A case of unusual blindness

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The Case

- 35 year old male who presented with
  - Bilateral complete vision loss - 6 day duration.
  - It started as blurry vision while vacationing in Antigua and progressed to complete vision loss in 24 hrs.
History (Contd.)

- **Review of Systems**
  - Positive for *pressure like sensation behind the eyes*
  - Negative for diplopia, discharge, pain.
- **Relevant History**
  - Denies any history of
    - Recent ear infections
    - Treatment for acne
    - Vitamin A supplement use
    - Lead exposure
    - Seizure disorder
    - Steroid use
History (Contd.)

- **Past Medical History**
  - ADHD
- **Allergies**
  - None
- **Medications**
  - Adderall
- **Family History**
  - Non contributory
- **Social History**
Physical Examination

- **Vitals:** HR 56, BP 137/83, RR 20, PO2 97% on RA, T 98.6
- **General:** Anxious, overweight young male
- **Respiratory:** Clear to auscultation
- **Cardiovascular:** RRR. S1 and S2 present.
- **Ophthalmic:**
  - No light perception
  - IOP and Anterior segment - WNL
  - Pupils round, equal but **non reactive to light**
  - **Fundoscopy:** Bilateral papilledema with retinal hemorrhage and venous tortuosity
- **Neurological:** Motor, Sensory and Cerebellar - WNL.
**Labs**

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<td>142</td>
<td>108</td>
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- AST - 18 U/L, ALT - 24 U/L, Alk Phos - 101 U/L
- ESR - 16 mm/hr
- CRP - 1.8 mg/dl
- ACE level - 24 U/L
Imaging

- MRI, MRA and MRV were within normal limits.
Labs (Contd.)

**CSF Analysis**

- **Opening Pressure** - 330 mm of H2O
- Appearance - clear and colorless
- Protein - 34 mg/dl
- Glucose - 64 mg/dl
- Gram stain - Negative
- Cytology - 2 mononuclear WBC
Negative CSF Workup

- Bacterial culture
- Viral culture
- Fungal culture
- VDRL
- HSV1
- HSV 2
- West Nile Virus IgM
- Varicella Zoster PCR
- Toxoplasma PCR
- Cryptococcal antigen
- B. burgdorferi Ig
Based on the clinical picture and investigations secondary causes of raised ICP like intracranial mass lesions (tumor, abscess), choroid plexus papilloma, arachnoid granulation adhesions after subarachnoid hemorrhage or bacterial meningitis, obstructive hydrocephalus, venous sinus thrombosis were ruled out.

Other differentials like glaucoma, optic neuritis, multiple sclerosis were ruled out.
Pseudotumor Cerebri
Discussion

- **Pseudotumor cerebri (PTC)/ Idiopathic intracranial hypertension (IIH)/ Benign Intracranial hypertension**, is a disorder of unknown etiology, first described by Quincke in 1897. (1)

- It is characterized by *symptoms and signs of increased intracranial pressure*, elevated intracranial pressure with normal CSF composition and no identifiable etiology of neuroradiologic studies. (2)

- The incidence of IIH ranges from 0.9-1 per 100,000 in the general population (3). *More than 90%* of PTC cases are seen in *obese women of child bearing age* (4).

- Pathogenesis is *unclear*. 
Clinical Presentation

**Symptoms**
- Headache (non specific and variable) (8),
- Horizontal diplopia (CN VI palsy),
- Pulsatile tinnitus
- Vision loss (acute or progressive), transient visual obscuations.

**Physical exam**
- Papilledema
- Visual field defects (usually peripheral)
- Abducens palsy (unilateral or bilateral).
Diagnosis

- MRI with/without contrast with post contrast MR venography is the imaging modality of choice.
  - Flattening of the posterior sclera is the most reliable indicator on MRI (9-10)
    - Specificity - 100%
    - Sensitivity - 43.5%
  - Lumbar puncture
    - Opening pressure of >250 mm of H2O.
  - CSF analysis - Normal
Associations...

- **Medications and exogenous substances**
  - Growth hormone
  - Tetracycline
  - Retinoids
  - Nalidixic acid
  - Lithium,
  - Nitrofurantoin
  - Thyroid replacement
  - Corticosteroid withdrawal.

- **Associated systemic illnesses**
  - Addison’s disease
  - Hypoparathyroidism
  - Anemia
  - Polycystic Ovarian Syndrome
  - Systemic Lupus Erythematosus
  - Behcet’s syndrome
• Our patient did not have any associated systemic illnesses.
• Only medication he was taking was **Adderall** for ADHD.
Adderall

- Adderall (Dextroamphetamine and amphetamine) is a non catecholamine, sympathomimetic amine that promotes release of catecholamines from presynaptic nerve terminals.
- Adderall XR was approved to treat ADHD in adults in 2004.
Reports

- Based on reports from FDA, since 2003 there were 6 cases of people using Adderall who developed pseudotumor cerebri
• **Females** - 4
• **Males** - 2
• **Age range**
  - 10-19 yrs - 4 cases
  - 30-39 yrs - 2 cases
Treatment

- **Treatment goals**
  - Alleviation of symptoms
  - Preservation of vision

- **Initial Treatment**
  - Stop potential agents that can cause IIH.
  - Acetazolamide
  - Lasix and topiramate
  - Short term steroids/serial lumbar punctures
  - Bridge to definitive surgical therapy in patients with rapidly progressive symptoms.
Treatment (Contd.)

- **Surgical Treatment**
  - *Optic nerve sheath fenestration (ONSF)*
    - Less severe side effects and significantly better outcomes (11).
    - Benefits may not be long lasting (12).
  - *CSF shunts (Lumboperitoneal/Ventricular)*
    - Failure of maximal medical therapy/ONSF
    - Intractable headaches
    - Available expertise
    - Patient preference.

- **Newer modalities**
  - *Intracranial dural venous stenting.*
Prognosis

- The prognosis for visual outcome in IIH is generally good. Prompt intervention results in a favorable outcome (13).
- People at higher risk of permanent visual loss:
  - Males
  - Younger age
  - Black race
  - Systemic hypertension
  - Anemia
  - Significant vision loss at presentation
  - Obesity
  - Higher opening pressures

- Recurrence of symptoms occur in 8-38% of patients
Back to the Patient...

- **Adderall** was stopped.
- With lumbar puncture, pressure sensation behind his eyes was relieved and his vision improved slightly.
- He was started on **acetazolamide** 250 q6h and also on steroids initially (for suspicion of multiple sclerosis).
- Steroids were later tapered off and discontinued after negative workup for MS.
- An **LP shunt** was placed due to the severity of visual loss with continuous, gradual improvement in his vision.
- Post discharge his **vision improved dramatically with almost complete resolution** of his symptoms at 1 month follow up.
References

Thank You