Bilateral cochlear enhancement in Cogan syndrome

A 43-year-old man presented for neurologic evaluation with acute bilateral hearing loss. Two weeks before presentation, he had bilateral eye pain diagnosed as scleritis. He also noted a subacute history of fevers, night sweats, and diarrhea. Neurologic examination revealed bilateral sensorineural hearing loss, confirmed with audiology. MRI demonstrated enhancement of the cochlea bilaterally (figure). Extensive evaluations for rheumatologic and infectious etiologies were negative.

Cogan syndrome is a rare systemic disease classified among the vasculitides characterized by audiovestibular and ocular involvement. The patient stabilized with oral prednisone. If refractory, treatment with steroid-sparing immunosuppressants or cochlear implants can be considered.

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Disclosure: The authors report no disclosures.

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Neurology 2009;73;75
DOI 10.1212/WNL.0b013e3181aaea6c

This information is current as of June 29, 2009