Case Report

I. Case History
- Patient demographics: 69 year old Caucasian male
- Chief complaint:
  - Ocular irritation: left eye more than right eye, onset one week ago, moderate severity, accompanying blurry vision, tearing, discharge, redness
  - Associated with onset of facial droop left side (08/09/16), followed by facial droop right side (08/16/16)
- Ocular history:
  - Patient reported history of lazy left eye as a child with patching. No history of ocular trauma or ocular surgery.
  - Monitored by outside eye care provider for glaucoma suspicion without treatment, and cataracts
- Medical history:
  - CAD s/p CABG 2013, HTN, paroxysmal atrial fibrillation, obesity, OSA, myoclonic epilepsy s/p remote TBI, hyperlipidemia, PTSD, major depressive disorder, rosacea
- Inpatient medications:
  - Atorvastatin, cholecalciferol, clonazepam, lamotrigine, metoprolol, sodium chloride IV
- Other salient information:
  - Timeline of facial palsy onset:
    - (08/09/16) Left ear pain. Left sided facial drooping noted by wife. Patient evaluated at Salem Hospital, diagnosed with Left sided Bell’s Palsy, initiated medication (valacyclovir and prednisone) and discharged
    - (08/17/16) Follow up with private doctor who noted right sided facial drooping
    - (08/17/16) Admitted to West Roxbury, VA in Massachusetts, for progressive bilateral Lower Motor Neuron (LMN) cranial nerve VII palsy

II. Pertinent findings
- Clinical:
  - Visual acuity with correction (Feinbloom chart) OD 10/20 OS 10/40; PH NI
  - Normal: confrontation fields, pupils, versions
  - Slit lamp:
    - External: good Bell’s Reflex OU
    - Lids: lagophthalmos with exposed sclera inferior OS 6mm>OD 2mm
    - Conjunctiva: moderate diffuse injection 360 of bulbar conjunctiva OU, stringy mucous discharge inferior lid margin OS>OD
    - Cornea: OD moderate SPEE inferior third with diffuse fine punctate staining on fluorescein inferior. OS moderate SPEE inferior half with coalesced punctate staining on fluorescein inferior temporal
    - Open angles. No anterior chamber reaction. Iris flat and intact
  - IOP with Perkins: 17 mm Hg OD/OS at 5:25pm
  - Dilated fundus exam
    - Lens: mild cortical changes with grade 3+ nuclear sclerotic changes OU
    - Media: vitreal syneresis, no posterior vitreous detachment OU
    - Unremarkable: macula, vessels, posterior pole OU
Periphery: scattered reticular degeneration OU

Physical:
- Cranial Nerve 7 Testing:
  - Eye brow raise unappreciable right and left
  - Eyelid closure: incomplete left > right. Orbicularis oculi strength: right > left
  - Puff cheeks: unable to purse lips, greater filling right vs left
  - Show teeth: right nasolabial fold when trying to smile, unappreciable on left

Laboratory studies:
- CBC: elevated WBC, neutrophils, monocytes, prothrombin time; depressed lymphocytes
- Lyme antibody serum: detected
- Lyme IgG & IgM: positive
- EBV IgG: detected
- CSF: positive for Lyme, elevated protein and WBC
- RPR, HIV 1&2, VDRL, ANA, Hep B, VZV: non-reactive

Radiology studies:
- CT head without contrast: negative for acute intracranial abnormality
- MRI with contrast: diffuse cranial nerve enhancement involving the bilateral oculomotor, trigeminal, and facial nerves, as well as bilateral 9-11th cranial nerve complexes. No evidence of acute infarct, hemorrhage, intracranial mass, or hydrocephalus
- CT thorax: no evidence of sarcoidosis. Bilateral pulmonary nodules. Left sided nephrolithiasis without evidence of hydrourephrosis

III. Differential diagnosis
- Primary: Simultaneous Bilateral Cranial Nerve VII Palsy secondary to Lyme Neuroborreliosis
- Others: cerebrovascular accident; infectious EBV, HIV, tuberculosis, meningitis; granulomatous sarcoidosis; demyelinating Guillain-Barre Syndrome; neoplastic carcinomatosis or lymphoma

IV. Diagnosis and discussion
- Condition began with left ear pain followed by sudden left facial droop. Unilateral LMN left cranial nerve VII palsy with no other neurological factors drove the diagnosis of unilateral Bell’s Palsy and appropriate treatment was initiated. Eight days later, sudden right facial droop was noted changing the condition to simultaneous bilateral LMN cranial nerve VII palsy, which presented a diagnostic dilemma prompting further work up. The etiology was revealed by positive Lyme titers in the setting of Lyme endemic Massachusetts during the summer months. Appropriate treatment and slow resolution of neurological and ocular symptoms followed
- Cranial nerve VII
  - Provides innervation to facial musculature: frontalis, orbicularis oculi, buccinators, orbicularis oris, platysma, posterior belly of digastrics, and stapedius muscle
  - LMN lesion affects all the facial muscles on the ipsilateral side to the lesion due to peripheral innervation by the ipsilateral VII nerve. A LMN lesion would appear as smoothing of the brow, open eye, flat nasolabial fold, and drooping of the mouth
  - Upper Motor Neuron (UMN) lesion does not affect the brow and eyelid muscles on the contralateral side to the lesion due to central innervation by the contralateral cerebral cortex. Central innervation is bilateral for the upper face, and unilateral for the lower face. An UMN lesion would appear as flattening of nasolabial fold and drooping of the mouth
• Etiologies of unilateral palsy: idiopathic Bell’s Palsy, viral: HSV, HZV, CMV, EBV, adenovirus, coxsackievirus, mumps, rubella; Lyme disease, otitis media, blunt facial trauma, iatrogenic injury (parotid/mastoid surgery), hemangioma/neuroma or tumor surrounding the facial nerve, Ramsay Hunt Syndrome

• Etiologies of bilateral palsy: infectious from: Lyme disease, Epstein-Barr virus, HIV, syphilis, mononucleosis, meningitis; Guillain-Barre Syndrome, multiple idiopathic cranial neuropathies, sarcoidosis, brain stem encephalitis, benign intracranial hypertension, diabetes mellitus, vasculitis, bilateral neurofibromas, leukemia, intrapontine/prepontine tumor, Melkersson-Rosenthal syndrome

• Lyme borreliosis
  • Tick transmitted multisystem inflammatory disease caused by the *B. burgdorferi* spirochete. Lyme disease has become the most common vector-borne disease in the United States with approximately 20,000 new cases reported each year
  • Prevalence varies by
    - Geography: in 2011, 96% of Lyme disease cases were reported in 13 states: CT, DE, ME, MD, MA, MN, NH, NJ, NY, PA, VT, VA, WI
    - Season: peaks during May and June
  • Incubation varies from 3-32 days followed by stages
    - One: erythema migrans (hallmark target like rash), flu like symptoms: fever, headache, malaise, myalgias
    - Two occurs after several weeks or months: neurologic abnormalities in 15%, cardiac involvement in 8%
    - Three: chronic monoarticular or oligoarticular Lyme arthritis involving large joints
  • Diagnosis:
    - History of tick exposure, epidemiology, clinical signs and symptoms, and immunologic assay using antibody titers against the spirochete
    - Erythema migrans: most specific and sensitive than serology in early Lyme
    - Evidence of antibodies against *B. burgdorferi*: enzyme immunoassay (EIA), Western blot with abnormal IgM (2 bands) and IgG (5 bands)
    - CSF: elevated CSF lymphocytes, monocytes, plasma cells; elevated protein; PCR detected *Borelia* species antigens
  • Lyme Neuroborreliosis (LNB) occurs in 10-15% of untreated Lyme disease, when systemic infection results in neurologic involvement
    - Presentation includes: subacute meningitis within weeks to months of untreated erythema migrans; most commonly, CN VII palsy, unilateral more than bilateral
    - Orbital and Ocular LNB: rare, can occur at all three stages with the most common being uveitis and optic neuritis
      - First: follicular conjunctivitis and episcleritis
      - Second: uveitis and neuro-ophthalamic disorders
      - Third: keratitis, chronic intraocular inflammation, orbital myositis

• Unique features
  • Unilateral CN VII palsy is common with an incidence of 25 per 100,000. Seventy percent of those are attributed to idiopathic Bell’s Palsy
  • Bilateral CN VII palsy is rare (only 2% of all CN VII palsies) with an incidence of 1 per 5,000,000. The majority of those are attributed to an underlying systemic condition. Only
23% are attributed to Bell’s Palsy. Bilateral CN VII palsy is considered to have simultaneous onset when the opposite side becomes involved within 30 days of the first side.

- Unilateral corneal exposure keratopathy can be addressed with multiple treatment options including: lid taping, ophthalmic ointment, bandage or scleral contact lens, amniotic membrane, tarsorrhaphy, or copious lubrication. Bilateral corneal exposure keratopathy is a challenge due to the functional compromise in visual acuity with the multiple treatment options mentioned above. Thus, the patient’s visual needs must be taken into consideration when determining treatment modality.

V. Treatment, management

- Medical:
  - Initial diagnosis of Unilateral Bell’s Palsy: Valacyclovir and Prednisone PO x 8 days: poor response
  - Ultimate diagnosis of Bilateral CN VII Palsy secondary to Lyme Neuroborreliosis: Ceftrizxone 2mg IV: at 2 week follow up, improvement in right sided palsy, no improvement on left sided palsy

- Ocular:
  - OD: Carboxymethylcellulose 0.5% Q4H, Ophthalmic ointment QHS
  - OS: Ophthalmic ointment TID, Carboxymethylcellulose 0.5% Q4H, Ophthalmic ointment QHS; lid taping QHS
  - Marked improvement in patient comfort OU. Full resolution of exposure keratopathy OD. Improvement OS with moderate residual superficial punctate keratitis, and sectoral bulbar conjunctival injection

VI. Conclusion

- Clinical pearls
  - CN VII palsy
    - Unilateral LMN peripheral palsy results in paralysis of one side of the facial musculature: smoothing of the brow, open eye, flat nasolabial fold, and drooping of the mouth. The most common etiology is idiopathic Bell’s Palsy, or herpetic infection. Other possibilities to consider: h/o facial trauma or surgery, slow onset due to tumor, h/o tick bite or outdoor activities in Lyme disease endemic regions
    - Unilateral UMN central palsy results in paralysis of one side of only the lower facial musculature: flattening of nasolabial fold, and drooping of the mouth. The most common etiology is cerebrovascular accident
    - Bilateral LMN palsy results in paralysis of both sides of the face. The majority of these cases have a serious underlying medical condition and need to be admitted for further work up and management
  - Exposure keratopathy
    - Unilateral: lid taping, ophthalmic ointment, bandage or scleral contact lenses, amniotic membrane, tarsorrhaphy, or copious lubrication
    - Bilateral: challenging to maintain functionally acceptable visual acuity during treatment
      1. Lid taping/aggressive ointment instillation/amniotic membrane into worse eye. Copious lubrication into better eye
2. Alternate which eye is to receive aggressive lubrication day to day
3. Bandage or scleral contact lenses on both eyes
4. Reversible marginal tarsorrhaphy to both eyes with copious lubrication

- **Update:**
  - **Follow up 1 (08/22/16)**
    - Visual Acuity (Snellen): OD 20/40 OS 20/100
    - Cornea: improvement OD > OS; OD mild superficial punctate keratitis inferior; OS moderate superficial punctate keratitis central to inferior half
    - CN VII testing: no improvement from baseline exam
  - **Follow up 2 (08/31/16)**
    - Visual Acuity (Snellen): OD 20/30+ OS 20/100+
    - Cornea: marked improvement OU; OD clear; OS moderate superficial keratitis inferior-nasal quadrant extending to fixation
    - CN VII testing: improvement on right side in all parameters, no change on left side
References:


