CLINICAL VIGNETTE

Auricular Chondritis as a First Feature of Granulomatosis with Polyangiitis

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A 40-year-old Caucasian female presented to rheumatology for evaluation of recurrent inflammation of the auricles. Prior to evaluation, the patient's only past medical history was intermittent migraine headaches. However, for the past several months, patient had been presenting to her local primary care physician and urgent care with external ear redness and swelling. Several different courses of antibiotics had been prescribed to the patient, including Clindamycin, Levofloxacin, and lastly Sulfamethoxazole/trimethoprim. She first presented to the UCLA health system as a transfer from her local hospital for suspicion of Stevens-Johnson syndrome and Toxic Epidermal Necrolysis (TEN) following sulfamethoxazole/trimethoprim. She was admitted to the ICU, with biopsy confirmation of TEN. She received one dose of etanercept subcutaneously, as well as aggressive wound care. Her hospital course dramatically improved after etanercept, and patient was discharged within the week.

At her first outpatient dermatology follow-up appointment, her prior history of ear swelling prompted a referral to rheumatology. Consultation confirmed recurrent episodes of auricular swelling, as well as a history of intermittent sinusitis. Laboratory evaluation found high titers of p-ANCA/myeloperoxidase antibodies. Biopsy of the auricle revealed pathological features consistent with chondritis, specifically mild perichondral inflammation and superficial and deep perivascular and peri-adnexal lymphoplasmacytic inflammatory infiltrate with scattered eosinophils. Additional labs for chondritis included proteinuria, in the setting of a normal creatinine. Subsequent kidney biopsy was consistent with pauci-immune glomerulonephritis consistent with ANCA vasculitis. The patient received six rounds of cyclophosphamide and corticosteroids as treatment for glomerulonephritis. Currently, she is on a steroid sparing regimen of azathioprine with normal urine protein levels.

Granulomatosis with Polyangiitis (GPA, formerly known as Wegener's Granulomatosis) is a systemic vasculitic condition that is part of a group of conditions called AAV (ANCA-associated vasculitis). The other conditions within AAV include Churg-Strauss Vasculitis, and Microscopic Polyangiitis, which are linked by overlapping pathology, small and medium vessel involvement, and the presence of ANCA antibodies.[1] Common initial manifestations of GPA often include renal, pulmonary, or ENT involvement. However, it is important to be aware of atypical initial manifestations, such as recurrent chondritis of the auricles, which was detailed in this case presentation. Once the diagnosis is established, it is important to screen for end organ damaging conditions including necrotizing glomerulonephritis. As in our case, normal creatinine does not exclude renal involvement.

Approximately 90% of patients with GPA develop upper airway or ear abnormalities.[2] Early nasal symptoms include nasal pain, rhinitis, epistaxis, and brown or bloody crusts. The inflammation of nasal cartilage can lead to septal erosion, septal perforation, or nasal bridge collapse, which can then lead to a saddle nose deformity. Ear disease in GPA most commonly involves either conductive or sensorineural hearing loss. Conductive hearing loss may be due to Eustachian abnormalities, while sensorineural hearing loss occurs in the setting of inner ear disease. The mechanism of action of inner ear disease in GPA is still poorly understood.

Auricular chondritis is a primary feature of the condition relapsing polychondritis (RPC). 25% to 35% of patients with RPC have a concurrent rheumatic disease, including AAV.[3,4] Evaluation with lab markers, as well as clinical and histological manifestations, occur with overlapping rheumatic conditions. ANCA antibodies can be detected in relapsing polychondritis, although the percentage and pattern of staining of the antibody (perinuclear "p-ANCA" vs cytoplasmic "c-ANCA" staining) is still not clear. One study found 24% of patients (8/33) with primary relapsing polychondritis to be ANCA positive.[5] This data was challenged by a study showing ANCA reactivity in three of six patients in whom RPC appeared in conjunction with a primary systemic vasculitis (specifically GPA and microscopic polyangiitis).[6]

Still, the ability of changes in ANCA titers to predict disease flares is poor. In addition to auricular chondritis, some of the shared clinical features of GPA and RP are laryngotracheal bronchial disease, necrotizing glomerulonephritis, episcleritis, and saddle nose deformity. Histologically, AAV are distinguished from other systemic small-vessel vasculitic conditions by the absence of immune deposits. In the case discussed, patient had a kidney biopsy confirming pauci-immune disease.

GPA and Relapsing polychondritis are two conditions that frequently overlap in their clinical symptoms and pathologic data. The overlap can be difficult to manage and challenging to categorize patients. Checking for ANCA antibodies is part of the work up of auricular chondritis, whether it is due to primary relapsing polychondritis or secondary to a systemic vasculitic condition. Criteria classification for vasculitis are important in large studies, but not as crucial in the treatment of individual patients. In cases where patients share manifestations of both conditions, different treatment regimens are considered. In recurrent ear swelling and erythema, consider the evaluation of auricular chondritis and systemic rheumatic conditions.

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