Diplopia Due to Skew Deviation Following Neurotologic Procedures

Maura K. Cosetti, MD1, Mohammad Fouladvand, MD2, J. Thomas Roland, Jr., MD1,3, and Anil K. Lalwani, MD1
Department of Otolaryngology1, Departments of Ophthalmology and Neurology, Department of Neurosurgery3, New York University Langone Medical Center, New York, NY, USA

ABSTRACT

Skew deviation refers to an acquired ocular hypertropia without comitant extra-ocular muscle abnormality that produces vertical diplopia. First associated with the vestibular system in 1979, Halmagyi et al. documented skew deviation following unilateral stapedectomy. The authors hypothesized that unilateral damage to a peripheral vestibular end organ, specifically the utricle, would lead to asymmetric imbalance in tonic vestibular tone thereby causing an ocular tilt reaction (OTR). Subsequent animal and human research into the OTR has confirmed that it can follow significant unilateral damage anyway along the vestibular pathway, including the labyrinth/vestibule. CN VIII, lateral vestibular nucleus, interstitial nucleus of Cajal, median longitudinal fasciculus, and cortex. The OTR or head- eye synkinesis involves 3 components: plantar reflex, vertical, ocular torsion or skew deviation and postural head tilt. Diplopia in the absence of cranial neuropathy following CPA procedures is rare. We present 4 cases of skew deviation following CPA surgery or labyrinthectomy.

INTRODUCTION

Skew deviation refers to an acquired ocular hypertropia without comitant extra-ocular muscle abnormality that produces vertical diplopia. First associated with the vestibular system in 1979, Halmagyi et al. documented skew deviation following unilateral stapedectomy. The authors hypothesized that unilateral damage to a peripheral vestibular end organ, specifically the utricle, would lead to asymmetric imbalance in tonic vestibular tone thereby causing an ocular tilt reaction (OTR). Subsequent animal and human research into the OTR has confirmed that it can follow significant unilateral damage anyway along the vestibular pathway, including the labyrinth/vestibule. CN VIII, lateral vestibular nucleus, interstitial nucleus of Cajal, median longitudinal fasciculus, and cortex. The OTR or head-eye synkinesis involves 3 components: plantar reflex, vertical, ocular torsion or skew deviation and postural head tilt. Diplopia in the absence of cranial neuropathy following CPA procedures is rare. We present 4 cases of skew deviation following CPA surgery or labyrinthectomy.

Table 1. Summary of patient characteristics and neuro-ophthalmologic findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, sex</th>
<th>Pathology</th>
<th>Tumor size: AP x transverse x cranio-caudal (mm)</th>
<th>Surgical approach</th>
<th>Pre-/post-op HL (dB)</th>
<th>Skew deviation: dipteric hypertropia (mm)**</th>
<th>Therapy</th>
<th>Diplopia resolution (weeks)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>55, F</td>
<td>Vestibular Schwannoma</td>
<td>22 x 24 x 32</td>
<td>Right TL (subtotal)</td>
<td>30/NoR</td>
<td>4</td>
<td>---</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>64, F</td>
<td>Petroclival meningioma</td>
<td>9 x 11 x 13</td>
<td>Left RS</td>
<td>15/25</td>
<td>10; 14, 8</td>
<td>14, 10</td>
<td>Prism therapy</td>
</tr>
<tr>
<td>3</td>
<td>59, F</td>
<td>Vestibular Schwannoma</td>
<td>11 x 4 x 4.3</td>
<td>Left RS</td>
<td>15/NoR</td>
<td>2; 3, 0</td>
<td>3, 2</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>50, M</td>
<td>Endolymphatic hydrops</td>
<td>n/a</td>
<td>Right labyrinthectomy</td>
<td>95/NoR</td>
<td>4; 8, 4</td>
<td>4, 6</td>
<td>none</td>
</tr>
</tbody>
</table>

*All patients experienced onset of diplopia immediately post-operatively
**The affected or hypertropic eye is contralateral to the side of surgery

METHODS

Retrospective review of patients with complaints of diplopia following CPA surgery or surgery for endolymphatic hydrops. All patients underwent neuro-ophthalmologic consultation and examination, including optokinetic testing, confrontational visual field assessment, color plate, pupillary reflex, slit lamp examination and Head Tilt Test. Diagnosis of skew deviation was made following neuro-ophthalmologic exam. If skew deviation persisted after 6 weeks, prism lenses were prescribed. Additional patient data including demographics, surgical approach, tumor size (where applicable), pre- and post-operative hearing level were recorded. Profound hearing loss was designated as NR (no measurable response to audiologic stimuli on the affected ear.)

DISCUSSION

The vestibular system maintains gaze stabilization during rotational (the vestibulo-ocular reflex and semicircular canals) and lateral (utricle and saccule) head movements (figure 2). Unilateral damage from endolymphatic hydrops surgery, including vestibular neurectomy or labyrinthectomy, can lead to skew deviation in some cases. A recent report found chemical labyrinthectomy with intra-lymphatic gentamicin also caused skew deviation, although onset was delayed rather than immediately following the procedure. Regardless of approach, CPA surgery also has the potential for vestibular nerve injury and resulting skew deviation. From this small case series it does not appear that tumor size predicts skew development, although a larger patient population is necessary. Patients 1-3 demonstrated only mild pre-operative HL. It is possible that patients with significant hearing preoperatively, a probable marker for residual vestibular function, may be specially at risk for developing skew deviation. As vestibular ablation occurs routinely with each of these procedures, skew deviation likely occurs more frequently than is currently diagnosed. Complaints of diplopia should prompt neuro-ophthalmologic consultation to reliably diagnose skew deviation and exclude cranial neuropathy. Patients can be reassured as spontaneous resolution typically occurs within 10 weeks.

REFERENCES