

Case Reports

Sympathetic neural hyperalgesia edema syndrome as a cause of autoimmune hearing loss

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Summary

Purpose: To describe another previous unreported manifestation of the sympathetic neural hyperalgesia edema syndrome – autoimmune hearing loss. **Materials and Methods:** Dextroamphetamine sulfate 30 mg extended release capsules was given to a woman to try to help her conceive since, with her pelvic pain, chronic fatigue syndrome, and diarrhea, it was thought that an inflammatory condition related to permeation of unwanted chemicals into endometrial tissue related to the sympathetic neural hyperalgesia edema syndrome could be inhibiting her failure from conceiving despite three cycles of embryo transfer. **Results:** Not only did the symptoms mentioned above disappear, but she also noted marked improvement of hearing loss that had been present for several years. The improvement in hearing was documented by audiology tests and had not responded to many months of 15 mg/day prednisone. **Conclusions:** Autoimmune hearing loss (diagnosis established by her ear nose and throat specialist) should be added to the long list of manifestation of the sympathetic neural hyperalgesia edema syndrome.

Key words: Autoimmune hearing loss; Autoimmune inner ear disease; Sympathetic neural hyperalgesia edema syndrome; Pelvic pain; Chronic fatigue.

Introduction

A recent editorial suggested that dextroamphetamine sulfate may be the single most effective treatment for a large variety of disorders especially prevalent in women [1]. Probably the most common use for the amphetamine by the gynecologist is pelvic pain, but the editorial mentioned a large variety of conditions both specific to women, more common to women, or also present in men [1-4]. It is known as the sympathetic neural hyperalgesia edema syndrome [5, 6].

The message from the editorial was that it is the gynecologist who is the specialist most familiar with the syndrome. More and more women are using the gynecologist as their primary physician. Thus, when symptoms present that seem outside the scope of the gynecologist, the patient expects the gynecologist to refer her to the proper specialist.

Unfortunately most specialists will not be nearly as familiar with the sympathetic neural hyperalgesia edema syndrome as the gynecologist, and thus may subject the patient to a multitude of invasive tests and risk medical or surgical treatments that frequently prove inadequate when simple safe treatment with dextroamphetamine sulfate could have been given. Thus the gynecologist should become familiar with all of the various manifestations of the neural hyperalgesia edema syndrome.

The purpose of this present case report was to describe a new manifestation of the sympathetic neural hyperalgesia syndrome. Unfortunately the name which refers to pain and swelling does not fully describe all of the permeability disorders related to this syndrome. This particular patient had a variety of other symptoms, e.g., dysmenorrhea, dyspareunia, diarrhea, and chronic fatigue syndrome that also improved following dextroamphetamine sulfate.

The patient did successfully conceive at a later time but probably not related to her treatment with sympathomimetic amines. Her interesting case of how she conceived and the important message that comes with her successful pregnancy will be reported at another time.

Case Report

The patient presented at age 41 for three years of infertility. Her past medical history included autoimmune hearing loss, Hashimoto's thyroiditis, mitral valve prolapse, and endometriosis. She also complained of lightheadedness, headaches, palpitations, pelvic pain, fatigue, and episodes of diarrhea. She was taking L-thyroxine 75 mcg/daily and 15 mg prednisone daily.

After three failed in vitro fertilization (IVF) cycles, the sympathomimetic amine dextroamphetamine sulfate 15 mg daily was added to her regimen in an effort to attempt to improve implantation. Within three months, the dosage had been slowly increased

to 30 mg daily. At this point, the woman noted that, while she had not initially noticed hearing improvement, when she went off the dextroamphetamine sulfate for acute appendicitis and had an emergency appendectomy the previous month, she could tell that her hearing deteriorated. She then noticed hearing improvement upon restarting the medication. She subsequently went for an official hearing test, which showed documented hearing improvement since starting dextroamphetamine sulfate.

Four months later, the woman noted a two-kg weight loss, decrease in headache frequency, decreased palpitations, decreased diarrhea, decreased pelvic pain and pain with intercourse, and an increase in overall energy.

The woman had never noted any improvement with prednisone. With the marked improvement in hearing with the dextroamphetamine sulfate, she stopped the prednisone. She has remained on dextroamphetamine sulfate for four years and she has not had any deterioration in her hearing.

Discussion

Autoimmune inner ear disease (AIED) is a rare disease marked by rapidly progressive, often fluctuating, bilateral sensorineural hearing loss over a period of weeks to months. Occasionally only one ear is affected initially, but in 79% of patients of patients the disease will eventually progress to bilateral hearing loss [7]. Vestibular symptoms, such as generalized imbalance, motion intolerance, and vertigo are concurrently present in about half of patients [7]. Autoimmune inner ear disease accounts for less than one percent of all cases of hearing impairment of dizziness and is more common in females. Diagnosis may be overlooked due to lack of a specific diagnostic test, but is generally based on clinical criteria, a positive response to steroid treatment, as it is reversible with immunosuppression, or evidence from broader tests of autoimmunity [7, 8].

Systemic autoimmune disease coexists in 15-30% of patients [7]. Autoimmune inner ear disease can be broken down into two forms. Primary AIED exists in the absence of systemic autoimmune disease and is inner ear-specific. Secondary AIED is thought to be the consequence of systemic immune abnormalities that also involve the inner ear [8]. The woman had been treated with glucocorticoids but did not respond. Immunosuppressants were discussed but she declined because of desire to conceive.

The name sympathetic neural hyperalgesia edema syndrome seems to be related to certain tissues being more susceptible to permeability by toxic chemicals, coupled with hypofunction of the sympathetic nervous system (which is the main neurologic control of cellular permeability). This is the basis of many autoimmune disorders or allergic reactions leading to hives, rheumatoid arthritis, or other "autoimmune disorders" [1-3, 9-11]. One young woman "dying" from autoimmune hepatitis immediately responded to dextroamphetamine sulfate and is now perfectly normal [12]. The woman's life was saved merely by the fortuitous conversation she had with a gynecologist for whom she was filming a YouTube presentation [12]. Thus autoimmune hearing loss should be added to the long list of chronic disorders that all

respond to dextroamphetamine sulfate. The long-term use of dextroamphetamine has not been associated with any health deterioration, but without the gynecologist intervention the woman would have been subjected to the risks of long-term of steroids and/or immunosuppression without anywhere near the same improvement in hearing. Most of the literature about this syndrome has been in gynecological journals, and without a pharmaceutical company advocating the drug, it is not likely that the knowledge of the benefits of dextroamphetamine sulfate will be shared by other specialists. Thus, it became the responsibility for the gynecologist, the primary caregiver for women, to be aware of all manifestations of this disorder.

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