**Abstract**

Wegener’s granulomatosis is a systemic vasculitis with known manifestations in the head and neck. This case report aims to present a very rare instance of otitis media with facial nerve paralysis and sinusitis with subperiosteal abscess of the orbit as an initial presentation of Wegener’s granulomatosis. The otolaryngologist should maintain a high index of suspicion for autoimmune disease in these diagnostically challenging cases.

**Case Presentation**

An 18 y.o. female presented to the ED with a 3 week history of left ear pain and left retroorbital pain. She had been seen by outpatient ophthalmology who noted a normal exam with normal IOP. She was treated with oral antibiotics, however symptoms worsened to include left-sided proptosis, facial pain, nasal congestion, and sore throat. CT scan (Figure 1) demonstrated left pansinusitis with invasion of the lamina papyracea. Nasal endoscopy demonstrated purulence draining from the middle meatus, and diffusely edematous, erythematous mucosa. The left ear demonstrated serous effusion, but no perforation. She was treated with nasal sprays, IV steroids, and IV Unasyn. She demonstrated clinical and radiological improvement over the next 24 hours. All cultures were negative. The patient was sent home with oral Levaquin, a Medrol dose pack, and nose sprays.

An in-office myringotomy was performed the next day for the persistent middle ear effusion. After serous fluid was suctioned, granulation tissue within the middle ear space was noted. Three days later, she presented to the ED with left eye proptosis and complete left facial paralysis. A repeat CT scan demonstrated progression of the left orbital subperiosteal abscess(Figure 2). She then underwent left anterior ethmoidectomy, maxillary antrostomy, and sphenoidotomy with findings of purulent drainage and nasal polyposis. She also underwent left medial orbital wall decompression and drainage of subperiosteal abscess. An audiogram demonstrated a left moderate sloping to severe mixed hearing loss with a 30db conductive component over 3 frequencies (Figure 3). Two days later facial pain and pressure were improving, and she was discharged on her previous antibiotic regimen.

**Case Presentation (cont’d)**

Six days later, the patient was readmitted for worsening left eye proptosis and pain. CT scan demonstrated recurrent subperiosteal abscess (Figure 4). She was brought back to the OR for revision total ethmoidectomy, sphenoidotomy, and orbitotomy with biopsy of periorbita. The exam was notable for diffuse fibrinous tissue in the nasal cavity, abnormally thickened and inflamed periorbita, and congealed underlying orbital fat. The medial rectus was extremely inflamed. No discrete abscess was identified.

Due to the negative cultures and abnormal intraoperative findings, the rhinological service was consulted. Subsequent laboratory values were positive for c-ANCA, PR3, and rheumatoid factor. Both CRP and ESR were elevated. A CT of the chest demonstrated several cavitation and non-cavitations nodules, as well as perihilar lymphadenopathy. A right upper lobe wedge resection was then performed, and the pathology demonstrated findings consistent with Wegener’s granulomatosis. This was helpful given the non-specific findings from the nasal tissue biopsies (Figure 5)

The patient was managed with weekly Rituximab treatments, as well IV steroids. All symptoms resolved over the course of 1.5 months.

**Discussion**

Wegener’s granulomatosis, or granulomatosis with polyangiitis, is an autoimmune vasculitis. It typically presents with a classic triad of involvement of the upper respiratory tract, lower respiratory tract, and kidneys. Its incidence is 1-3 in 100,000, and has no gender predilection (1). Head and neck manifestations account for the presenting symptoms in 73% of patients ultimately diagnosed. The eye is the most common site for initial presentation of the disease in the head and neck, present in up to 19% of patients (2). Serous otitis media is the most common otologic manifestation, and is often secondary to eustachian tube dysfunction. Suppurative otitis media results from mucosal involvement of the middle ear and mastoid air cells, with facial palsy resulting in 8-10% of cases. Inner ear manifestations are common in later stages, with SNHL present in 35% of patients secondary to vasculitis of cochlear blood supply and immune complexes in cochlea (3).

Initial sinonasal manifestations range from non-specific mucosal changes and edema to necrosis of the turbinates and septum, ultimately resulting in saddle nose deformity. Chronic sinusitis is not typical at initial presentation. Oral manifestations are very rare and may include ulcerations, or evidence of ‘strawberry’ gingival hyperplasia. Subglottic stenosis is present in 16% of patients, and is five times more frequent when disease presents in childhood.

The most common ocular manifestations of the disease are orbital pseudotumor, episcleritis, and ulcerative keratitis which occur in 15% of patients. The presence of c-ANCA on lab values is typical in several autoimmune disorders, however the presence of antibodies to the neutrophil granule component PR3 is pathognomonic for Wegener’s. Definitive biopsy characteristics include granulomatous inflammation, palisading granulomas and scattered giant cells. Vasculitis and/or necrotizing granulomatous may also be see. Biopsies are non-diagnostic in 50% of specimens, and often repeat biopsies from different sites are required for diagnosis.

**Conclusions**

Wegener’s granulomatosis often presents with head and neck manifestations at some point during the disease course. Initial presentation of the disease with refractory chronic sinusalitis, subperiosteal abscess, and serous otitis media with facial paralysis has not been described in the literature. Otolaryngologists should be familiar with all possible manifestations of the disease in the head and neck, and demonstrate appropriate levels of suspicion in these cases.

**References**