Granulomatosis with polyangiitis: An example of diagnostic (confirmation) bias

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Abstract

58-year-old woman with a history of bilateral mastoiditis presented with worsening malaise, headache, and mastoid tenderness concerning for either vasculitis or an infection. Given her history, elevated inflammatory markers, and nuclear medicine bone scan concerning for osteomyelitis, she was started on empiric antibiotic therapy and underwent an extensive infectious workup. After the workup failed to reveal an infectious etiology and her condition worsened despite treatment, the team reconsidered vasculitis. An elevated c-ANCA titer, PR3 >30, a urinalysis without RBC casts, and a computerized tomography without pulmonary nodules confirmed the diagnosis of granulomatosis with polyangiitis with limited involvement.

This case highlighted the impact of diagnostic (confirmation) bias in clinical decision-making as the clinical history and imaging findings led to an extensive infectious workup despite negative microbiology.

Keywords: Granulomatosis with polyangiitis, Case report, Confirmation Bias

Background

Granulomatosis with polyangiitis (GPA) is a rare auto-immune small-vessel vasculitis that classically presents with upper and lower respiratory tract symptoms and renal involvement.1

**CLINICAL CRITERIA**
- Nasal involvement: bloody discharge, ulceration, nasal congestion, blockage, or nasal septum perforation
- Cervical or peribronchial inflammation or pain on swallowing, hoarseness, voice or asthma, endobronchial involvement, or saddle nose deformity
- Conductive or mixed hearing loss

**LABORATORY, IMAGING, AND BIOPSY CRITERIA**
- Positron-emission tomography (PET) scan, enhanced computed tomography (CT), or magnetic resonance imaging (MRI)
- Granuloma, extravascular granulomatous inflammation, or giant cells or biopsy
- Inflammation, congestion, or obstruction of the nasal/stem/tent (nose) or intranasal imaging
- Petrous temporal bone, mastoid, or calvarial involvement
- Infectious or inflammatory granulomatous inflammatory disease, giant cells or biopsy

Case Presentation

13/20/22 • 58-year-old Spanish-speaking woman with several months of smoldering ear symptoms suspected to be an infection but refractory to several rounds of antibiotics and steroids.

06/11/22 • Admitted for bilateral mastoiditis and facial nerve paralysis. Underwent bilateral mastoidectomies with facial nerve decompression, tympanotomy, and treatment with antibiotics despite extensive negative infectious workup.

08/25/22 • Persistent boring mastoid pain and significant hearing loss causes concern for osteomyelitis. Started on antibiotics and a nuclear medicine bone scan was ordered.

09/03/22 • Presented to the ED with malaise, headaches, mastoid tenderness, and neck pain.

09/04/20 • NM bone scan was concerning for osteomyelitis of the left temporal bone. Diagnostic differential included infection vs. GPA/GCA.

09/05/22 • Chest x-ray without pulmonary nodules and urinalysis without RBC casts decreased concern for GPA. Primary concern for osteomyelitis given the clinical history, elevated inflammatory markers, bone scan, CT, and UA findings. The patient was started on empiric antibiotic therapy and underwent an extensive infectious workup.

09/04/23 • MRI brain demonstrates nonspecific fluid and enhancement in mastoidectomy beds representing either granulation vs. infection.

09/08/22 • Despite treatment with antibiotics, progressing condition and extensive negative infectious workup, the treating team continued to pursue an osteomyelitis.

09/07/22 • CT scan of the patient’s chest showed no evidence of pulmonary nodules.

09/09/20 • Treatment with high-dose IV steroids and Rituximab infusions with good effect.

09/14/20 • Patient received her first Rituximab infusion. The diagnosis of AAV with limited involvement was made with a c-ANCA of 1:12560, PR3 >30, UA without RBC casts, and CT without pulmonary nodules.

08/10/21 • Tolerating therapy well with no signs of active disease.

Discussion

This case is noteworthy for the initial delay in diagnosis that was likely due to confirmation bias.

Diagnostic (or confirmation) bias is the tendency to give more weight to findings that support a preliminary diagnosis while failing to seek out or minimize contradictory evidence.4

In this case, treatment and investigations primarily focused on the management of infection due to the presumed clinical history and results of the NM bone scan. Despite the progression of the patient's condition despite empiric antibiotic workup and an extensive negative infectious workup, the treating team continued to pursue an infectious etiology as a result of diagnostic bias. It was not until the patient's condition had significantly progressed that the team broadened their differential and was able to make the diagnosis of GPA.

Several approaches could have been used to counteract bias in this case.
1. Seeking contradictory evidence and broadening the diagnostic differential.
2. Utilizing metacognitive interventions or balanced testing.3

This case is useful for reflective purposes in avoid diagnostic bias as a timely diagnosis would have prevented significant patient morbidity.

Competing Interests

The authors declare no competing interests, nor any financial interests.

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