Primary CNS Melanoma

Kevin Leikert
Shravani Mikkilineni
Barry Skarf

Follow this and additional works at: https://scholarlycommons.henryford.com/merf2020caserpt
Primary CNS Melanoma

Kevin Leikert DO, Shravani Mikkilineni MD, Barry Skarf MD PhD

Henry Ford Health System, Detroit, Michigan

Patient Presentation

- 49-year-old female referred for bilateral disc edema
- Bilateral disc edema noted by an optometrist in April of 2018, associated with a headache
- Referred to an ophthalmologist
- Has gained 20-30 lbs. in the last 4 years
- Saw an outside ophthalmologist, visual field showed some constriction in the right eye
- Did not start diams at that time, referred to neurology
- Neurology ordered an MRI and LP:
  - MRI/ MRV – Mild bilateral disc edema, no intracranial mass, no venous thrombosis
  - LP – opening pressure of 27
  - Started her on Diamox 1000 mg BID, then decreased to 1000 mg AM and 500 mg PM due to not tolerating the diamsx
- Returned to optometrist 2 weeks later with continued decreased visual acuity
- Discussed with neurology and increased diams back to 1000 mg BID
- Referred to neuro-ophthalmology

Diagnostic Considerations

- Elevated disc margins
  - Pseudopapilledema
  - Disc Drusen
  - Malignant hypertension
- Hyperviscosity syndromes
- Hypotension/blood loss
- Early in the course of toxic optic neuropathies

Elevated Intracranial Pressure

- Idiopathic intracranial hypertension
- Intracranial mass lesions
- Obstruction of venous outflow
- Venous sinus thrombosis
- Jugular vein compression
- Obstructive hydrocephalus
- Decreased CSF absorption
- Arachnoid granulation adhesions after bacterial or other infectious meningitis
- Subarachnoid hemorrhage
- Increased cerebrospinal fluid (CSF) production
- Choroidal plexus papilloma
- Malignant systemic hypertension

Workup for Bilateral Optic Disc Edema

- Full ophthalmologic exam including:1
  - Visual acuity
  - Visual field testing
  - Evaluation for afferent pupillary defect
  - Color vision testing
  - Dilated fundoscopic exam
- Optical coherence tomography (OCT) to quantify the elevation of the retinal nerve fiber layer
- MRI/ MRV and lumbar puncture6

Initial Imaging

Follow Up Imaging

Figure 1: (1) Optic disc pallor with surrounding watershed area. (2) Optic disc edema with hyperemia and vessel-obstruction (arrows). (3) OCT nerve showing diffuse optic atrophy in the right eye and elevated retinal nerve fiber layer in the left eye. (4) Ganglion cell layer section of OCT showing significant atrophy in the right eye and full ganglion cell layer in the left eye. (5) Humphrey visual field of the left eye showing a full visual field. (6) Humphrey visual field of the right eye showing diffuse constriction with a small infra-temporal island remaining

Follow Up Visits

<table>
<thead>
<tr>
<th>Date</th>
<th>Interval History</th>
<th>Exam</th>
<th>Testing</th>
<th>Plan</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/14/19</td>
<td>No changes in vision</td>
<td>Last 5 lbs. Taking Diamox 1000 mg BID</td>
<td>3+ APD OD Full color plates HVF unchanged Optic atrophy OD with possible refractive h. Persistent edema OD</td>
<td>Stable RNFL and GCL on OCT Decrease Diamox to 1000 mg AM and 500 mg PM</td>
</tr>
<tr>
<td>3/26/19</td>
<td>No changes in vision</td>
<td>Headsaches have improved</td>
<td>Last 5 lbs. Taking Diamox 1000 mg BID</td>
<td>3+ APD OD Full color plates HVF unchanged Optic atrophy OD Persistent edema OD</td>
</tr>
<tr>
<td>6/20/19</td>
<td>Nausea and vomiting</td>
<td>Gait abnormalities</td>
<td>Taking Diamox 1000 mg BID</td>
<td>3+ APD OD Full color plates HVF early depression in the left eye Optic atrophy OD Persistent edema OD</td>
</tr>
</tbody>
</table>

Subsequent Course

- Laminectomy from C7 to T3 was performed on 07/29/2019
- Pathology confirmed metastatic melanoma
- Gait improved after debulking of tumor
- Full dermologic exam revealed no other sites of melanoma
- Supports presumptive diagnosis of primary CNS melanoma
- Started on systemic chemotherapy per medical oncology
- Received one infusion
- Had a rapid decline and died on 8/25/19

Primary CNS Melanoma

- Melanocytes exist in the uvea, cerebral parenchyma, leptomeninges, mucous membranes, and skin4
- Uncommon and constitute approximately 1% of all melanoma cases and 0.07% of all brain tumors3
- Primary cerebral melanomas often develop in patients under 50 years of age and uncommonly metastasize to systemic organs
- Hyperintense on T1 weighted images and hypointense on T2 weighted images due to presence of melanin2
- Once the cancer spreads to leptomeninges, the overall median survival is generally only 10 weeks9

Bibliography