International Journal of Pediatric Otorhinolaryngology 107 (2018) 160–163

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Case Report

Granulomatosis with polyangiitis presenting as facial nerve palsy in a teenager

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ARTICLE INFO

Keywords:
Granulomatosis with polyangiitis
GPA
Wegener's
Facial nerve palsy

ABSTRACT

Granulomatosis with polyangiitis (GPA, previously known as Wegener's granulomatosis) is an autoimmune systemic small-vessel vasculitis, associated with the presence of anti-neurophil cytoplasmic antibodies with a cytoplasmic staining pattern (c-ANCA). It is characterized by necrotizing granulomas, usually affecting the airways and kidneys. GPA should be considered when patients do not improve despite adequate treatment of otologic symptoms, when patients have unspecific symptoms suggesting systemic disease (e.g. fever, malaise), or when other organs are involved (kidney, lungs, etc.). We present an interesting case of a 14-year-old female with eight-weeks of bilateral otalgia, unilateral facial nerve palsy, decreased appetite, and fatigue refractory to steroid, anti-viral, and antibiotic treatment ultimately diagnosed with GPA.

Case Presentation: A 14-year-old female presented to our emergency room with eight-weeks of bilateral otalgia, unilateral facial nerve palsy, decreased appetite, and fatigue. She was initially treated by her primary care physician with ciprodex for presumed otitis externa but returned with right facial swelling and paresthesia. She was subsequently diagnosed with Bell's palsy and treated with a course of prednisone; House-Brackmann score was not documented but there was incomplete eye closure per the family. Over eight-weeks, she completed 3 courses of prednisone (10mg BID), valacyclovir, and ciprofloxacin prescribed by her primary care physician. At first, she completely resolved; however, paralysis recurred and by the third course she had persistent incomplete eye closure. Two days prior to admission, she developed increased ear pain, right neck pain and new-onset severe back pain. On review of systems, she reported fever, appetite change, decreased activity, shortness-of-breath, cough, emesis, bilateral ear pain and pressure, with decreased right-sided hearing. On physical exam, extraocular movements were intact, pupils were equal, round and reactive to light, she had normal neck range of motion, strong pulses, and no respiratory distress. Ear evaluation revealed no proptosis, swelling, erythema, or mastoid tenderness; she had bilateral acute otitis media (OM) with thickened tympanic membranes. When she presented to us, her facial paralysis was scored as a House-Brackmann 5/6 (unable to wrinkle her right forehead or completely close her right eye with flattening of her right nasolabial fold). The patient received an inpatient audiogram after admission. Bilateral ears revealed moderate to severe conductive hearing loss in the low frequencies and a mixed hearing loss from 2000 to 4000 Hz with the right ear slightly worse than the left ear. Bilateral tympanometry was flat with no peak. Bilateral DPOAE (2000–8000Hz) were absent at all test frequencies.

Her white count was elevated at 17.7 K/mcl with 86% segmented, and a C-reactive protein was elevated at 9mg/dl. She was started on empiric ceftriaxone. Her chest x-ray revealed multiple, bilateral pulmonary nodules (Fig. 1A), while MRI of the head (Fig. 2) and CT of the temporal bones (Fig. 3) revealed a diffuse inflammatory/infectious process with extensive opacification of the middle ears and mastoid air cells.

Chest CT showed well-defined nodules throughout both lungs including two nodules with small foci of central air, suggesting cavitation. (Fig. 1B). Doppler ultrasound of the neck, CT neck, and echocardiogram were otherwise unremarkable; there was no evidence of sigmoid/tranverse sinus or jugular vein thrombosis. The infectious disease workup for aerobic, anaerobic, acid-fast bacteria, histoplasmosis, and fungi were negative; however, the preliminary percutaneous lung biopsy results suggested an infectious etiology. Due to opacification of

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https://doi.org/10.1016/j.ijporl.2018.02.009
Received 5 December 2017; Received in revised form 1 February 2018; Accepted 3 February 2018
Available online 07 February 2018
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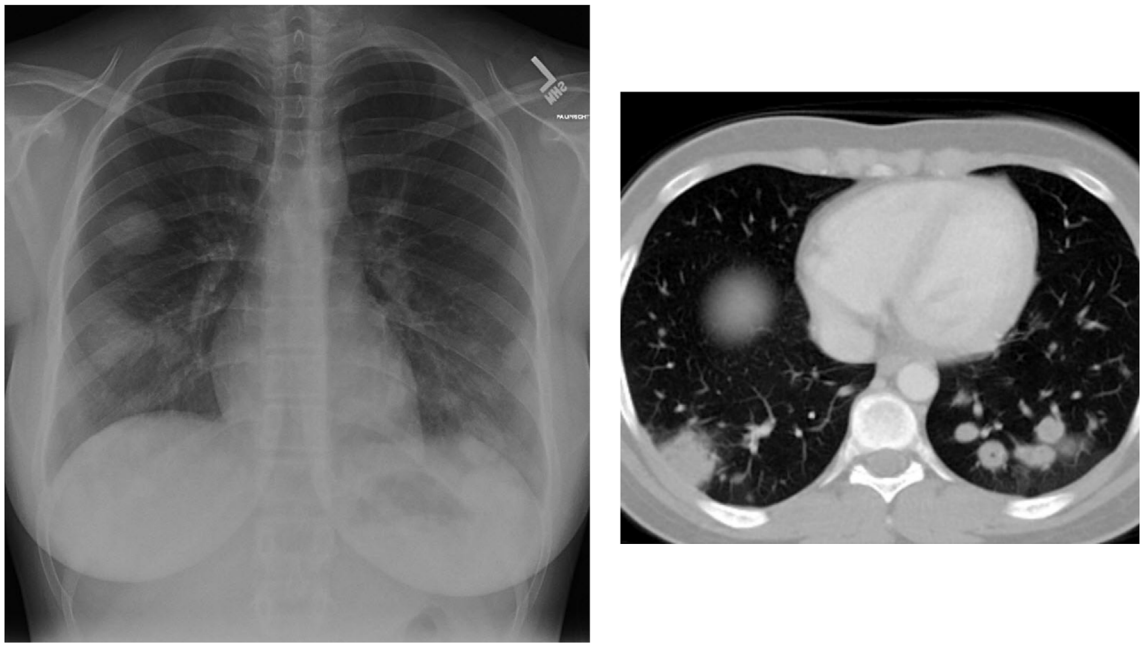

Fig. 1. A. Chest x-ray and **1 B.** axial postcontrast CT lung window image show multiple, bilateral nodules concerning for septic emboli, fungal disease or less likely metastatic disease.

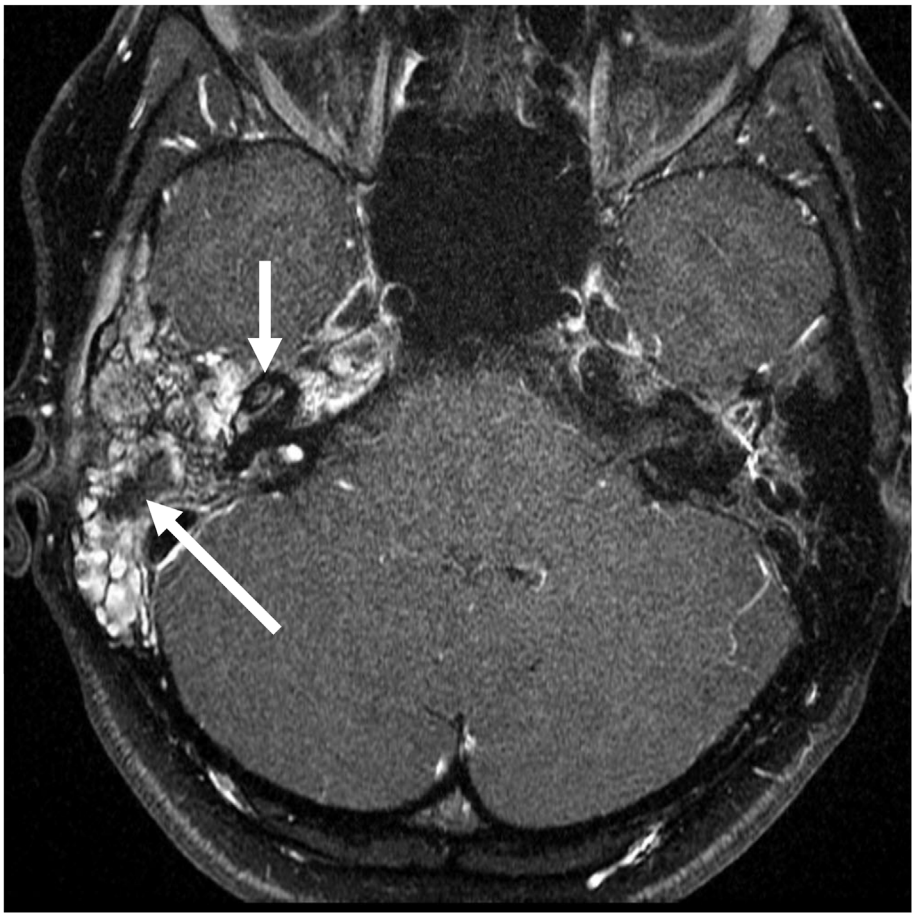

Fig. 2. Axial post-contrast fat-saturated MRI demonstrates extensive disease involving the right greater than left mastoid air cells, middle ear cavities and right cochlea **(short arrow)**. The majority of the disease enhances, consistent with inflammatory otomastoiditis and labyrinthitis. Non-enhancing focus in the right mastoid **(long arrow)** demonstrated restricted diffusion (not shown) suggesting focal area of frank pus or cholesteatoma.

the middle ears and mastoids, the patient was brought to the operating room for bilateral tympanostomy tube placement and intraoperative ear cultures, which revealed very few Gram-positive cocci in pairs and clusters, and few Gram-positive rods. She did not improve and returned to the operating room for tympanomastoidectomy with removal of spongy yellow-white tissue with significant granulation tissue filling the mastoid and middle ear. During her hospital course, she also developed gingival swelling and multiple urinanalyses that revealed moderate to