Zoster Imposter: Corneal Involvement in a Patient with Ramsey Hunt Syndrome

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Abstract:

A patient presents to optometry from the emergency room due to new onset left side facial paralysis. Subsequent appointments reveal pseudo-dendrites and pain in ipsilateral ear leading to the diagnosis of Ramsay Hunt Syndrome.

I. Case History
   1) Patient demographics: 56 year old African-American male.
   2) Chief complaint: Left side facial paralysis and incomplete lid closure for 5 days.
   3) Ocular history: Primary Open Angle Glaucoma Patient, lost to follow up. Compound Myopia.
   4) Medical history: Hypertension, diabetes, tinnitus, and two right shoulder surgeries. Patient has history of noncompliance with medications.
   5) Medications: Patient had been prescribed latanoprost QHS OU and Cosopt BID OU with a history of noncompliance over the last year. Patient is also taking 1000mg of metformin and 100 units of insulin for diabetes. Lastly, our patient was also taking 25mg of hydrochlorothiazide for hypertension control. After the diagnosis of acute Bell’s palsy was made at the emergency room, the patient was started on Acyclovir 400mg for two days and Prednisone 10mg six times a day for one day.
   6) Other salient information: Inpatient assessment prior to optometric referral revealed facial pain. Subsequent visits at the optometry clinic revealed further complications as discussed below.

II. Pertinent Findings
   1) 5 days post-acute Bell’s palsy onset
      a) Clinical: Presenting BCVA was 20/40 in habitual glasses OD/OS which pinholed to 20/25 OD/OS. Pupillary testing, extraocular muscles, and confrontation visual fields were normal. Adnexa showed left side facial paralysis including incomplete smile and incomplete lid closure of the left side. Slit lamp exam showed mild inferior staining OS and no other ocular pathology. Intraocular pressures were 20/21 and dilated fundus exam revealed glaucomatous cupping. Optical coherence tomography of the optic nerve head confirmed no edema or elevation in both eyes (see radiology results below). Patient reported equal color OD/OS on red cap desaturation. Reinstated latanoprost QHS OU and Cosopt BID OU for glaucoma control. In addition, patient was started on lubrication for minimal exposure keratopathy. Patient was to return to clinic in one month for Bell’s follow up and to update visual field.
b) Laboratory Results: When patient presented to ER five days prior to optometry appointment, HbA1C was 8.3. Other blood work was unremarkable.

c) Radiology studies: MRI studies showed no evidence of acute infarction. Optic nerve sheath enlargement with widening of the CSF space around the optic nerves was questioned along with a partially empty sella turcica. No flattening of the posterior globes or protrusion of the optic nerves into the posterior aspect of the globes is noted. Carotid Doppler and CT findings were negative.

2) 27 days post-acute onset Bell’s Palsy

a) Clinical Findings: Presenting BCVA was 20/40 OD, PH 20/25, and 20/80 OS which showed no improvement with pinhole. Pupillary testing, extraocular muscles, and confrontations were again normal. Adnexa showed improved left side facial paralysis; however, smile was still reduced and lid closure was not complete on the left side. Slit lamp exam showed extensive pseudodendrites through the visual axis which stained with rose bengal and fluorescein. There was no facial lesion or Hutchinson’s sign and the anterior chamber was quiet. Intraocular pressures were 18/20 with questionable compliance taking latanoprost and Cosopt. Dilated fundus exam was unchanged from the previous exam. Patient briefly mentioned reduced hearing in left ear since onset of Bell’s, but no further action was done at this time.

b) Meds: Patient started on oral acyclovir 800mg 5x/day x 10 days, Ocuflox QID OS, and Bacitracin ung QHS OS. Patient instructed to continue surface lubrication and Cosopt BID OU and to discontinue latanoprost.

3) 30 days post-acute onset Bell’s Palsy

a) Clinical Findings: Presenting VA was 20/40 OU with best corrected vision of 20/20 OD/OS. Pupillary testing, extraocular muscles, and confrontations were again normal. Adnexa showed improved left side facial paralysis; however, smile was still reduced and lid closure was not complete on the left side. Slit lamp exam showed complete resolution of pseudo-dendrites and no staining with rose bengal. Otoscopic examination revealed no vesicles; however, patient still complained of severe ear pain on the involved side. Intraocular pressures were 17/20 with questionable compliance taking Cosopt. One sample of salt and one sample of sugar were indistinguishable by patient using the tip of his tongue confirming impairment of CN 7.

b) Others: Discussion with ENT physician confirmed Ramsay Hunt syndrome and proper management with oral acyclovir. ENT physician deemed oral steroids were not indicated.

c) Meds: Patient to finish 10 day course of acyclovir and continue bacitracin ung QHS OS. Pt to also continue surface lubrication and Cosopt.
III. Differential diagnosis

1) Primary/leading: Ramsay Hunt Syndrome. Ramsay Hunt syndrome is a facial nerve paralysis from herpes zoster with ear or throat involvement. Patients develop a painful prodrome and ENT exams will reveal hearing loss on the ipsilateral side of the palsy in addition to a vesicular eruption in the ear canal and/or pharynx (1).

2) Others: For patients presenting with an acute Bell’s palsy, extensive cranial nerve testing should be performed along with MRI/CT if additional neurological abnormalities are found to rule out an infarction. Other differentials to consider would be Lyme disease; however, the patient did not have any recent exposure to ticks and didn’t present with any rash. Facial nerve palsy’s can also be caused by chronic otitis media. This type of facial paralysis occurs gradually and is accompanied with ear pain and fever. Our patient’s facial paralysis was acute and he did not possess a fever. Laboratory testing should be performed if the patient is suspected to have Lyme disease or diabetes. More than 10 percent of patients with Bell’s palsy have diabetes mellitus (1). Our patient is a known type 2 diabetic, and his medication was altered after laboratory testing revealed an elevated A1C.

IV. Diagnosis and discussion

1) Elaborate on the condition: Bell’s palsy is an isolated 7th nerve palsy previously attributed to compression of the facial nerve by unknown swelling or edema. More recently, it has been suggested that Bell’s palsy may be associated with the reactivation of herpes simplex type 1 (2). When Bell’s palsy is associated with varicella-zoster, it is referred to as either Zoster Sine Herpete or Ramsay Hunt syndrome with the latter being associated with hearing loss (3). Annually, approximately 15 to 30 per 100,000 persons develop a Bell’s palsy. The peak age occurrence is in the 40s affecting both men and women and left and right sides of the face equally. Patients who have had one episode of Bell’s palsy have an 8 percent risk of recurrence (1). A study looking at over 1500 Bell’s patients found that 12% had the triad of facial palsy, ear pain, and vesicular eruption, thus giving them the more specific diagnosis of Ramsay Hunt (4).

2) Expound on unique features: In Ramsay Hunt syndrome, only 5% of patients have residual hearing loss (5) Reports have found that ocular involvement in Ramsay Hunt syndrome is rare (6). A review of ocular findings in Ramsay Hunt syndrome listed nystagmus, acute retinal necrosis, uveitis, retrobulbar optic neuropathy, ophthalmic artery obstruction, acute ipsilateral glaucoma, and CN 3,4, and 6 nerve palsies as the reported ophthalmic complications (6). However, our unique case demonstrates that corneal involvement is also a possibility.

V. Treatment, management

1) Treatment and response to treatment: For a Bell’s palsy that has been associated with a herpes zoster infection, 800mg of acyclovir five times a day for 7-10 days is recommended in addition to 60mg of oral prednisone a day for four days (4). With that being said, the vesicular outbreak often occurs days after onset of facial weakness making it difficult to determine what caused the initial palsy. One study found that only 14% of Ramsay Hunt patients developed vesicles after the facial nerve involvement making it originally indistinguishable from a basic Bell’s palsy (5). Timely administration of oral acyclovir and
Prednisone has been shown to improve recovery chances in Ramsay Hunt patients. If administered within the first three days of the palsy, patients have a 75% chance of full facial nerve recovery (7). If treatment is initiated between day 4 and 7 patients have a 48% chance of full recovery, and after 7 days, the chance of complete recovery falls to 30% (6).

2) Research/Literature review/ Bibliography: References are attached at the end of the outline and the findings in research and review of the literature are mentioned in the discussion and treatment sections of this outline.

VI. Conclusion

1) In addition to extensive cranial nerve testing, a patient who has an acute Bell’s palsy should be screened for vesicular eruption in the ear canal and/or pharynx to rule out Ramsay Hunt syndrome. Furthermore, the proper administration of acyclovir and prednisolone is paramount in achieving the best facial nerve recovery.

2) With close monitoring of our patient, we were able to determine the true underlying cause of the Bell’s palsy and initiate proper treatment. Optometrists should consider timely follow up appointments with their Bell’s palsy patients to detect such serious conditions as Ramsay Hunt. Co-management with ENT should be considered with these patients.

3) While ocular findings with Ramsay Hunt syndrome are rare, optometrists should be aware of the wide variety of serious ocular presentations. Our case demonstrates that corneal findings are a possible ophthalmic complication in Ramsay Hunt syndrome and respond to timely treatment.

References

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