Reconstruction of Sphenoid Wing Dysplasia with Pulsating Exophthalmos in a Case of Neurofibromatosis Type 1 Supported by Intraoperative Navigation Using a New Skull Reference System

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ABSTRACT

A patient with neurofibromatosis type 1 had pulsating exophthalmos of the right eye with diplopia resulting from severe dysplasia of the sphenoid bone and consecutive herniation of the right temporal lobe. The right orbital tectum was reconstructed with titanium mesh and iliac spongiosa via a lateral orbitotomy using intraoperative navigation. For intraoperative referencing a cortical fixed-reference system and a skin scanning laser device were used. Postoperatively, the diplopia was reduced, but the patient asked for further treatment and the procedure was repeated 6 months later. Intraoperatively, the previously implanted titanium mesh was deformed and most of the transplanted bone was resorbed, probably because of pressure. A more extended mesh was implanted and iliac spongiosa was placed on both sides. Intraoperative navigation was used during both procedures. The adverse effects of diplopia were minimized and follow-up computed tomography after seven months confirmed that the bone graft was in place.

KEYWORDS: Neurofibromatosis type 1, sphenoid dysplasia, intraoperative navigation

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Fewer than 1% of patients suffering from neurofibromatosis type 1 (NF-1) are reported to have abnormalities of the orbit.¹ Usually, this manifestation occurs unilaterally and is often associated with plexiform neurofibromas in the distribution of the trigeminal nerve.² However, Riccardi reported that sphenoid wing dysplasia can occur either with or without a peripheral neurofibroma (pNF).³ Jackson et al⁴ classified orbitotemporal neurofibromatosis into three groups (group 1, orbital soft-tissue involvement only with a seeing eye; group 2, orbital soft-tissue and significant bone involvement with a seeing eye; and group 3, orbital soft-tissue and significant bone involvement with a blind or absent eye). Bone involvement mostly consists of a partial or complete absence of the greater wing of the sphenoid, being an enlargement of the superior orbital fissure with consecutive herniation of the temporal lobe.⁴

Surgical correction can be performed through an intracranial approach but is associated with a high complication rate¹ or when an upper lid incision or coronal flap is used.⁴ We suggested the lateral orbital approach as an alternative technique, using an available intraoperative navigation system. The use of computer-assisted simulation and navigation systems for the exploration of complex anatomic areas like the orbit has been reported to shorten overall operation time and to increase the safety of surgical maneuvers near delicate structures.⁵⁻⁷ However, in maxillofacial surgery established reference methods like head sets often interfere with established transfacial approaches, or as in case of using a Mayfield holder, do not keep the head mobile. Therefore, we demonstrate the use of a cortical fixed-reference system during reconstruction of sphenoid wing dysplasia in a patient with NF-1.

CASE REPORT

A 25-year-old man with NF-1 presented with pulsating exophthalmos of the right eye and diplopia resulting from severe dysplasia of the sphenoid

bone and consecutive herniation of the altered right temporal lobe (Fig. 1). Therefore, he belongs to group 2 according to Jackson.⁴ No cranial nerve deficit was present. Further signs of NF-1 were multiple café-au-lait spots and several Lisch nodules. The aim of the therapy was not primarily aesthetic improvement but to reduce the progressive diplopia while maintaining vision. Therefore, conservative reconstruction of the right orbital tectum using a mesh and iliac spongiosa via a lateral orbitotomy and intraoperative navigation (VectorVision², BrainLAB) was planned.

For intraoperative referencing a cortical fixed-reference system was used, one day after CE-certification. This "Latero Reference Star" obtained from BrainLAB was easily fixed in the hairy skin with one monocortical screw (Fig. 2). Therefore, the mobile intraoperative positioning of the head was affected only minimally. After its fixation, the periorbital skin area was scanned using a patient-registration laser device (z-touch, BrainLAB).⁸ The accuracy of this referencing method was checked by anatomical landmarks.

A modified lateral orbitotomy as revisited by Maroon and Kenderdell was used.⁹ However, we preferred an anterior curved incision with a lateral extension in the eyebrow. After the lateral orbital rim was replaced, fixation was performed with a miniplate (2.0, Mondeal Mini 2000) because intra-orbital pressure was suspected to be high. After exploration of the intraorbital displacement of the temporal lobe, severely thickened meningeals were visible (Fig. 3). Microscopic analysis of biopsies of the thickened meningeals and adjacent orbital fatty tissue excluded pNF at the site of the bony dysplasia. By opening the dura, the temporal lobe was repositioned and titanium mesh with iliac spongiosa was inserted as an overlay.

The patient's postoperative course was uneventful. His diplopia improved and he requested further treatment to enhance the chances of preserving his vision. Therefore, the procedure was repeated 6 months later. The lateral approach was extended

Figure 1 Preoperative 3D-model shows the osseous defect caused by the dysplasia of the right sphenoid wing (A). Primary axial CT reconstructions show the caudal dislocation and protrusion of the right eye (B) resulting from sphenoid wing dysplasia and consecutive herniation of the altered right temporal lobe (C).

Figure 2 The "Latero Reference Star" from BrainLAB is fixed on the skull with one monocortical screw. It is composed of three reflecting markers.

slightly medially. Intraoperatively, the titanium mesh was found to be turned and most of the transplanted bone had resorbed, probably because of pressure. A more extended mesh was implanted, and iliac spongiosa was placed on both sides (Fig. 4). His postoperative course was again uneventful and the adverse effects of diplopia were further minimized. Intraoperative navigation was used during both procedures (Fig. 5).

Follow-up computed tomography after seven months showed that the titanium mesh was in place. Sagittal reconstructed images revealed bone at the upper side of the titanium mesh bridging to the caudal border of the initial defect (Fig. 6).

Figure 3 The thickened meningeals of the right temporal lobe were explored using a lateral orbitotomy during the first surgical procedure. pNF was excluded histologically.

Figure 4 The titanium mesh implanted for reconstruction of the right orbital tectum during the second surgery was grafted with iliac spongiosa on both sides.

DISCUSSION

Reduction of diplopia in this patient with NF-1 associated with sphenoid dysplasia was successful and may be the first such case using the lateral orbital approach. In cases of NF-1 associated with altered anatomy of the skull base and therefore missing landmarks, intraoperative navigation offers a valuable tool.¹⁰ Because functional improvement preserving the vision was the main goal in treating this patient, a smaller approach was used instead of the standard surgical techniques. Small approaches provide only limited exposure and are a promising field for intraoperative navigation systems.⁵

When intraoperative navigation systems are used, their limitations must be kept in mind. Resolution of imaging data sets, errors of image fusion, and discrepancies during referencing increase inaccuracy before surgery. However, the main problem associated with intraoperative navigation is that topographic changes caused by the surgery can create discrepancies between the preoperative image data and the surgical site. Problem-oriented solutions for this problem involve intricate measures like intraoperative MR- or CT-imaging or the data transfer of navigated ultrasound information.^{11,12} With high-contrast structures, fluoroscopic systems enable intraoperative three-dimensional imaging, and they are easy to use and familiar.¹³

Preoperatively in our patient it was unclear whether the planned approach would be adequate or whether a more extended approach would be needed. Therefore, the use of established referencing methods like headset or the Mayfield headrest device would restrict intraoperative alternatives. Using the skull reference array proved to be easy and without complications.

If the surgeon is aware of the limitations of the intraoperative navigation system and regularly recalibrates using anatomic landmarks, then systems like the VectorVision² can be a useful supplement during the surgical exploration of complex anatomic regions, even for experienced surgeons. The skull reference

Figure 5 Intraoperative screenshot during second surgery shows retrobulbar navigation using fused CT and MR data sets.

array fulfills the demands of computer-assisted skull base surgery. In cases of NF-1, which is often associated with altered anatomy of the skull base, intraoperative navigation seems beneficial for orientation.

PAPER PRESENTED

Results were presented orally at the 10th Congress of the German Skull Base Society, Heidelberg, November 2002.

Figure 6 Follow-up computed tomography obtained 7 months after the second surgery. Secondary reconstructed axial (A) and sagittal (B) images revealed bone at the upper side of the mesh, bridging the caudal border of the initial defect. Laterally, the graft is connected to the lateral orbital wall.

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Commentary

Friedrich and colleagues describe a novel approach using frameless stereotaxy and facial incision for reconstruction of a sphenoid defect in a patient diagnosed with neurofibromatosis type I. This approach is less invasive than the extensive coronal incision/intracranial option. Another approach to this lesion would be to use a curvilinear incision extending superiorly from the root of the zygoma, creating a small craniotomy or craniec-tomy, and then placing the mesh into this region extradurally. Cosmetically, this incision may be less disfiguring because it is posterior to the hairline,

unlike the lateral extension of the eyebrow used in this case report.

The technique described in this case report is useful in the treatment of exophthalmos resulting from a sphenoid wing defect. However, we prefer an intracranial approach using frameless stereotaxy to treat cases of fibrous dysplasia/intraosseous meningioma, which have similar presentations. It is important to tailor the approach to the pathology. The authors also mention a minimally invasive ap-

proach as appropriate for the problem, but caution should be used in extending this approach to other causes of exophthalmos. This case illustrates the utility of frameless stereotaxy and the corresponding decrease in invasiveness in the correction of skull base defects.

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