

Abstract

Red ear syndrome is a very rare neurological disease that since first described in 1994 there is about 100 cases that has been reported. We are presenting a case of Red Ear syndrome that was associated with migraine and was very challenging in treatment.

Case

33 y/o M Adopted Right handed with PMH of depression, Asperger syndrome, gout, ADHD and scoliosis who is following in the neurology clinic for Headache which started 13 years ago, located "all around", most pronounced in the bilateral front parietal regions and feels like a "vice grip", used to occur 2 times a month when they started 10 years ago, then became daily about 6 years ago. They last anywhere from 15 min to hours. Throbbing in nature with some tightness, start in the back of the head more on the LT than the right, has photophobia, some mild phono phobia, no nausea, vomiting, flashing lights. Sometimes he gets tingling down his RT arm. 4 years later the patient started having redness and burning of the external ears of fluctuating severity, present throughout the day and are severe which per the patient words " it feels hot and burns like someone is injecting hot lava into the veins". That he frequently has to apply cold water and rub lidocaine gel to his external ears to relieve these symptoms. He also experiences "hot flashes" as well as flushing in the face and increase in baseline hyperhidrosis. 2 months before presentation patient has also been having bilateral L>R TMJ pain. Rheumatology ROS is specifically negative for sicca, eye dryness, OU, Raynaud's, photosensitivity, parotid or lacrimal gland swelling, abdominal pain or nausea, vomiting, diarrhea or constipation. General Physical exam was unremarkable except for redness of both ears as shown in (figure 1) and sometimes intermittent face rashes. Informed Consent was taken from the Patient. Per the IRB regulations of the hospital at University of Chiago, no need to take IRB approval for submitting a case report. The work up was very extensive including positive Cannabinoids +ve, Anti nuclear IFA borderline +ve 40 (normal range <40), Am cortisol 6.1 (normal range 6.2) & Fatty acid Profile is Low but the rest of the work up was unremarkable including CBC and CMP, CRP & ESR, ACE , TSH, T3 and T4, Urine catecholamine, Thyroglobulin AB, Thyroid peroxidase Ab, Vitamin b12, A and E , Zinc, Endomysial Ab, C1 esterase inhibitor ab, Chromogranin A, ANCA Panel for vasculitis (Myeloperoxidase Ab and proteinase 3 ab), Calprotectin, Uric acid, ANA, ANCA, Anti Ds DNA, C3 & C4, Cryoglobulin, Tissue transglutaminase IGA and IGG, Immunoglobulin A, M, G, Endomyosial Ab, FFTYC ab to collagen II, Hepatitis B, C, Syphilis, Quanteferon gold test, ACTH, Calcitonin and RF. Also the MRI brain and whole spine was unremarkable, Cardiac CT and Pulmonary function testing.

The patient also was tried a lot of medications with no much improvement including Lexapro, Tegretol, Diazepam, Clonazepam, Topiramate, Ketorolac, Propranolol, Indomethacin, Verapamil, Prednisone, Amitriptyline, levoceterizine, Desoximetasone, Topical Motemaone and Depakote. Also tried Cold water application, Vaseline application and was finally prescribed Medical Marijuana transiently. Then he was diagnosed with migraine and red ear syndrome. For the Migraine, He has been taking gabapentin for years which on(12/2017) increased to 900mg bid with minimal response. Tried botox injections on 3/2/2018 which only helped for the 1st 3 doses then after that he has changed his neurologist and started to notice less response to Botox and started to take Rizatriptan then shifted to Amiovig 140 mg monthly with very good improvement. For the Ear pain treatment he has the following Procedures:

1. Bilateral Sphenopalatine ganglion block 2018 s/p 100% relief for few days
2. LT Pterygopalatine angle block 2019 >75 % improvement for 3 month
3. Gasserian Nerve Block 2020 > 75% improvement for few days
4. Bilateral Sphenopalatine ganglion block 2022 s/p 90-95% relief for 3month and 27 days
5. Bilateral Sphenopalatine ganglion block 3/2023 s/p 100% relief to date



Discussion

- Red Ear Syndrome (RES) is a Unilateral/bilateral erythema of the ear with associated pain and burning sensation where the pain may radiate to the mandible, cheek, and occiput from the pinna.
- The attacks can last for seconds to hours but commonly occur for 30 min to an hour. It was 1st described by Lance in 1994 and since then about 100 cases has been reported.
- The pathophysiology of the RES is either brainstem trigemino autonomic circuit dysregulation theory where there is dysregulation between the connections between the facial parasympathetic fibers and the trigeminal nerve excitatory fibers vs thalamic syndrome resulting into disinhibition of the trigemino thalamic neurons leading into pain vs antidromic irritative discharges from C3 root vs Temporo mandibular joint dysfunction as an example of angry back firing c nociceptors.
- There are 2 types of RES, either primary "idiopathic" usually occurs with migraine or secondary usually occurs in older population, has a trigger and not usually associated with migraine.
- On Diagnostic criteria it is a diagnosis of exclusion.
- Treatment is a challenge as there is no standardized treatment but usually in primary RES gabapentin is the drug of choice and the other options include C3 block, greater auricular nerve block, verapamil, NSAIDs, indomethacin, Triptans and sphenopalatine ganglion block which was the treatment of choice for our patient.

References

1. Lance JW. The red ear syndrome. Neurology. 1996;47:617–20.
2. Hirsch AR. Red ear syndrome [comment]. Neurology. 1997;49:1190.
3. Arjona A, Fernández-Romero E. Red ear syndrome: a new case. Rev Clin Esp. 2000;200:704–5.
4. Purdy RA, Dodick DW. Red ear syndrome. Curr Pain Headache Rep. 2007;11:313–6.
5. Velasco EM, Luis Lopez Mesonero L, Pedraza Hueso MI, Ruiz Pi ñero M, de Lera Alfonso M, Guerrero Peral AL. Red ear and more: facial and extrafacial erythema accompanying migraine attacks. Headache. 2016;56:174–75.
6. Barragan-Martinez D, Urbanos-Nunez A, Martinez-Salio A. Red ear syndrome: a case report with good therapeutic response. Rev Neurol. 2015;61:239–40.