Hearing Loss and Teprotumumab

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Background: Thyroid eye disease (TED) is an unpredictable autoimmune inflammatory disease which can be sight-threatening, debilitating, and disfiguring. Teprotumumab (IV infusion every 3 weeks x 8 doses) was recently approved as the first and only FDA-approved drug for TED in 2020. Phase 2 and 3 studies showed significant improvement in proptosis, double vision, soft tissue inflammation and quality of life for patients with active moderate to severe TED. Side effects were experienced by 85% of patients on teprotumumab. Hearing loss symptoms were reported in 10% of patients and were reported to be reversible upon stopping the drug.

Objective: To explore the incidence of hearing loss symptoms and sensorineural hearing loss in patients treated with teprotumumab. Methods: All patients, followed at one institution, treated with at least 4 infusions of teprotumumab were evaluated. Charts were evaluated for baseline hearing symptoms and hearing symptoms during or after therapy with teprotumumab. Those patients with hearing symptoms were referred for audiogram testing and patulous eustachian tube (PET) testing.

Results: Twenty-eight patients were included in this analysis. Thirteen patients (46%) complained of hearing symptoms. The most common symptoms were autophony or an ear plugging sensation and hearing loss or muffled hearing. Hearing symptoms developed after a mean of 3.6 infusions. Of the patients with hearing symptoms, three patients (23%) had sensorineural hearing loss documented on audiogram (n=2) or patulous eustachian tube (n=1) documented on PET testing. To date, the patient with PET has experienced some improvement, but not resolution, of her symptoms. The two patients with documented sensorineural hearing loss have not experienced a significant improvement in hearing, on audiogram, on average 3 months after stopping teprotumumab.

Conclusion: Teprotumumab is a promising new therapy for active moderate to severe thyroid eye disease. Providers should consider performing a baseline audiogram with PET testing and performing audiograms with PET testing for patients that develop hearing symptoms during or after therapy. Hearing loss is a concerning adverse event and its mechanism and reversibility should be further studied.