Abstract Title: Ocular complications associated with idiopathic hypertrophic cranial pachymeningitis (IHCP) and subsequent vision rehabilitation

Abstract:
A patient presents with near total visual and hearing impairment secondary to IHCP. This rare case is unique for IHCP because of the severity of dual sensory loss and highlights challenges to rehabilitation.

I. Case history
  • Demographics:
    1. 69 year old white male veteran
  • Chief complaint:
    1. Wants to live independently; h/o total visual impairment right eye with near total vision loss in the left eye. Profound hearing loss right ear. Severe hearing loss left ear.
  • Ocular history:
    1. Idiopathic hypertrophic cranial pachymeningitis resulting in dual sensory impairment (blindness and deafness)
      - Presented with rapidly reducing vision OD in 2005, initially diagnosed as optic sheath meningioma.
        ○ Resulted in optic atrophy OD with NLP vision.
      - Vision OS started to deteriorate in early 2009, with vision decreasing from 20/30 to 5/200+2 on 01/13/10 and progressing to HM in 12/2014
      - s/p trans-sphenoidal biopsy of left optic nerve sheath in 09/24/09: inconclusive
      - s/p biopsy and decompression of left optic nerve in 07/06/10: evidence of chronic inflammation and meningiothelial proliferation
      - s/p biopsy of right optic nerve in 08/04/14: evidence of inflammatory cuff surrounding optic nerve sheath
      - Most recent MRI in 05/20/15 shows diffuse thickened nodular pachymeningeal enhancement along anterior skull base with involvement of CN II, V, VII, and VIII.
      - PET scan in 12/19/14 shows abnormal uptake in pachymeninges adjacent to pons, brain stem, and cervical canal, correlating with abnormal thickening on MRI and consistent with pachymeningitis of undetermined etiology
    2. Primary open angle glaucoma OD>>OS, diagnosed 2004
      - Untreated IOP (mmHg): OD: 31, OS: 25
  • Medical history:
    1. HTN, PTSD, depression, hyperlipidemia, colon polyps, history of thyroid cancer s/p hemithyroidectomy, history of positive PPD and negative quantiferon
  • Ocular medications:
    1. Latanoprost 0.005% qhs OU, timolol maleate gel 0.5% qam OU
  • Systemic Medications:
    1. Famotidine, lisonopril, simvastatin, temazepam, ibuprofen, alendronate, aluminum hydroxide/magnesium hydroxide/simethicone, vitamin D, magnesium supplement

II. Pertinent findings
  • Clinical
    1. BCVA: NLP OD, LP OD
2. Pupils: no reaction OD, 2+ reaction OS with 4+ APD OD
3. Goldmann tonometry: 14 mmHg OU
4. C/D: 0.85 with significant rim thinning sup and inf OD, 0.55 OS
5. ONH: pallor OU

- **Radiological**
  1. MRI 05/20/15: Diffuse thickened nodular pachymeningeal enhancement and thickening of anterior skull base with involvement of cavernous sinus, internal auditory canals, and posterior fossa. Tram track enhancement of the optic nerves (right greater than left) that extends to orbital apices and abnormal enhancement of cranial nerves V, VII, and VIII. Progression since 10/14/11 MRI.

**III. Differential diagnosis**
- **Primary/leading:**
  1. Idiopathic hypertrophic cranial pachymeningitis (IHCP)
- **Others:**
  1. IgG4 mediated disease, neurosarcoïdosis, granulomatous diseases, autoimmune diseases, carcinomatous meningitis, Tolosa-Hunt, intracranial hypotension

**IV. Diagnosis and discussion**
- **IHCP**
  1. Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare chronic and progressive inflammatory disease characterized by a diffuse or localized thickening of the dura mater. Usually seen in older males, the most common symptom at presentation is headache with or without neurologic complications including cerebellar ataxia and multiple cranial nerve palsies.
- **Ocular complications**
  1. Neuro-ophthalmic manifestations due to optic neuropathy are common, resulting in central vision loss, visual field loss, diplopia, or ophthalmoplegia. Ocular symptoms can present as one of the initial signs of the condition, although this is rare. Damage occurs from either compressive fibrosis around the nerve or inflammatory extension of dural involvement surrounding the intracranial optic nerve and cavernous sinus, resulting in optic nerve edema or atrophy.
- **Workup**
  1. Gadolinium enhanced magnetic resonance imaging (MRI) is extremely useful for diagnosis and identification of appropriate biopsy site, but may remain clinically silent for up to two years. Imaging shows thickened abnormally enhancing dura mater, usually with sphenoid wing, tentorium cerebelli, or falx cerebri involvement. Diagnosis is made on the exclusion of other etiologies that can lead to similar MRI findings such as intracranial hypotension, infectious diseases, autoimmune diseases, and malignancies. A dural biopsy is invaluable and often recommended to confirm findings of chronic inflammatory cell infiltrates and to rule out other possible causes.

**V. Treatment, management**
- **Treatment**
  1. The patient was given a three week course of oral methotrexate and a five-day trial of IV methylprednisolone followed by a three-month course of oral prednisone with no overall improvement. Family members appreciated slight subjective improvement of hearing with corticosteroids. While there is no standardized treatment of IHCP, current literature shows improvement of symptoms with long-term corticosteroid and immunosuppressant therapy. Despite this, rare cases have shown no response to therapy, as seen in our patient. More research is needed to better understand the pathophysiological mechanism behind IHCP and identify effective treatment options.
Rehabilitation

The unique presentation of this patient offered challenges to rehabilitation. A step-wise priority-driven approach was adopted to meet his goal of independent living. The primary focus was communication, giving the patient the foundation needed to effectively learn other necessary rehabilitation skills, especially orientation and mobility. The patient responded well to tactile communication with print-on-palm and was able to successfully interact with others while gaining speed and fluency with continued training. Progress was considerably slower compared to most low vision patients and frequent repetition of techniques was required.

VI. Conclusion

IHCP is an extremely rare condition that can produce severe neurologic complications, including irreversible vision loss. Extensive rehabilitation can be utilized to teach these unique patients the skills and concepts needed to regain independence.

Bibliography


Residency affiliation: UC Berkeley Optometry
Residency Name and Location: Primary Eye Care/Low Vision Rehabilitation,VA Palo Alto Health Care System
Available images:
  1. Fundus photos OD, OS
  2. SD-OCT of optic nerve head OD, OS
  3. MRI (05/20/15)

Authors:
- Tina Zheng, OD
- David Yang, OD, FAAO
- Lee Vien, OD, FAAO