Optical Coherence Tomography for the Management of Fibrous Dysplasia with Suspected Optic Nerve Compression

Joshua Loewenstern1, Christopher Hernandez1, Carolyn Chadwick1, Amish Doshi, MD2, Rudrani Banik, MD3, Christopher Sarkiss, MD1, Joshua Bederson, MD1, and Raj K. Shrivastava, MD4

1Department of Neurosurgery, 2Radiology, and 4Ophthalmology, Icahn School of Medicine at Mount Sinai, New York, NY

BACKGROUND

- The skull base is the most common site of the craniofacial skeleton to be affected by fibrous dysplasia (FD).
- A significant proportion of such FD patients present with compression of the optic nerve, which can cause visual impairment, but most patients initially do not experience vision loss.
- Optical coherence tomography (OCT), which is a physiologic imaging modality widely used to assess the condition of the retinal nerve fiber layer (RNFL), has been useful in the monitoring of other compressive lesions on the optic nerve including pituitary tumors and craniopharyngioma, and may have a beneficial role in managing patients with FD involving the optic nerve.

METHODS

- 6 patients with suspected optic nerve compression who had received OCT imaging as part of their neuro-ophtalmic exam were reviewed over a 2-year period.
- Patients were evaluated for the presence of optic neuropathy grouped by either normal age-adjusted RNFL thickness (above 5th percentile) or abnormal (below 5th percentile) as measured by OCT imaging.
- Other visual exam measures included visual acuity, visual field deficits, color vision, and optic cup-to-disc ratio in addition to radiographic measures including optic canal size and presence of optic nerve encasement or compression on CT.

RESULTS

Two patients were found to have mild optic neuropathy in one eye each. The mean (± SD) RNFL thickness on OCT was 93.1 ± 12.0 µm, with 3 of 12 eyes falling below the age-adjusted 5th percentile.

Abnormal age-adjusted RNFL thickness was associated with the presence of optic neuropathy (ρ < 0.05, Table 1). Visual field defects, optic canal size, and optic nerve encasement differed by the presence of optic nerve compression on CT (ρ's < 0.05), but the occurrence of optic neuropathy did not differ by optic nerve compression (ρ = 0.45).

CONCLUSIONS

- Based on the case series of this rare condition, abnormal RNFL thickness as measured by OCT better predicted the presence of optic neuropathy than CT alone.
- OCT may therefore be a valuable imaging modality to monitor FD patients for development of optic neuropathy during periods of conservative "watchful waiting" in order to determine the best timing for intervention if necessary.

Table 1. Clinical and Radiographic Comparisons

<table>
<thead>
<tr>
<th>Clinical Measure</th>
<th>Age-adjusted OCT RNFL Thickness Percentile</th>
<th>Top 5% Below 5th (µm)</th>
<th>p-value</th>
<th>Optic Nerve Compression</th>
<th>Yes</th>
<th>No</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Top 5% Visual Field MD (µm)</td>
<td>7.7 (5.1)</td>
<td>13.2 (5.6)</td>
<td>0.14</td>
<td>0.047</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Optic Neuropathy</td>
<td>2 Yes, 1 No</td>
<td>2 Yes, 1 Partial, 0.003</td>
<td>0.046</td>
<td>0.046</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Radiographic Evaluation: Mean Optic Canal Cross-Area (mm²)</td>
<td>7.7 (5.1)</td>
<td>13.2 (5.6)</td>
<td>0.14</td>
<td>0.047</td>
<td>0.046</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Optic Nerve Encasement</td>
<td>2 Yes, 1 No</td>
<td>2 Yes, 1 Partial, 0.003</td>
<td>0.046</td>
<td>0.046</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Optic Nerve Compression</td>
<td>2 Yes, 1 No</td>
<td>2 Yes, 1 Partial, 0.003</td>
<td>0.046</td>
<td>0.046</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Figure 1. Representative retinal nerve fiber layer (RNFL) thickness measures from OCT

Figure 2. Intra-operative images of optic nerve decompression for FD

This work has been accepted for publication: Loewenstern, J., Hernandez, C.M., Chadwick, C., Doshi, A., Banik, R., Sarkiss, C.A., Bederson, J. and Shrivastava, R.K., 2018. Optical coherence tomography in the management of skull base fibrous dysplasia with optic nerve involvement. World Neurosurgery, 109, pp.e546-e553.