Grand Rounds

James Murphy, MD
SUNY Downstate, Dept of Ophthalmology
Thursday, November 14, 2013
Case

CC: EOM abnormalities (per neurology)
HPI: 58yo F h/o DLBCL s/p chemo 1-8/2013, PE consult from neurology service for lateral gaze weakness left eye noted on admission for leg weakness and LBP/leg pain. Patient reported pain and diplopia in left gaze but not in primary.

POH/gtts: denies past trauma, ocular surgery, strabismus
PMH/meds: DLBCL diagnosed 1/2013 s/p R-CHOP 12 cycles terminating 8/2013, PE 2/2013 on Lovenox, L-spinal stenosis
NKDA
FH: migraines-sister, mother
SH: (-) tobacco, EtOH, illicits, IVDA
ROS: (+) chronic leg weakness, numbness, pain, LBP; (-) fevers, night sweats, weight loss
Case

dVAsc: 20/25 ou
Pupils: 4-2 errl ou (-) RAPD
EOM: OD full; OS -1 infra/supra-duction, -4 ABduction, full AD
CVF: FTFC ou
Tapp: 14/14 @ 14:00

HEENT: (-) bony irregularity, lymphadenopathy, swelling. (+) fullness of orbit esp. laterally L-side
Hertel: 19/17 @ 115mm

SLE
LLA: wnl B/L
CS: w/q ou
K: clear ou
AC: d/q ou
IP: flat, rrl ou
Lens: tr ns ou

DFE with scleral depression: unremarkable

Patient Care, Professionalism, Interpersonal Communication, Practice-Based Learning & Improvement
What next?
Systems-Based Learning, Practice-Based Learning & Improvement
Differential?
Differential

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thyroid orbitopathy</td>
<td>50</td>
</tr>
<tr>
<td>Inflammatory lesion</td>
<td>11</td>
</tr>
<tr>
<td>Cystic lesion</td>
<td>10</td>
</tr>
<tr>
<td>Lymphoproliferative</td>
<td>5</td>
</tr>
<tr>
<td>Vascular neoplasm</td>
<td>4</td>
</tr>
<tr>
<td>Secondary tumors</td>
<td>4</td>
</tr>
<tr>
<td>Mesenchymal tumor</td>
<td>4</td>
</tr>
<tr>
<td>Optic Nerve tumor</td>
<td>3</td>
</tr>
<tr>
<td>Lacrimal gland lesion</td>
<td>2</td>
</tr>
<tr>
<td>Vascular anomalies</td>
<td>1</td>
</tr>
<tr>
<td>Other</td>
<td>5</td>
</tr>
</tbody>
</table>

- IOI, Wegener, sarcoidosis
- Primary
- Lymphoma
- Lacrimal gland tumor
- Rhabdomyosarcoma
- Extension from globe
- Melanoma
- Uveal mets
- Glioma, meningioma

Metastatic orbital disease
Overview

- Represent 2-11% of all orbital tumors
- 30-60% orbital mets establishes diagnosis before primary tumor. Why?
- Mets reach the orbit via hematogenous spread
- Orbital mets << uveal mets (1:8)
- Bilateral only 4% of the time (c/c: uveal mets)
- Infrequently involve the EOMs (ex: melanoma)
- Common: proptosis, abaxial globe displacement, ptosis, diplopia, pain, chemosis, vision loss
- Most common location: superotemporal orbit

<table>
<thead>
<tr>
<th>Primary Orbital Tumor</th>
<th>Metastatic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proptosis, vision loss</td>
<td>Diplopia, pain</td>
</tr>
</tbody>
</table>

Practice-Based Learning & Improvement
Etiology

Breast
Prostate
GI (adenoCa)
Lung (bronchogenic>SCLC)
Other: RCC, thyroid Ca, neuroblastoma, Ewing sarcoma, Wilms tumor

<table>
<thead>
<tr>
<th>Origin</th>
<th>Per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breast</td>
<td>53</td>
</tr>
<tr>
<td>Prostate</td>
<td>11</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>11</td>
</tr>
<tr>
<td>Lung</td>
<td>4</td>
</tr>
<tr>
<td>Sarcomas and other</td>
<td>21</td>
</tr>
</tbody>
</table>

# Epidemiology

<table>
<thead>
<tr>
<th>Diagnostic Group</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Childhood and Adolescence (0–20 years)</td>
</tr>
<tr>
<td>Adenoid cystic carcinoma of lacrimal gland</td>
<td>18</td>
</tr>
<tr>
<td>Capillary hemangioma</td>
<td>100</td>
</tr>
<tr>
<td>Cavernous hemangioma</td>
<td>10</td>
</tr>
<tr>
<td>Cystic lesions</td>
<td>77</td>
</tr>
<tr>
<td>Fibrous histiocytoma</td>
<td>25</td>
</tr>
<tr>
<td>Infectious processes</td>
<td>35</td>
</tr>
<tr>
<td>Inflammatory lesions</td>
<td>12</td>
</tr>
<tr>
<td>Lymphangiomas</td>
<td>6</td>
</tr>
<tr>
<td><strong>Lymphoproliferative diseases</strong></td>
<td><strong>1</strong></td>
</tr>
<tr>
<td>Optic nerve glioma</td>
<td>5</td>
</tr>
<tr>
<td>Optic nerve meningioma</td>
<td>4</td>
</tr>
<tr>
<td>Pleomorphic adenoma of lacrimal gland</td>
<td>0</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>98</td>
</tr>
<tr>
<td>Secondary and metastatic malignancies</td>
<td>1</td>
</tr>
<tr>
<td>Thyroid orbitopathy</td>
<td>4</td>
</tr>
<tr>
<td>Trauma</td>
<td>7</td>
</tr>
</tbody>
</table>

Diagnostics

- Comprehensive Eye Exam
- Forced-ductions
- Imaging
  - MRI
    - T1: isointense to EOMs
    - T2: hyperintense to EOMs, hypo to orbital fat
    - T2+Gad: variable enhancement
  - CT: similar to MRI, less resolution
- Biopsy
  - Centroblastic (>80%)
  - Immunoblastic
  - Anaplastic
Management

- Comprehensive H&P
- Advanced Imaging
- **When is surgery indicated?**
  - Biopsy for diagnosis
  - Anatomically malignant/threatening globe/orbital contents
Surgical Approaches
Our Patient: recurrent orbital DLBCL

- DLBCL is the most common adult form of NHL
- Most are Bcl-6 positive
- Prognosis: 5-year survival >50%
- LP with cytology confirmed recurrence of DLBCL
- Full body imaging revealed no other foci of recurrence
- Currently undergoing radiation to cavernous sinus and left orbital apex co-managed by RadOnc and Oncology services, chemo planned to follow
- Patient recovering as expected from surgery, EOM deficits resolved with the exception of abduction, which has shown improvement

Patient Care, Professionalism, Interpersonal Communication, Practice-Based Learning & Improvement
The case presented today offers an excellent learning case in that it affords and demands a comprehensive review of orbital tumors, their presentation and a review of their wide array of etiologies. The examination, patient communication, and clinical decision-making made for an educational and valuable experience. The case involved communication of bad news with the patient, which is an experience any clinician should review and practice. The case also demanded interdisciplinary communication with oncology, ENT, neurology, neurosurgery and radiation oncology, which at times was challenging but rewarding in the end, as the patient received appropriate care and experienced empathetic interactions with all staff involved.
ACGME Core Competencies

- **Professionalism**: compassionate and ethical care was delivered at all times. The patient’s needs were placed above all others.
- **Patient Care**: appropriate, effective, and state-of-the-art care was delivered at every clinical and surgical visit. Promotion of patient health and autonomy was maintained throughout the course of treatment and evaluation.
- **Medical Knowledge**: up-to-date medical knowledge was established through review of the most current medical literature and practice of the most recent, safe and effective surgical techniques.
- **Interpersonal and Communication Skills**: effective communication was maintained at all times. A detailed discussion was held with the patient regarding risks, benefits and alternatives of all medical and surgical options and an informed decision and consent were made based on thorough consideration of this information.
- **Practice-Based Learning and Improvement**: a systematic review of the patient’s progress at each visit was carried out and comparison with the published body of literature and norms were reviewed throughout the course of management.
- **Systems-Based Practice**: the patient was approached and evaluated with respect to overall well-being. Care was taken to ensure that the patient was actively monitoring and treating her other co-morbidities with proper and appropriate physician supervision.

Professionalism
References

- BCSC 4: Ophthalmic Pathology and Intraocular Tumors
- BCSC 7: Orbits, Eyelid and Lacrimal System, 2011
Thank you to:

- Dr. Shinder
- Dr. Burstein
- Dr. Farber
Questions?