Presentation and Progression of Papilledema in Venous Sinus Thrombosis

Katy C. Liu, 1 M. Tariq Bhatti, 1,2,3 Mays A. El-Dairi1

Departments of Ophthalmology, 1 Neurology2 and Neurosurgery, 3 Duke Eye Center and Duke University Medical Center, Durham, NC

Purpose

Venous sinus thrombosis (VST) is a rare cerebrovascular disease that generally affects younger patients. In adults, 32-48% of patients with VST have been reported to have papilledema. 1,2 Papilledema can result in potentially devastating vision loss. Thus, the natural history of papilledema in VST and its response to treatment are important for ophthalmologists, neurologists, and neurosurgeons to recognize in order to maximize visual outcomes. We describe the presentation and progression of papilledema in a retrospective chart review of patients with VST who underwent serial eye examinations. Current guidelines for VST management broadly recommend monitoring for progressive visual loss, but with little specifics. 3

Methods

Approval for this study was obtained from the institutional review boards at the Duke University Medical Center. A retrospective chart review was performed on all patients who presented to the Neuro-Ophthalmology clinic from January 1, 2008 through December 31, 2015. Patients were included in the study if they had a diagnosis of VST confirmed by neuroimaging (MR or CT venography) and received serial ophthalmic examinations including dilated fundoscopy. Any predisposing factors for VST were recorded. In total, 7 adult and 3 pediatric patients (age range: 9-58 years) were identified. Papilledema was graded according to the Frisen scale (0 to 5). Outcome measures included: time from diagnosis to development of papilledema; progression of papilledema (Frisen grade and time frame); time to resolution of papilledema; and final visual outcome (best-corrected visual acuity and visual fields).

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Figure 1: MR Venoography of VST (coronal and axial sections shown). A) Patient 1 (top): Thrombosis of superior sagittal sinus and right transverse sinus, sigmoid sinus with extension to the right jugular bulb and right jugular vein. B) Patient 2 (bottom): Thrombosis of right transverse sinus, sigmoid sinus, and proximal jugular vein.

Figure 2: Fundus photos of progression of papilledema and response to treatment. Patient 1 (left): A) week 1 after VST diagnosis, grade 2 OU; B) week 2: grade 3-4 OD, grade 4 OS (red line: LP in week 2); C) week 3: grade 3 OD, grade 2 OS; D) 5 months; E) week 1: grade 1 OD; F) week 2: grade 5 OU; G) week 3: grade 5 OU (red line: ONSF OS in week 4); H) week 5: grade 4 OD, grade 2 OS (sp ONSF OS).

Results

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age/ Gender</th>
<th>Clinical presentation</th>
<th>Preexisting factors</th>
<th>Initial papilledema grade</th>
<th>Time VST diagnosis to papilledema</th>
<th>Progression of papilledema over time</th>
<th>Treatment</th>
<th>Time from treatment to final visual outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>43 F</td>
<td>N/A</td>
<td>N/A</td>
<td>n/a</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>2</td>
<td>40 F</td>
<td>Right transverse sinus, sigmoid sinus, and proximal right jugular vein</td>
<td>N/A</td>
<td>3</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>3</td>
<td>52 M</td>
<td>N/A</td>
<td>N/A</td>
<td>4</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>4</td>
<td>29 F</td>
<td>N/A</td>
<td>N/A</td>
<td>2</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>5</td>
<td>26 M</td>
<td>N/A</td>
<td>N/A</td>
<td>4</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>6</td>
<td>35 M</td>
<td>N/A</td>
<td>N/A</td>
<td>3</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>7</td>
<td>35 M</td>
<td>N/A</td>
<td>N/A</td>
<td>2</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>8</td>
<td>56 M</td>
<td>N/A</td>
<td>N/A</td>
<td>4</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>9</td>
<td>42 M</td>
<td>N/A</td>
<td>N/A</td>
<td>5</td>
<td>0 days</td>
<td>1. LP alone 10 months</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>

• Bilateral papilledema grade ≥3 was present in 9 of 10 patients. One patient had grade 1 papilledema.

• The time from diagnosis of VST to papilledema ranged from the same day to three months. In two patients (2 & 3), papilledema was not present on initial examination. In other cases, a baseline fundoscopic exam was not performed; papilledema was noted on outpatient follow-up.

• Two patients (1 & 2) showed worsening of papilledema grade prior to intervention. Worsening of papilledema was detected at 1-week follow-up in both patients, and were examined weekly until response to treatment.

• Three patients (5, 9, 10), were initially diagnosed with papilledema in the setting of idiopathic intracranial hypertension but were subsequently found to have VST.

• Four patients required surgical intervention: 2 patients (2 & 7) had optic nerve sheath fenestration (ONSF), 1 patient (9) had a ventriculoperitoneal shunt placed, and 1 patient (7) underwent ONSF followed by a lumboperitoneal shunt.

• Time to resolution of papilledema ranged from 3 weeks to 1 year.

• Final visual acuity ranged from 20/20 to light perception (LP) with 5 of 10 patients with visual impairment.

Conclusions

• Serial ophthalmic evaluation is important in all patients diagnosed with VST regardless of the absence of visual symptoms; because VST can result in significant visual impairment.

• Papilledema has varying presentations in VST:
  • Papilledema may be present at time of presentation
  • Papilledema may not be present at time of presentation by may develop subsequently
  • Papilledema may worsen after presentation

• Based on the results of this study we recommend:
  • Baseline ophthalmic examination at time of diagnosis
  • Follow-up examinations weekly for 1 to 2 weeks

• Response to treatment should be followed with ophthalmic examination after medical or surgical intervention(s).

References