showed a defect in orbital roof (Figure-3) and vertical orbital dystopia. We suspected a thin radiolucent material used to separate the orbital cavity and the brain at the level of lesser wing of sphenoid which was visible in one of the slices of the CT scan. Consent for removal of the alloplastic material, if required and the repair of dura was taken from the patient. The patient didn’t consent to be operated for vertical dystopia as he did not complain of double vision and wanted to delay the corrective surgery for that. His main concern was CSF leakage and correction of exophthalmos. The patient was put under general anaestheia with orotracheal intubation, note the extent of exophthalmos in supine position (Figure-4). Previous incision was modified into a zigzag incision and extended bilaterally in the preauricular crease. Craniotomy was performed by the Neurosurgeon to access the defect. A titanium mesh used to reconstruct the frontal bone defect on the left side was removed to access the cranial cavity. A large sheet of artificial dura used on the frontal lobe to reconstruct the dura was removed (Figure-5). CSF was found to be leaking from a large defect in the dura, which was repaired with fascia lata from the thigh. The nasofrontal duct was found plugged with bone wax for obliteration, this plug of wax was removed. The remanants of the frontal sinus and nasofrontal duct opening were demucosalized and free fat graft from the thigh was used to replug the defect. Bone dust was placed over the free fat graft and reinforced with fibrin glue. The defect site at the roof of the orbit was exposed, a PTFE sheet was removed. The patient’s medical records did not specify the material used. The bone defect after removal of the titanium mesh was reconstructed with clavarial bone graft from the non-dominant side (Figure-5). A separate piece of calvarial graft was used to reconstruct the supra orbital ridge left side, which was deficient and had been covered by the hanging edge of the titanium mesh to give it a smooth outline. The bony defect in the orbital roof was filled with a split thickness calvarial graft fixed with mini-screws and mini-plate bent to configuration. Post operative CT scans of the reconstructed orbital roof and cranial defect (Figure-7,8). Effort was made to follow the contour of the orbit roof to normalize the orbital volume. The wound was closed in layers after achieving haemostasis. Two non-suction drains were placed and were removed when the drainage was less than 20 ml per day. Patient reported immediate cessation of the watery discharge from the nose and an improvement in facial profile with respect to a better cosmetic outcome of the bulging eye (Figure-6).

**Figure-1**: Preoperative 3D CT scan of the patient, there is a gross difference in orbital volumes, the left orbit is shallower with low orbital roof. Vertical orbital dystopia can be appreciated. Also note the titanium mesh in place from the previous surgery.
Figure-2: Titanium mesh extending into the infratemporal fossa. It has also been utilized to cover the supraorbital bone deficit on left side. The red arrow shows the defect in temporal bone.

Figure-3: Removal of PTFE (black arrow) and free fat graft from previous surgery (blue arrow).

Figure-4: Inadequate placement of titanium mesh, an unknown graft material (yellow arrow) also visible in CT scan slice, this came out to be PTFE membrane. Also notice the depressed orbital roof reconstructed with synthetic material (blue arrows).

Figure-5: Exophthalmos left eye even on supine position.
Figure 6: Marked reduction in exophthalmos following surgery.

Figure 7: Artificial dural substitute being removed.

Figure 8: Split thickness calvarial bone graft harvested from parietal bone (blue arrow), a separate graft was used for supraorbital bar on same side (yellow arrow). The graft was secured into the frontal bone defect (black arrow) and secured with AO miniscrew/miniplate system.

Figure 9: Post operative superior view.

Figure 10: Frontal view with adequate eye movement.
Figure-11: Defect in orbital roof reconstructed by split thickness calvarial bone graft (yellow arrow), fixed with osteosynthesis.

Figure-12: Frontal defect reconstruction done by split thickness calvarial graft (red arrow), two bars of split thickness calvarial graft (yellow arrows) were harvested and were used to reconstruct the frontal bar and the temporal bone (blue arrows).

Table-1: Demographic and clinical data of patients

<table>
<thead>
<tr>
<th>S No</th>
<th>Sex</th>
<th>Age</th>
<th>Diagnosis</th>
<th>Primary surgery/redo surgery</th>
<th>Surgical procedure</th>
<th>Comments and/or complications</th>
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| 1    | 7 y | Male| Sphenoid and Temporal bone defect of right side   | Primary surgery             | Calvarial bone graft through transcranial approach                                | 1. Resolution of pulsatile exophthalmos  
2. Bleeding episode causing peri orbital swelling, resolved without any operative intervention. |
| 2    | 36  | Male| NF type 1 (neurofibromatosis right side face, involvement of right eye) | Primary surgery             | De-bulking of facial/orbital tissues                                              | Satisfactory esthetic result, patient did not consent for sphenoid defect surgery               |
| 3    | 19  | Male| Craniofacial Fibrous Dysplasia                    | Redo surgery                | 1. Bone graft in sphenoid bone defect after removal of infected PTFE implant and artificial dura  
2. Dural repair with fascia lata, sphenoid defect reconstructed with calvarial bone graft.  
3. Frontal sinus was curetted, bone wax removed, packed with fat graft, bone dust and fibrin glue | 1. Patient was satisfied with the result. Post operatively, there was no CSF rhinorrhea  
2. The patient did not consent to the correction of vertical dystopia |

DISCUSSION

Sphenoid wing defects can occur due to a number of reasons, they can be isolated or associated with Neurofibromatosis 1. Raised intracranial pressure can also contribute to this defect. Another hypothesis is that the malformations could arise due to mesodermal and neur ectodermal deformities. Neurofibromatosis 1 is a genetically inherited disorder, it results from mutation in NF-1 tumour-suppressor gene. The diagnostic criteria of NF 1 consist of two or more than two of the clinical features such as family history of NF2, vestibular schwannomas alone or in combination with meningioma, glioma, juvenile cortical cataract etc. The skeletal deformities of Neurofibromatosis are present in almost half of the patients, these are absence of greater wing of sphenoid bone, scoliosis, tibial pseudoarthrosis, meningocoele and macrocephaly. The partial or complete absence of the greater wing of the sphenoid
bone is one of the features of NF 1. This defect could extend to include the superior orbital fissure, hence enlarging it. The absence of this barrier between the orbital contents and the brain can present as proptosis or vertical orbital dystopia (if long standing). Other manifestations visible on the face are lisch nodules and plexiform neurofibromas. The treatment for a patient with Neurofibromatosis type 1 is multidisciplinary. The team consists of Ophthalmologist/Oculoplastic Surgeon, Oral and Maxillofacial Surgeon, Plastic and Reconstructive Surgeon, Orthopaedic Surgeon and a Neurosurgeon. Other specialties like Psychiatry and Genetesisist can also be involved in a case-to-case base. Optic gliomas are diagnosed on MR scan and most are asymptomatic and require no surgical intervention. The patients are concerned with the facial deformity caused by the plexiform neurofibromatosis. The surgical correction is often unsatisfactory and incomplete. This can be due to the involvement of nearby nerves and the tendency for recurrence. 23-25

Fibrous dysplasia is a benign disease in which bone is replaced by fibrous connective tissue. Craniofacial fibrous dysplasia can cause functional as well as aesthetic problems. These can present as swelling of midface and frontal bone. Functional deficit can arise due to compression of optic nerve, nasal blockade and lacrimal duct blockage. The decision to perform surgery depends upon a number of factors including urgent orbital decompression as well as the degree of aesthetic deformity. In case of orbital decompression there can be a post-operative defect in one of the orbital walls, we encountered a case in which there was a defect in sphenoid bone and hence loss of barrier between the content of two cavities.

Whatever the cause, the primary aim of the surgery is to place an inert material as a permanent and stable barrier between the orbital cavity and the middle cranial fossa. This can be achieved through a lateral approach or a transorbital/transcranial approach. 26 For good access transorbital/transcranial approach is preferred. There are mainly 2 types of materials to be used as barrier materials that can be placed between the middle cranial fossa and the orbital cavity, that is autogenous bone for example iliac crest or calvarium and the alloplastic materials like titanium mesh and acrylic based materials. Some surgeons have reported good aesthetic outcome due to the ease of manipulation of iliac crest into the desired shape27, there is however risk of increased donor site morbidity. Outer cortex of mandible has also been used as a donor site for bone graft28. The advantages of a calvarial bone graft include the availability of bone near the operation site, the calvarial bone has less graft resorption as compared to other autogenous graft materials as it has more cortical content and less spongyous/cancellous bone. Major disadvantage is the difficulty in bending the graft to the required shape and size. However, calvarial bone has a natural convexity which was utilized in this series to reconstruct the orbital roof and provide adequate volume for the herniating orbital contents. Overall, the bone grafts have the advantage of being cost effective and inert as opposed to alloplastic materials. The advantage of using an alloplastic material such as a titanium mesh is the ability to bend the malleable titanium mesh according to the shape of the defect. 29,30 The risk of infection with titanium implants is extremely low but a potential problem could arise during the removal of implant due to adhesions through the mesh. 31 However the metallic mesh can easily be seen on a CT scan and the post operative CT scan can verify the position of the mesh. To our knowledge the chances of titanium implants getting infected is negligible. Histological studies have proved the safety of titanium implants/mesh.31 All efforts should be made to cover the implant surface with soft tissue and the implant/screw should be fixed adequately.

CONCLUSION
Sphenoid wing defects can occur in isolation, as a part of Neurofibromatosis or they can occur in postsurgical defects (as in our case, an operated case of Fibrous dysplasia). A stable and non-resorbable material such as a cortical graft alone or in combination with titanium mesh is recommended. Artificial dura should be avoided whenever possible and autogenous tissue preferred. Brain tissue should be isolated from orbital tissue with dural seal and permanent barrier for a stable result.

REFERENCES