Recurrent facial palsy in Melkersson-Rosenthal syndrome: a case report

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Abstract:

Melkersson-Rosenthal syndrome (MRS) is a rare condition of unknown etiology with recurrent facial palsy, fissured tongue (lingua plicata) and orofacial edema symptoms. This case report identifies a fifty year old male with recurrent MRS.

Case Outline:

I. Case History

- Patient demographics: 50 year old African American male, Mr. N.D.
- Chief complaint: left side facial weakness for 1 week accompanied by light sensitivity, headaches on his left side, intermittent sharp pain in the left ear, dry eye OS.
  - Mr. N.D. reports that this is his sixth episode of facial palsy with his last episode in 2006 and that it is secondary to his long standing diagnosis of Melkersson-Rosenthal Syndrome.
  - Mr. N.D. has self-treated his dry eye with lubricating ung OS qhs, patching OS qhs and artificial tears 1 gtt OS>OD q3h x 1w. However, he has run out of artificial tears and ung is expired.
- Ocular history:
  - Normal tension glaucoma suspect OU
  - History of exposure keratitis OS and left facial palsy secondary to Melkersson-Rosenthal syndrome with residual left orbicularis weakness
  - Refractive error and presbyopia OU
  - Retinal lattice degeneration OS
- Medical history:
  - Coronary artery disease status post bypass in 2011
  - Dyslipidemia
  - Hypertension
  - Melkersson-Rosenthal Syndrome
  - Asthma
  - Depression
  - Lower back and knee pain
- Medications
  - Albuterol 90Mcg 200D, inhale 2 puffs by mouth every 4 hours prn
  - Aspirin 325mg ec Tab, 1 po qd
  - Atorvastatin Calcium 40 mg tab, 1 tab po qhs
  - Bupropion Hcl 75mg Tab, 1 tab po bid
  - Carboxymethylcellulose Na 0.5% oph soln, 1 gtt OS qid prn
  - Cetirizine 10mg Tab, 1 tab qd
  - Ketotifen 0.025% oph soln, 1 gtt OU bid prn
  - Metoprolol Succinate 50mg Sa Tab, ½ tab po qd
  - Nitroglycerine 0.4mg Sl Tab, dissolve 1 tab under tongue q5 min prn
  - Zolpidem Tartrate 10mg Tab, 1 tab po qhs prn
- Other salient information:
  - Per Neurology exam 05/12/06:
    - First episode ~ 1999, woke up with face tingling which developed into left facial droop which lasted for 3 months. During this time he couldn’t close
eyes, experienced drooling, had swelling on left face, lips and tongue. He had continuous headaches and had no sense of taste.

- Second episode ~ 2001 and third episode 05/2002: Similar symptoms which lasted 3 months and was successfully treated with oral prednisone and antivirals.

- Fourth episode ~ 2004: symptoms started as a left ear ache and progressed rapidly over next 3 days to his most severe attack with ear drum rupture, runny nose, entire facial numbness, left facial palsy, left face and lip swelling. Mr. N.B was treated with Salsalate 500mg po qid, Valcyclovir 500mg bid x 10d, Prednisone 5 mg tid x 3d, 5 mg bid x 3d, 5 mg qd x 3d.
  - Previous workups included 3 unremarkable MRIs and CTs, serology work up that was normal for CBC, electrolytes, negative for HVF, RPR, HCV and lyme, elevated cholesterol and LDL, normal CXR, and negative ACE.
  - Due to occurrence of lingual plicata, facial swelling and facial nerve palsy he was diagnosed with Melkersson-Rosenthal syndrome.
  - The tentative plan was to wait for the next attack and then surgically decompress the left CN VII after discharge from the army in 2005.

- Exam findings:
  - Pupils, EOMs, VF, C/Ds normal
  - facial sensation reduced on left face forehead, V2 and V3 distribution
  - forehead folds flatter on left, orbicularis oculi weaker on left, nasolabial sulci flatter on left
  - corneal sensation intact bilaterally
  - tongue is midline
  - palate elevation is symmetrical and uvula is midline
  - gag reflex is intact
  - shoulder shrug and head turning are equal
  - unremarkable cerebellar, sensory and gait

- Assessment: Melkersson-Rosenthal syndrome with left facial nerve and trigeminal nerve impairment associated with left facial and tongue swelling
  - Per neurology exam 01/17/08:
    - Patient reported 3 mild attacks of left facial numbness with lingual swelling over past year, most recently 1 month previous to exam
    - Patient was treated with acyclovir, prednisone and hydroxyzine and symptoms resolved in all episodes
    - Left V2-V3 hypesthesia and mild dysarthria unchanged
    - CT ordered and completed on 04/17/08: unremarkable
  - Per neurology exam 02/03/09:
    - Patient reported 2-3 mild attacks of left facial numbness with lingual swelling over past year, most recently 2 months prior to exam
    - Patient was treated with acyclovir, prednisone and hydroxyzine and symptoms
    - Left V2-V3 hypesthesia and mild dysarthria unchanged

II. Pertinent Findings

- Clinical:
  - Pupils, EOMS and VF normal
  - OS: 1+ SPK
  - Cranial nerve assessment:
    - II normal: VAs OD & OS 20/20
    - III, IV, VI normal: Cover test at distance cc orthophoria
• V abnormal: patient reports slight numbness and tingling on all 3 dermatomes on left side when stimulated with finger tips
• VII abnormal: reduced eyebrow raise, smile and puffed cheeks on left side
• VIII normal: equal hearing of rubbed fingertips on left and right side
• IX normal: equal palate raise
• X normal: able to swallow
• XI normal: equal shoulder lift and strong resistance
• XII normal: no tongue deviation
  o IOP OD: 17 mmHg OS: 18 mmHg at 10:54am via Goldmann applanation tonometry
• Physical
  o Left eyelid more sluggish with incomplete blink
  o Speech fluent and intact
  o Alert and oriented
  o Mild facial asymmetry at corners of mouth
  o Blood pressure: 107/70
  o Weight 215 lb
  o Height 71 in
• Laboratory studies: none
• Radiology studies: none
• Others: (-) family history of Melkersson-Rosenthal syndrome

III. Differential Diagnosis

• Primary/leading: Recurrent acute left facial nerve palsy with left V2 and V3 trigeminal nerve involvement secondary to Melkersson-Rosenthal syndrome causing trace exposure keratopathy OS
• Others: Recurrent left Bell’s palsy, transient ischemic attack, trigeminal neuralgia

IV. Diagnosis and Discussion

• Elaborate on the condition:
  o Melkersson-Rosenthal Syndrome (MRS) is a rare condition with unknown exact etiology. Classic signs of MRS include recurrent facial nerve palsy, facial edema and lingua plicata (fissured tongue). However, the triad is present concurrently in less than a third of patients and instead may present with one or more symptoms or with consecutive symptoms. Facial palsy may present on one or both sides of the face. The incidence of MRS with facial palsy is 0.36 in 100,000 patients per year and does not have a higher incidence in certain ethnicities or genders. While MRS can affect any age group, those between 25 and 40 years old are more commonly affected. 75% of MRS patients with facial palsy have at least 2 episodes indicating that recurrent facial palsy is quite common. Research indicates that facial palsy in MRS is possibly caused by viral infection, similar to Bell’s palsy, however recurrent facial palsy in MRS has worse prognosis than recurrent Bell’s palsy. Facial nerve impairment may become permanent following multiple episodes of MRS.
  o Diagnosis of MRS if very difficult and is likely underdiagnosed. MRS is often a diagnosis of exclusion or only confirmed after recurrent episodes. Diagnostic investigations including histology may not be necessary because MRS is a clinical syndrome. CT, MRI and Chest X-ray may be necessary to rule out associated conditions.
• Expound on unique features:
  o Although not a classical feature, Mr. N.D. exhibited recurrent concurrent impairment of the trigeminal nerve during active MRS episodes. Patients in other such reports
have exhibited involvement of other cranial nerves including the hypoglossal, glossopharyngeal, auditory and olfactory nerves.

V. Treatment and Management

- Treatment and response to treatment:
  - Prescribed artificial tears prn OU, lubricating ung OS qhs with eye patch OS qhs and Ketotifen bid OU.
  - Advised evaluation by primary care physician who prescribed Prednisone 10mg Tab 6 tabs po x 7 days and Acyclovir 800mg 1 tab po x 7 days.
  - Symptoms resolved with treatment.
- Refer to research where appropriate:
  - Steroids, steroid-antibiotic combinations (minocycline), steroid-antileprosy combination (clofazimin, dapsone) and NSAIDS have been used for treatment. Facial nerve decompression may become necessary for patients with recurrent MRS facial paralysis.
  - Prednisolone is the leading treatment for facial palsy in MRS but its benefit is not fully determined and outcomes can still be poor with treatment.

VI. Conclusion

- Clinical pearls, take away points if indicated:
  - Melkersson-Rosenthal Syndrome, although rare and poorly understood, is an important condition for eye care providers to be aware of in the context of a patient with recurrent facial paresis in order to provide proper treatment. Possible multiple cranial nerve involvement requires understanding of a concise clinical cranial nerve assessment. As with many conditions, an accurate history is imperative in diagnosing MRS.

VII. Bibliography

