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Frameless stereotaxy in a transmandibular, circumglossal, retropharyngeal cervical decompression in a klippel-feil patient: technical note

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Abstract Frameless stereotaxy, while most commonly applied to intracranial surgery, has seen an increasing number of applications in spinal surgery. Its use in the spine has been described to a greater degree in posterior rather than anterior surgical approaches, presumably due to the relative paucity of anatomical landmarks appropriate for frameless stereotactic registration in the anterior spine. This technical note illustrates the previously undescribed, successful use of frameless stereotaxy to the transmandibular, circumglossal, retropharyngeal surgical approach in a patient with Klippel-Feil syndrome.

Keywords Frameless stereotaxy · Klippel-Feil syndrome · Transmandibular, circumglossal, retropharyngeal approach

Introduction

The successful use of frameless stereotaxy has been described for placement of transarticular screws [13] and pedicle screws [7, 10] in the cervical, thoracic, and lumbar spine. Applying this technology in anterior spine surgery is more problematic because the anatomical landmarks necessary for device registration are relatively sparse. Nevertheless, its use has been described in cadaveric studies [1, 6, 8] and in a few clinical cases involving osteophyte resection, screw insertion for anterior cervical plate fixation [4], and transoral approaches to spinal pathology [11, 12]. The transmandibular, circumglossal, retropharyngeal (TCR) approach provides excellent exposure of the skull base and upper cervical spine, and may similarly benefit from

advantages of intraoperative navigation technologies, especially in patients with complex anatomy. Here we describe the successful application of computerized tomography (CT) wand intraoperative navigation to a TCR approach in a patient with Klippel-Feil syndrome with a previous occiput to T9 vertebra fusion.

Case report

This is a 44-year-old gentleman who presented with hoarseness and progressive inability to walk and swallow. The patient had a tracheostomy in place and was nearly wheelchair bound at the time of presentation. The patient was diagnosed with Klippel-Feil syndrome with craniocervical junction compression at 8 years of age. At

that time, a two-stage procedure was performed: a posterior craniocervical junction decompression followed by an anterior transmandibular circumglossal decompression. The patient did well following the procedures, but over the 2 years prior to his current presentation, he noted gradual decline in his ability to walk, swallow, breathe comfortably, and use his upper and lower extremities, eventually necessitating tracheostomy and percutaneous endoscopic gastrostomy (PEG) tube placement. An MRI revealed progressive basilar invagination and brain stem compression (Fig. 1).

A two-stage procedure was then performed on the patient. During the first stage, a revision posterior approach to the occipital cervicothoracic junction was performed. The patient underwent a C1-T1 laminectomy, decompression of the posterior craniocervical junction, and posterior fixation and fusion from occiput down to T9 (Fig. 2). During the second stage, a revision TCR approach to the anterior craniocervical junction was performed with the aid of a frameless CT intraoperative navigation system. The decompression of the anterior craniocervical junction was accomplished by complete corpectomies of the C1–C3 vertebrae and a partial C4 corpectomy.

The Surgical Navigation Networks (SNN) Computerized Tomography (CT) wand system (ISG Technologies, Inc., Mississauga, ON, Canada) was used for intraoperative navigation. [Of note, this company is no

longer viable. We are now currently using the Brain-LAB® navigation system for such indications (Brain-LAB, Inc.®, Westchester, Illinois, USA)]. This SNN CT wand system, like similar systems used for surgical navigation, involves preoperatively obtaining computed tomographic images of the patient's surgical anatomy, which are then integrated and correlated with patient's external anatomic structures. Thereafter, by using a probe ("wand") linked to this system, the operator may navigate within the surgical field by correlating the gross anatomy with computed tomography images obtained preoperatively. For this patient, preoperative CT of the head and upper C-spine with two-dimensional sagittal reconstructions was performed with fiducial markers placed on the forehead, anterior to the tragus of both ears, and on each of the mastoid tips. The patient's head was fixed in the three-point Mayfield head clamp. A registration was then completed, and accuracy was confirmed visually (Fig. 3a).

An incision was made through a natural skin crease in the right lateral neck extending up through the anterior neck dividing the lower lip in the plane of the patient's old incision from his previous operation (Fig. 3b). Next, the skin and muscle were elevated off the anterior portion of the mandible. With a micro-oscillating sagittal saw, an osteotomy was made in the right parasymphseal area just proximal to tooth #28, the right mandibular first molar, and small chisels were used to complete the osteotomy (Fig. 3c). Soft tissues of the floor of the mouth were divided using electrocautery, and care was taken to protect the lingual and hypoglossal nerves. The sublingual gland was encountered laterally and divided to avoid injury to the submandibular gland. Posterior to the glossopharyngeal tendon, the base of the tongue was freed from its attachments. Retractors were placed to displace the tongue laterally to provide access to the posterior wall of the pharynx (Fig. 3d).

At this point the SNN intraoperative navigational system was utilized, and the planned rostral and caudal ends of the decompression were formulated (Fig. 4). The posterior pharyngeal wall was incised using electrocautery, and the anterior portions of C1 to C4 were exposed. Self-retaining retractors were placed, aiding exposure. Using the wand system for guidance, C2–3 corpectomies were performed. This drilling process was carried back to the posterior longitudinal ligament (PLL). In addition, the arch of C1, the caudal end of the clivus, and a portion of the ventral C4 vertebral body were removed. The PLL was then opened at the C3–4 junction caudally, and the ligament was rostrally reflected off the ventral aspect of the dura up to the craniocervical junction and removed using rongeurs and curettes. At this point it was noted that the tip of the odontoid process was penetrating into the foramen magnum. Decompression was accomplished by drilling

Fig. 1 Sagittal T2-weighted MRI of the skull base and cervical spine illustrating stenosis at the craniocervical junction with increased intramedullary signal intensity (arrow)

Fig. 2 Radiographs of cervico-thoracic spine showing posterior fixation and fusion from occiput down to T9. **a** Lateral projection. **b** Anterior-posterior projection.

down the odontoid process with a high-speed diamond burr and by removing the cruciate, the transverse, and the apical ligaments. Operative ultrasound revealed complete decompression of the pons, the medulla, and the upper cervical spine.

Following the decompression, the posterior pharyngeal wall was closed with successive closure of the mylohyoid muscle, gingiva, labial mucosa, musculature of the lip, and moist vermilion of the lip. Craniofacial plates were used to reapproximate the mandible. The dry vermilion was then realigned, and the platysma and neck were closed (Fig. 3e).

Postoperative imaging revealed adequate decompression (Fig. 5). At 23 months' follow-up, the patient is able to stand and ambulate with the assistance of a walker and can speak clearly. There is no facial droop, no tongue deviation, and he possesses strong cough and gag reflexes. He complains of no malocclusion. Despite good bulbar function, however, he still uses his PEG tube for nutrition. His resting posture remains secure, and he complains of no neck pain.

Discussion

The TCR approach allows exposure to the middle and lateral compartments of the skull base and cranial nerves from nine through twelve. The exposure allows access

from the ipsilateral infratemporal fossa to the contralateral medial pterygoid plate, and may also be extended down to the C-7 vertebra, allowing control of nearby vasculature. The approach was originally elucidated by Biller et al. [3], and later by Krespi et al. [9] and Ammirati et al. [2]. Some have also advocated its use in chordomas of the clivus and upper cervical spine [5]. Here we have described its successful use in spinal and brain stem decompression in a patient with Klippel-Feil syndrome.

The use of frameless stereotaxy, while used most extensively in approaches to intracranial pathology, has a growing number of applications in spinal surgery, especially posterior surgeries including transarticular and pedicle screw placement in the cervical and lumbar spine [7, 10, 13]. Widespread use in the anterior spine is complicated by the relative lack of anatomical landmarks necessary for device registration. Nevertheless, novel approaches to this problem in the upper cervical spine have been described in the literature, including navigation-assisted transoral odontoidectomy in patients preoperatively fixed and scanned in a HALO device to permit immobility during registration [11]. Successful resection of a chordoma of the cranial base and upper spine was also achieved using frameless navigation in a patient with dislocation of the dens and medullary compression [12]. In addition, cadaveric studies have suggested that the accuracy of spinal

Fig. 3 Intraoperative approach to the anterior cervical spine. **a** Positioning of the patient showing 3-point skull fixation, placement of fiducial markers, and tracheostomy in place. **b** View of initial incision through lateral neck and lower lip at the site of previous incision. **c** Osteotomy with subsequent removal of right aspect of mandible. **d** Retractors placed to displace the tongue laterally to provide access to the posterior wall of the pharynx. **e** Postoperative photograph revealing multi-layered closure of soft tissues and reconstruction of mandible to achieve adequate cosmesis

instrumentation placement may be improved by the use of frameless stereotaxy [1, 4].

In this case, fixation was accomplished using a Mayfield head clamp, and anterior decompression was achieved with using the SSN navigation system. Despite obtaining adequate exposure to the anterior craniocervical junction using the TCR approach, it is sometimes difficult to accurately judge intraoperatively

Fig. 4 Illustration of the TCR approach with use of intraoperative navigation to plan extent of operative decompression

Fig. 5 Postoperative sagittal CT image reconstructed from axial images through the head and neck showing the extent of anterior and posterior decompression

how much bony decompression must be done. Given the adjacent brainstem and spinal cord, over-drilling in an attempt to achieve maximal bony decompression could have disastrous results. In this way, the use of such navigation guided the surgeons in obtaining adequate decompression without compromising neurological status. Such navigational accuracy may have been augmented in this case by cervical immobilization afforded by the patient's Klippel-Feil syndrome and previous posterior fusion.

Conclusion

Frameless stereotaxy, while most commonly applied to intracranial surgery, has seen an increasing number of applications in spinal surgery. Its application to the complex anatomy of the TCR approach may also provide benefit, especially in a patient with unusual anatomy, as described here. In this case, we illustrate the previously undescribed, successful use of frameless stereotaxy to the TCR approach in a patient with Klippel-Feil syndrome.

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