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## Abstract

- A 12-year-old boy presented with 3 weeks of progressive bilateral visual disturbances and examination revealed severe bilateral disc edema
- Medical history included group A streptococcal bacteremia and left pseudomonas otomastoiditis complicated by cerebral venous sinus thrombosis (CVST) involving left transverse/sigmoid sinuses and internal jugular vein
- Work-up confirmed markedly elevated intracranial pressure (ICP) and persistent venous sinus thrombosis
- Despite antibiotics, anticoagulation, acetazolamide, steroids, and venous sinus angioplasty, progressive vision loss required ventriculoperitoneal (VP) shunt placement
- Case highlights severe vision-threatening complications of pediatric CVST and need for aggressive ICP management and multidisciplinary care

## Case Presentation

### Presentation:

- A 12-year-old male presented to the ED with three weeks of bilateral visual disturbances including partial field blackouts and central scotoma in the left eye.
- Referred by outside optometry due to findings of bilateral optic disc edema

### Pertinent medical history:

- Recent hospitalization for Group-A streptococcus bacteremia and left-sided pseudomonas otomastoiditis complicated by thrombophlebitis of the left transverse/sigmoid sinus and internal jugular vein
- Underwent mastoidectomy, myringotomy and treatment with IV antibiotics and anticoagulation
- Visual symptoms started 2 months after discharge

### Ophthalmic exam:

	OD	OS
Near Visual Acuity	20/200 PH 20/30	20/400 PH 20/50
Intraocular Pressure	18	18
Pupils	Pharmacologically dilated	Pharmacologically dilated
Colors	Control + 5/7	Control+ 4.5/7
Extraocular Motility	Full	Adducted in primary -2 abduction
Visual Fields	Full	Full (reports central scotoma)

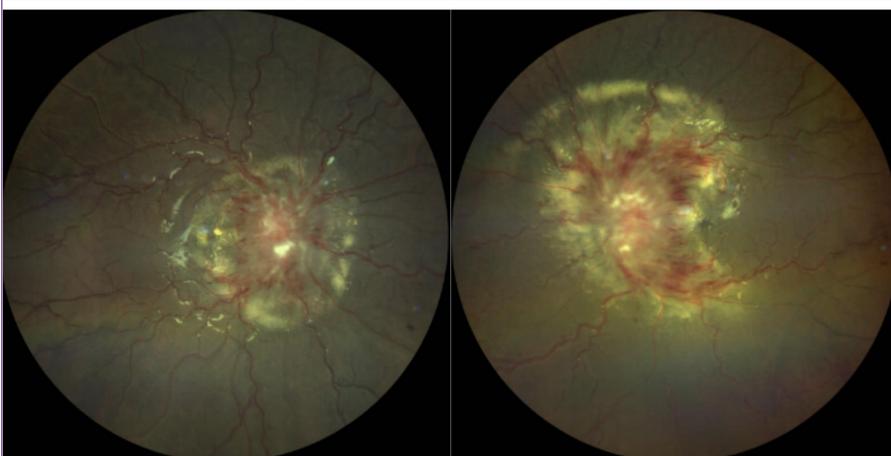


Figure 1: Fundus photographs of both eyes revealed severe bilateral disc edema with hemorrhages, vessel obscurations, peripapillary exudates and a serous retinal detachment in the left eye.

## Work-Up and Management

### Labs:

- Infectious/inflammatory serum studies within normal limits

### Lumbar puncture (LP):

- Opening pressure >55cm H<sub>2</sub>O
- CSF studies within normal limits (NMO/MOG negative)

### MRI Brain with and without contrast (Figure 2):

- Thickening of the optic nerve sheath complexes with bulging bilateral optic discs consistent with bilateral papilledema

### MRV Brain with contrast (Figure 3):

- Opacification within the left transverse and sigmoid dural venous sinuses and left proximal internal jugular vein consistent with persistent venous thrombosis

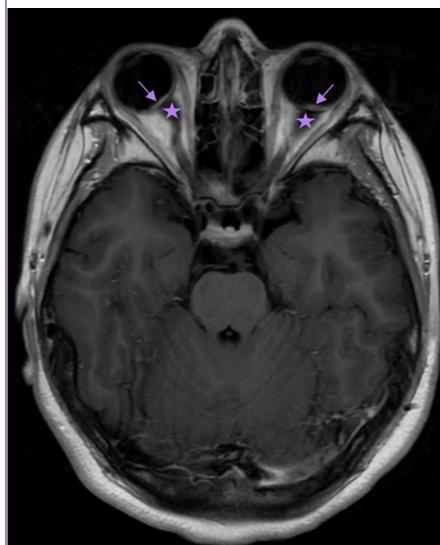


Figure 2

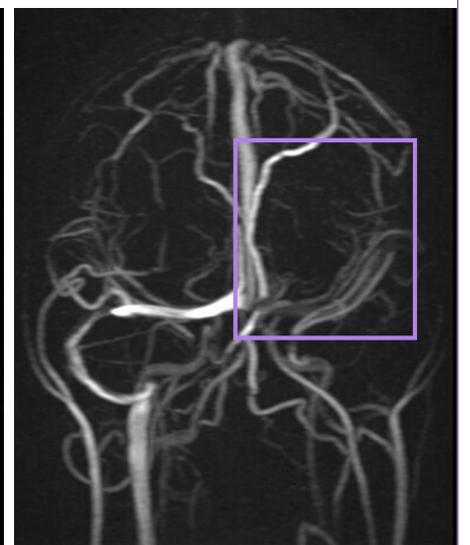


Figure 3

### Treatment Course:

- Started on acetazolamide and steroids for severe papilledema
- 2 months later, underwent repeat LP and venous sinus angioplasty due to concern for progression
- 3 months later, repeat angiogram demonstrated persistently elevated venous sinus pressure, underwent placement of VP shunt
- Developed optic nerve atrophy secondary to severe disc edema despite aggressive interventions
- Remains stable on acetazolamide therapy

## Discussion

- CVST is a serious complication of mastoiditis, presenting with subtle findings often masked by infection, making timely diagnosis critical<sup>1</sup>
- Papilledema occurs in 90% of pediatric CVST cases, with over half experiencing permanent optic nerve damage and vision loss<sup>2</sup>
- Aggressive ICP control is needed to minimize permanent vision loss
- Standardized treatment protocols remain undefined<sup>3</sup>
- Anticoagulation remains a mainstay in CVST medical treatment
- Papilledema treatment approach is similar to that of fulminant or malignant idiopathic intracranial hypertension (IIH):
  - Acetazolamide (alternatively, topiramate or furosemide)
  - IV and oral steroids
  - Temporizing CSF drainages (lumbar drain, serial large volume LPs)
  - CSF diversion procedures (venous sinus stenting/angioplasty, VP shunt)<sup>4,5</sup>

## Conclusions

- Pediatric CVST can present with severe papilledema and rapid vision loss, particularly when associated with otomastoiditis
- Early recognition and aggressive ICP management are critical to prevent permanent optic nerve damage
- Venous sinus angioplasty may provide temporary relief, but many patients require definitive CSF diversion with VP shunting
- Multidisciplinary care and early neuro-ophthalmologic involvement are essential to optimize visual and neurologic outcomes

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