Wegener’s Granulomatosis Presenting as a Parapharyngeal Mass

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Abstract

Rarely, atypical head and neck pathologies are the presenting feature of patients found to have Wegner’s Granulomatosis (WG). We describe the hospital course of a patient with a parapharyngeal mass and discuss how a stepwise approach led to the diagnosis of WG. To our knowledge, this report is the first description of a patient presenting with a parapharyngeal mass causing superior cervical chain compression with simultaneous Lemierre’s Syndrome who was ultimately diagnosed with WG. We advocate early consideration of WG in patients with atypical head and neck presentations and demonstrate how an interdisciplinary team leads to optimal coordinated care.

Introduction

Wegener’s Granulomatosis (WG), also known as Granulomatosis with Polyangiitis (GPA), is an autoimmune vasculitis characterized primarily by inflammatory necrotizing granulomatous lesions within the upper and lower respiratory tract and kidneys.1 Head and neck manifestations of this disease are the presenting symptom in 80-95% of cases and may include sinonasal, otologic, pharyngeal, and laryngeal pathologies.2 Sinonasal involvement may range from non-specific symptoms such as nasal obstruction, discharge, and crusting to more classic WG symptomology such as septal perforation and saddle deformity. Otologic manifestations are also common, as oris media effusion is described frequently within the literature.2 Although WG most often manifests in the head and neck as one of the aforementioned entities, a growing body of literature reveals a new subset of disease presentation.

Otolaryngological literature has increasingly recognized WG as a worthy diagnostic consideration for patients presenting with atypical lesions of the head and neck with secondary cranial neuropathies.1,15 Moreover, such lesions are notoriously difficult to diagnose, with only 15-25% of all biopsies described as meeting the pathologic criteria of WG.1 This can create a prolonged diagnostic interval and favors an interdisciplinary approach to ensure proper management. Here we discuss one such case of Wegener’s Granulomatosis, emphasizing the difficulties faced in diagnosis, the need for a thorough history and physical exam, and the value of the interdisciplinary medical team.

Case Report

A 24-year-old Caucasian male with no past medical history presented to clinic with a two-month history of odynophagia, dysphagia, and weight loss. He had previously been evaluated by otorhinolaryngologists at three separate outside facilities. The first presentation resulted in outpatient management with oral antibiotics. The second presentation resulted in a right tonsillectomy with placement of bilateral pressure equalization tubes at an outside hospital. The third visit led to an inpatient admission with administration of intravenous antibiotics and steroids for a suspected parapharyngeal abscess. Despite three very different treatment strategies, his odynophagia worsened to the point of dehydration and culminated in a 30-pound weight loss. At this time, he also developed bilateral conjunctivitis, dysuria, and persistent polyarthralgias involving his knees, elbows, and ankles. The patient was admitted to Otolaryngology for further examination and workup.

On physical exam, he had bilateral conjunctival injection with right anisocoria. There were no other focal findings in the remainder of his head and neck examination. This included a very thorough oropharyngeal, sinonasal, and subglottic examination performed with flexible fiber-optic laryngoscopy (FFL). Laboratory values were as follows: white blood cells of 7,400/μL, hemoglobin of 10.9 g/dL, hematocrit of 32.1%, platelet count of 476,000/μL, sodium of 132mEq/L, and albumin of 2.6g/dL. Blood cultures showed no growth. A CT scan of the neck with contrast revealed a right parapharyngeal soft tissue infiltrate concerning for infection or neoplasm, but no discrete abscess was evident (Fig. 1). The patient was then admitted to the hospital and started on broad-spectrum antibiotics.

Discussion

At this point, due to his constellation of symptoms and broad differential diagnosis, a multidisciplinary team was assembled initially involving internal medicine, infectious disease, oncology, and ophthalmology.

On hospital day 3, the patient was taken to the operating room for a right neck exploration. Reactive adipose tissue and several reactive lymph nodes at the level of the skull base were removed and sent for pathologic and microbiologic analysis. An MRI of the orbit, face, and neck demonstrated a right jugular vein thrombophlebitis consistent with Lemierre’s Syndrome (Fig. 2).

During the workup for the patient’s dysuria, urinalysis revealed microscopic hematuria without evidence of infection. In addition, his serum creatinine increased from 0.7 to 3.28 mg/dL despite adequate hydration and appropriate medication dosing regimens. A subsequent renal ultrasound was inconclusive, but could not exclude intraparenchymal disease. Rheumatology and nephrology consults were then placed for workup of potential WG, yielding the following result: negative antineutrophil antibodies (ANA), negative anti-mitochondrial antibodies, anti-streptolysin O titer within normal limits, C3 complement within normal limits, decreased C4 complement, elevated IgE, positive rheumatoid factor, positive cytoplasmic anti-neutrophil cytoplasmic antibody (c-ANCA), and positive anti-proteinase 3 antibodies (anti-PR3).

Surgical pathology of the neck exploration returned as acute lymphadenitis with eosinophilia, eosinophilic myositis, and no evidence of malignancy in any sample. For definitive diagnosis of WG, the patient underwent a renal biopsy on hospital day 7, ultimately revealing focal necrotizing glomerulonephritis (non immune-complex mediated).

With a firm diagnosis of WG, the multi-disciplinary treatment team started the patient on daily oral prednisone and twice daily Azathioprine. The patient made immediate improvements in his health status, both subjectively and objectively. He was discharged on hospital day 12 and continues to receive outpatient care with otolaryngology, nephrology, and rheumatology services.

Similarity, testing for the standard laboratory markers can also come with mixed value. The most helpful markers for diagnosing WG are by far anti-nuclear antibody (ANA), cytoplasmic anti-neutrophil cytoplasmic antibody (c-ANCA), and anti-proteinase 3 antibodies (anti-PR3). Gottschlich et al notes that while c-ANCA and anti-PR3 antibodies are positive in 95% of patients demonstrating the generalized phase of WG, only 50% of patients will be positive during the localized phase.1

The patient discussed in the above case presentation was one whose laboratory results and history were suggestive of Wegner’s Granulomatosis, though certain elements of his clinical picture were inconsistent with the classic presentation of this disease. Specifically, he did not display nasal manifestations such as epistaxis or saddle nose deformity, and he had no tracheopulmonary symptoms or signs such as subglottic stenosis, hemoptysis and/or pulmonary nodules on X-Ray. This patient was part of a subset defined by an ever-growing body of literature, one that includes parapharyngeal and retropharyngeal manifestations with subsequent cranial neuropathies and/or superior cervical chain compression.1,5

Conclusions

Though head and neck manifestations are often the chief complaint of patients presenting with WG, the classic triad of sinonasal, pulmonary, and renal pathology is not always evident at initial presentation and will not always develop during the course of disease. As demonstrated by this case, WG should be included within the differential diagnosis of atypical head and neck lesions refractory to multiple treatment regimens. Moreover, the assembly of an interdisciplinary medical team leads to an expedited diagnosis, optimized inpatient management, and coordinated outpatient care.

References


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Figure 1. Computed Tomography Scan of Parapharyngeal Lesion
Computed tomography scan of the neck with contrast. Axial (A) and coronal (B) images demonstrate a right parapharyngeal space soft tissue infiltrate. Note the opacification of the parapharyngeal fat compared to the normal left side.

Figure 2. Magnetic Resonance Imaging of the Brain demonstrating Lemierre’s Syndrome
Axial (A) view shows a right jugular venous thrombophlebitis. Coronal sections (B & C) reveal venous congestion with surrounding edema as well as opacification of the right tentorium cerebelli (C). This constellation of findings was consistent with Lemierre’s Syndrome.

Figure 3. Computed Tomographic Scan of the Chest showing right hilar lymphadenopathy with surrounding edema.