Etiology of Acute Unilateral Enlarged Blind-spot: Idiopathic or Hidden Culprit?
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Abstract:
A 26 year-old woman sees a large spot in her left eye, correlating with an enlarged blind spot on examination. Work-up is underway to rule-out a treatable etiology versus acute idiopathic blind spot enlargement (AIBSE) syndrome.

I. Case History
A 26 year-old Caucasian woman presented with a chief complaint of a black spot in the vision of her left eye, which had been present for three weeks. The onset was acute and not accompanied by pain, headache, or any other symptom. Her systemic history is remarkable for cervical cancer for which she underwent Loop Electrosurgical Excision Procedure (LEEP) 6 years prior. Medications include only an Ortho Evra patch. Social history includes smoking for 10 years and social alcohol consumption. Best-corrected visual acuity was 20/20 OD and 20/20 OS. Color vision testing demonstrated that the patient was able to identify 14/14 Ishihara plates correctly with each eye. Pupil testing revealed a 0.3 log unit relative afferent pupillary defect in the left eye. Confrontation fields were full OD and demonstrated a significantly enlarged blind-spot OS. Humphrey visual field testing confirmed the enlarged blind-spot, which was approximately 10 x 15 degrees. Ocular motility testing demonstrated normal ductions, versions, and saccades. Slit lamp examination revealed healthy anterior segment structures bilaterally. Intraocular pressures using Goldman applation tonometry were 12 mmHg OD and 16 mmHg OS. Blood pressure was 120/90 right arm sitting. Dilated fundus examination revealed hypoplastic, anomalous optic nerves (cupping of 0.1/0.1 OU). The neuroretinal rim was pink with no evidence of pallor in either eye. The maculae, retinal vasculature and peripheral retinal evaluation were unremarkable in each eye.

The patient was referred for work-up, including laboratory testing and neuro imaging. The MRI showed no structural abnormalities or abnormal enhancement in the brain or orbits. Laboratory testing was remarkable for an elevated white blood cell count, ESR, and C-reactive protein, as well as an ANA with a nucleolar pattern and a borderline 1:40 titer.
The patient returned in follow-up three weeks later, with no new symptoms. All aspects of her clinical appearance remained stable. Additional lab work was ordered to further investigate the reason for the elevated inflammatory markers and elevated white blood cell count, as well as to rule out an auto-immune process. The patient will continue to be closely monitored to determine if any treatment is warranted.

II. Pertinent findings
A. Radiology Studies
• MRI of brain and orbits with and without contrast.

B. Laboratory studies ordered:
• CBC with differential
• Platelet count
• ESR (Westergren)
• C-reactive protein
• Lyme titer
• RPR
• FTA-ABS
• ACE
• ANA
• p-ANCA
• c-ANCA

C. Additional lab work ordered
• ENA
• Anti-ds DNA
• SSA/SSB
• Rheumatoid factor
• Repeat CBC, ESR, C-reactive protein

D. Other tests to consider:
  • ERG
  • IVFA

III. Differential diagnosis of enlarged blind-spot
  • Acute Idiopathic Blind-spot Enlargement Syndrome
  • Multiple Evanescent White Dot syndrome (MEWDS)
  • Optic neuritis (papillitis) secondary to:
    - Multiple sclerosis
    - Systemic lupus erythematosus
    - Lyme disease
    - Sarcoidosis
    - Syphilis
    - Toxic reactions (i.e. to tobacco, methanol, quinine, arsenic, salicylates, lead)
  • Compressive lesions (i.e. retrobulbar mass)
  • Papilledema
  • Pseudotumor Cerebri
  • ONH drusen

IV. Diagnosis and discussion
The etiology of the patient’s enlarged blind-spot continues to remain elusive. This may be a presentation of acute idiopathic blind-spot enlargement (AIBSE). However, due to the laboratory test results, more testing is underway to rule out any other etiology

V. Treatment, management
No treatment at this time. The patient will continue to be monitored closely to determine if any treatable etiology can be uncovered. If the acute idiopathic blind-spot enlargement is confirmed, the treatment will consist of close monitoring as spontaneous resolution may occur.

VI. Conclusion/Clinical Pearls
Volpe et al. defined acute idiopathic blind-spot enlargement (AIBSE) as an “absolute symptomatic enlargement of the blind spot without commensurate swelling of the optic nerve head occurring in conjunction with presumed disease of the optic nerve and peripapillary retina”. Acute idiopathic blind-spot enlargement (AIBSE) is considered a diagnosis of exclusion and its cause remains unknown.

VI. Bibliography/literature review