Parotid Gland Tumor Masquerading as Dry Eye
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Abstract
Parotid gland tumors can present with seventh nerve palsy, generally a sign of malignancy. This patient, presenting with complaint of dry eye, is diagnosed with seventh nerve palsy caused by a benign parotid gland tumor.

Case History
- Patient demographics: 54 year old, AA, female
- Chief complaint: Patient has noticed dry, red eyes for the past few months that are sometimes watery. The left eye is worse than the right.
- Ocular Hx: none
- Medical history: HTN, hypothyroidism, headaches
- Medications: Levothyroxine, Atenolol, Amitriptyline, Midrin, Naproxen, Flexeril
- Other salient information: Current smoker

Pertinent findings
- Clinical:
  - Best corrected VAs: 20/20 OD and 20/25 OS.
  - Slit lamp exam:
    - lids/lashes: OD normal, OS lower lid laxity
    - conjunctiva: OD normal, OS normal
    - cornea: OD trace diffuse punctate keratitis, OS 3+ central and inferior punctate keratitis
    - lens: OD normal, OS normal
    - posterior segment: OD normal, OS normal
  - Observation: Decreased blink rate OS, lagophthalmos OS
  - Fissure size: 11 mm OD, 14 mm OS, +inferior scleral show OS
  - Exophthalmometry 19/18 base 116
- Cranial nerve exam:
  - CN I: normal, able to identify cinnamon and coffee scents
  - CN II: pupils normal, FDT visual field normal OD and OS
  - CN III, IV, VI: versions smooth and full
  - CN V: Motor – normal, equal on both sides; Sensory - normal, equal on both sides
  - CN VII: smile droops on L side, incomplete lid closure on L side, no resistance to lid closure on L side, asymmetrical brow wrinkling
  - CN VIII: auditory stimulus louder on R side compared to L
  - CN XI: shoulder shrug normal, equal on both sides
  - CN XII: normal, equal on both sides
- Radiology studies:
  - MRI of the head w/ and w/o contrast
1.7 x 1.3 cm enhancing mass of the deep portion of the left parotid gland and
mild asymmetric enhancement of the descending portion of the left facial nerve
as it enters the stylomastoid foramen

- Differentials: adenoid cystic carcinoma, paraganglioma

- Biopsy
  - Fine needle aspiration: hypocellular specimen with rare clusters of atypical cells
  - Non-diagnostic for malignancy, unable to classify type of tumor

**Differential diagnosis**

- Primary: Left incomplete CN 7 palsy with secondary exposure keratitis
- Others: Graves disease was considered given the lower lid laxity, but was ruled out with
  exophthalmometry. The gradual onset of symptoms would be atypical for Bell’s palsy, an
  idiopathic 7th nerve palsy. Bell’s palsy was fully ruled out with MRI.

**Diagnosis and discussion**

This case demonstrates a partial cranial nerve 7 palsy caused by a benign parotid gland
tumor. Although most parotid neoplasms (about 80%) are benign, a facial nerve palsy
caused by a parotid tumor is highly suggestive of malignancy. The presentation of this case is
unique in that the patient presented complaining of dry eye and had not noticed any other
signs of facial nerve dysfunction. Most often, patients with parotid gland tumors will notice a
mass or lump. In this case, the tumor was small and located in the deep lobe of the parotid
gland, so the patient presented with only ocular symptoms. This patient complained of
bilateral dryness which could be mistaken for typical dry eye. The observation of asymmetric
punctate keratitis and lagophthalmos raises suspicion of a problem with lid function or
apposition. On gross observation, a reduced blink rate on the left side and the patient’s
mildly asymmetric smile confirmed an incomplete cranial nerve 7 palsy. Once a new cranial
nerve palsy is diagnosed, the other cranial nerves must be properly examined. In this case,
the patient had a left sided hearing deficit which helped to localize the area of concern. By far
the most common cause of facial nerve palsy is Bell’s palsy, which presents as an acute,
isolated palsy. This is a diagnosis of exclusion. The etiology is presumed to be from herpes
simplex virus. In this case, the gradual onset of symptoms affecting two cranial nerves
eliminated Bell’s palsy as a differential and made further workup with imaging necessary. The
MRI results confirmed a tumor in the deep lobe of the left parotid gland compressing the
facial nerve.

**Treatment, management**

The tumor was surgically resected (parotidectomy) which resulted in worsening of the left
7th nerve palsy. Reconstructive surgery was planned for a later date. For the exposure
keratitis, the patient was initially treated with heavy lubrication instilling preservative free
artificial tears every 2 hours during the day and taping the lid at night. Since her symptoms
worsened after surgery, a gold weight was placed in the left upper lid. This may be removed,
if the nerve function recovers. A partial tarsorrhaphy may be considered if the function does
not improve.
**Conclusion**

- Clinical pearls
  - A parotid gland tumor is a potential cause for CN 7 palsy. A parotid gland mass may be palpable in the preauricular area or inside the mouth. A parotid gland tumor that presents with CN 7 palsy generally suggests malignancy and a poor prognosis.
  - Asymmetric punctate keratitis should alert the clinician to evaluate for an issue of lid function and apposition.
  - Cranial nerve exam is a quick and easy way to determine if a palsy is isolated and should be performed for any newly diagnosed cranial nerve dysfunction.

- Bibliography
“A Curious Case of Monocular Oscillopsia”
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Abstract: This 29-year-old female patient who suffered with superior oblique myokymia symptoms for 8 years reports worsening of her symptoms. She was prescribed topical timolol, which eliminated her symptoms within 1–2 days of starting the treatment.

I. Case History
   a. 29 year old white female
   b. CC: “diplopia/quivering of vision”
   c. HPI: OD only, onset 8 years earlier, worsening over past 10 weeks, multiple times per day, unpredictable onset, near work makes things worse, each occurrence lasts 3-10 seconds on average, (-) trauma
   d. Ocular Hx: unremarkable; does not wear Rx
   e. Medical Hx: unremarkable except 10 weeks post-partem delivery without complications
   f. Medications: denies
   g. Family Medical Hx: unremarkable
   h. Allergies: No Known Drug Allergies

II. Initial Exam
   a. Visual Acuity: 20/20 OD, OS, OU (sc)
   b. Pupils, Confrontation Fields, EOM’s: Normal OU
   c. Cover Test: orthophoric distance and near (sc)
   d. Subjective Manifest Refraction
      i. +0.50 sph OD
      ii. Plano OS
   e. Goldman Tonometry: 18 mmHg OU
   f. Dilated Fundus Exam: Normal OU
   g. Baseline Humphrey Visual Field Test: reliable and normal OU

III. Follow-up Exam
   a. Clinical findings same as previous
   b. Ocular Coherence Tomography: Normal OU
   c. Park’s 3-Step: UTT (no hyper deviation)
   d. Maddox Rod 9 DAF’s: Normal OU
   e. Marginal Reflex Distance: equal and normal OU
   f. Blood Pressure: 118/68
   g. Cranial Nerve Testing
      i. CN II → VIII tested; all normal upon testing
IV. Laboratory studies
   a. ANA: marginally elevated; reflex ds-DNA test
   b. Myasthenia Panel: normal
   c. ESR: normal
   d. Thyroid panel: normal
   e. RPR: normal
   f. ds-DNA: normal

V. Radiology studies ordered
   a. MRI/MRA ordered by neurology → normal

VI. Differential diagnoses
   a. Primary/Leading → Superior Oblique Myokymia (SOM)
   b. Acquired/congenital nystagmus
   c. Opsoclonus

VII. Diagnosis and discussion
   a. Superior oblique myokymia
      i. Background; definition; clinical findings; patient symptoms
   b. Demographics
      i. Females > males
      ii. Triggers
   c. Pathogenesis
      i. Option 1 → aberrant regeneration
      ii. Option 2 → microvascular compression
   d. Ominous Causes
      i. Tumors, CVA’s, A/V malformations, multiple sclerosis, hydrocephalus, cerebral cysts
   e. Anatomy Review
      i. Brainstem and Cranial Nerve IV anatomy
      ii. Blood supply to dorsal root exit zone of CN IV on dorsal side of brainstem
   f. Imaging Recommendations for suspected cases of SOM
      i. MRI high recommended
         1. Small slices required to see microvascular compression on MRI
      ii. Lab tests: unknown if valuable at this time in SOM
   g. Treatment
      i. Observation
      ii. Medical Management
         1. Gold/historical standard: carbamazepine
         2. Others: gabapentin, phenytoin, memantine, baclofen, clonazepam, propranolol, Botox
      iii. Surgery
         1. EOM surgery to weaken superior oblique
2. Multiple techniques used
3. Microvascular decompression
   a. Skull base/brainstem surgery; invasive
h. New treatment with topical beta-blockers?
   i. One case report from 1994 found relief of SOM with topical betaxolol
   i. What I did to manage patient
      i. Based off of one case report from 1994; I initiated topical timolol 0.5%
         BID OS
      ii. Symptoms/Signs resolved in 2 days
j. Localized vs. Systemic Theory of Beta-Blockers in Treatment of SOM
   i. Competing theories at this point
   ii. Localized $\rightarrow$ topical beta-blockers work only on the superior oblique
        muscle itself or nerve endings at the level of the globe
   iii. Systemic Theory $\rightarrow$ enough of the topical beta-blocker medication gets
        absorbed through conjunctival vessels to elicit an effect on a systemic
        level
   iv. I had patient use timolol in contralateral eye to test localized vs. systemic
       theory
       1. Within 2 days of use in the contralateral eye her SOM returned
       2. Within 2 days of re-initiating treatment in initial eye, her SOM
          disappeared again
       3. Good evidence to maybe suggest Localized Theory is more
          accurate than Systemic Theory
k. Potential New Associations with SOM
   i. Trauma $\rightarrow$ Men
   ii. Pregnancy $\rightarrow$ Women

VIII. Conclusions/Clinical Pearls
   a. Monocular oscillopsia?? Think of SOM
   b. Seal diagnosis with attentive case history and slit lamp exam
   c. Neuro imaging required to rule out ominous causes
   d. Consider topical beta-blocker therapy
      i. Dirt cheap! $4 list at Target/Walmart/etc.
   e. If no response then refer to neurology for trial of oral medications
   f. Surgery is a last resort only

IX. References
   a. 32 total references used
Abstract: In most cases of leukemic optic nerve infiltration, there is already evidence of CNS disease. In this patient, the optic nerve was the first sign of CNS involvement in an acute lymphoblastic leukemia relapse.

I. Case History
   a. Demographics: A 12 year-old Caucasian male was referred to our clinic by his oncologist.
   b. Chief Complaint: Pain and blurred vision OS, especially on eye movement, which started a few weeks prior.
   c. Ocular History: Mild myopia OU, otherwise unremarkable.
   d. Medical History: Significant for acute lymphoblastic leukemia (ALL), which had been diagnosed in 2012. He received a bone marrow transplant in August of 2014, and he had a relapse in October of 2014. Past surgical history was significant for tricuspid valve replacement, bowel resection, and cholecystectomy. He was also being treated for endocarditis of the prosthetic tricuspid valve. His chemotherapy had been put on hold as he was pancytopenic when he came to our clinic.
   e. Medications: amlodipine, ceftriaxone, clonidine, dexamethasone, escitalopram, micafungin, pentamidine, potassium chloride, and valacyclovir.

II. Pertinent Findings
   a. Clinical/Physical: BCVAs were 20/20 OD and 20/20-2 OS. Pupils were equal and reactive to light and accommodation OU, with 0.5 log unit RAPD OS. EOMS showed FROM OU. On anterior segment examination, mild dry eye syndrome was noted OU. Anterior chambers were clear, quiet and without iris abnormalities in either eye. Intraocular pressures were normal OU. Dilated fundus examination revealed extensive optic disc edema OS with an infiltrative lesion in the nerve. There was mild disc edema OD. Maculae appeared flat with good foveal light reflexes OD and OS. Retinal vasculature appeared normal OU. Optic nerve OCT showed mild optic disc edema OD (111 micron thickness) and extensive optic disc edema OS (267 micron thickness). Visual fields (Humphrey 24-2) showed enlarged blindspots OU.
   b. Laboratory Studies: No abnormal cells in latest CSF sample taken 3 days prior.
   c. Radiology studies: MRI of the brain and orbits showed enlargement of the left optic nerve and prominence suggesting optic disc edema or papillitis. No mass effect, hemorrhage or restricted diffusion to suggest ischemic event.
   d. Other: B-scan showed a medium reflective lesion at the left optic nerve 2.3 mm in size. No optic disc drusen OU.

III. Differential diagnosis
   a. Primary
      i. Optic Neuritis
      ii. Leukemic Infiltration of the Optic nerve
      iii. Papilledema Secondary to CNS Infiltrate
      iv. Ischemic Optic Neuropathy
b. Other
   i. Opportunistic Infection of the Nerve
   ii. Pseudotumor Cerebri

IV. Diagnosis and discussion
Diagnosis: Leukemic Infiltration of the Optic Nerve in a Patient with Acute Lymphoblastic Leukemia

Primarily seen in children, acute lymphoblastic leukemia (ALL) is a relatively common form of leukemia. 70-80% of children that are diagnosed with it are cured with current chemotherapy regimens. In the patients that have a relapse, CNS involvement is very common. Prophylactic brain irradiation and intrathecal therapy are used in patients in remission to prevent a CNS relapse of the disease. Studies have shown that the optic nerve could provide a sanctuary for leukemic cells during irradiation and intrathecal therapy. Because of this, the optic nerve has also been found in a few case reports to be the initial site of relapse of the disease.

Camera et al indicated that optic nerve infiltration occurs in approximately 1.4% of pediatric patients with ALL. Further studies have suggested that ocular involvement is usually an ominous sign in the setting of ALL. In 15 year study, 28 patients in a pediatric population of 131 children with leukemia developed ocular involvement. Of those 28 with ocular involvement, 27 died within 28 months.

In addition to optic nerve infiltration, there are several other ways that ALL can manifest in the eyes. Patients with ALL can also develop the following ocular conditions:
- Orbits: proptosis, diplopia
- Lids: edema, chemosis,
- Cornea: ring ulcers, infiltrates
- Conjunctiva: hyperemia and edema of lower sub-palpebral conjunctiva
- Anterior Chamber: uveitis, hypopyon
- Retina/Vitreous: hemorrhages, cotton wool spots, vascular occlusions, edema, vitritis

V. Treatment, management
a. Treatment and response to treatment:
Numerous cases in the literature indicate that treatments of leukemic optic nerve infiltration with irradiation and intrathecal chemotherapy achieve the best outcomes and preserve the most vision. They also suggest that the irradiation be done as promptly as possible. Intrathecal chemotherapy alone is associated with worse visual outcomes, but could be indicated if the patient’s vision is largely unaffected during an infiltrative episode.

After being seen in our clinic, the patient was sent to Neuro-Ophthalmology at the University of Iowa. They agreed with the assessment and decided to start intrathecal chemotherapy, given that the vision was still 20/20-2 in the affected eye (left eye). He responded very well, and the edema and pain subsided. A few months later, however, the patient had sudden loss of vision and
intense pain of the right eye. Clinical examination revealed optic nerve infiltration OD, and pallor OS. This time, Neuro-Ophthalmology proceeded with irradiation of the optic nerve OD and continued intrathecal chemotherapy. Despite the radiation, vision in the right eye never recovered (BCVA was count fingers at 3 feet). Luckily, the left eye BCVA was still 20/30 after recovery of the initial infiltration. Unfortunately, one month later, the patient’s health continued to decline, and he passed away from the leukemia.

VI. Bibliography


VII. Conclusion

a. Clinical pearls, take away points if indicated:

i. Oncologists should have an eye care professional examine any leukemia patient complaining of sudden vision loss or eye pain.

ii. Prompt irradiation of the optic nerve in conjunction with intrathecal chemotherapy is indicated for best chance of preserving vision in leukemic infiltration of the optic nerve.

iii. Ophthalmic involvement in ALL is usually associated with a poor prognosis for survival.