Wegener’s Granulomatosis Presenting with Bilateral Facial Nerve Palsy and Limited to the Head and Neck

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Abstract  Objectives. To illustrate the only reported case of Wegener’s granulomatosis limited to the head and neck, and to provide an update on current knowledge and treatment of Wegener’s granulomatosis. Case report. A 50-year-old man presented with bilateral chronic otitis and mastoiditis, eventually developing bilateral facial palsies. While he progressed with other head and neck symptoms, his disease was limited entirely to the head and neck. Diagnosis of Wegener’s granulomatosis was delayed for this reason, due to the lack of systemic manifestations and the inability to confirm the diagnosis with a tissue biopsy, despite an elevated c-ANCA level. Conclusion. The early diagnosis and initiation of appropriate treatment of Wegener’s granulomatosis is essential to prevent progression of the disease to a severe and irreversible phase. This case emphasizes that an awareness of this rare presentation is important to prevent the unnecessary delay of appropriate treatment in order to optimize the patient’s prognosis, particularly when classic therapy for otitis or facial palsy is ineffective.

Keywords  Wegener’s granulomatosis; facial palsy

1. Introduction

Wegener’s granulomatosis (WG) is a necrotizing vasculitis that develops granulomatous lesions affecting both arterioles and venules [3,8]. It is an uncommon immunologically mediated systemic disease that typically affects the upper airways, lungs, and kidneys [8]. Other organs less commonly affected include the skin, articulations, eyes, and nervous system. For the disease to present with only otolaryngological symptoms is uncommon [3,5].

Otolologic involvement during WG can be classified into five distinct patterns: serous otitis media, sensorineural hearing loss, chronic otitis media, vertigo, and facial palsy [2,7,13]. These manifestations are caused by the granulomatous process in the temporal bone and nasopharynx [10]. The incidence of otologic involvement varies from 19% to 61% in recent literature [1,3,6,11]. Facial palsy is a rare manifestation of WG (5%) and is far less common as the primary presenting symptom [7]. All of the reported cases initially presenting with otolaryngological symptoms alone have progressed to develop systemic manifestations of WG, one of which was fatal. To our knowledge, this is the only case presenting as bilateral facial nerve palsy and limited to the head and neck.

2. Case report

A 50-year-old man was referred to our ENT department with a three-month history of bilateral chronic otomastoiditis and bilateral conductive hearing loss, three-day history of worsening headaches centered around the left temple and mandible, and a 24-hour history of the left lower motor neuron facial palsy (House-Brackmann (HB) grade VI). He had two prior presentations to the emergency department with mastoid pain without referral to our department. Complete head and neck examination revealed bilateral middle ear effusions, and pure tone audiometry showed a mild bilateral conductive hearing loss. A computed tomography (CT) scan with contrast of the temporal bones showed an increased left mastoid fluid level and a new fluid level in the right mastoid (compared with one month previously). He was commenced on intravenous (IV) antibiotics (ticarcillin and clavulanate) and prednisolone 60 mg daily, and a left intact canal wall mastoidectomy with bilateral myringotomies and tympanostomy tube insertion was performed. At surgery, sterile mastoid fluid was drained bilaterally. No growth was returned on culture.

Postoperatively, his ipsilateral facial nerve had improved from HB grade VI to V. He was discharged on prednisolone 60 mg daily, to be tapered down by 5 mg per week, with an immunology review in 2 weeks’ time. The patient represented one month later with a 48-hour history of right facial nerve palsy (HB grade III) and associated right-sided otalgia, the left facial nerve palsy having shown no improvement since discharge; complete examination of the head and neck, including fibroptic nasoendoscopy, revealed only patent bilateral
Figure 1: MRI head, revealing diffuse pachymeningeal enhancement and abnormal enhancement of the right facial nerve.

tympanostomy tubes in situ. Although the nasal mucosa appeared normal, a random biopsy was taken and confirmed as normal mucosa. The patient was screened for autoimmune and granulomatous diseases (anti-nuclear antibodies, rheumatoid factor, cytoplasmic and perinuclear antineutrophil cytoplasmic antibodies, erythrocyte sedimentation rate, C-reactive protein, full blood picture, urea, electrolytes and creatinine, liver function tests, calcium, lactate dehydrogenase) and infectious serologies (herpes simplex virus, cytomegalovirus, human immunodeficiency virus, hepatitis, human T-lymphocyte virus, Epstein-Barr virus, Treponema pallidum, Lyme disease, Mycobacterium tuberculosis), a magnetic resonance imaging (MRI) with contrast and CT temporal bones with contrast were ordered, and he was recommenced on a daily dose of prednisolone 60 mg while results were pending. The c-ANCA level returned a positive result (16 U/mL (normal range < 5 U/mL)), and he was admitted to hospital with a provisional diagnosis of WG for review by the immunology and acute pain services. At this time, his ESR and CRP were not elevated, and this did not change during the course of his disease. A CT chest was performed, revealing an incidental finding of an anterior mediastinal mass most likely suggestive of a thymoma, with differential diagnoses of teratoma and granulomatous mass. CT guided FNA revealed abundant necrotic cellular material only. Though WG was suspected as the cause of bilateral facial palsy and supported by the finding of the modest elevation of c-ANCA, there was no other specific evidence of WG, and the presence of an undifferentiated mediastinal mass that was unable to be biopsied by FNA and would require midline sternotomy access consequently led the immunology team to defer treatment with cyclophosphamide; he was maintained on prednisolone.

The patient was readmitted one week later with worsening severe left facial pain. A repeat MRI/MRA (Figure 1) revealed diffuse pachymeningeal enhancement with thickening involving the left tentorium, middle cranial fossa, left hemispheric convexity extending into the Meckel’s cave on the left side involving the dura along the petrous ridge and also internal auditory canal. The right facial nerve showed abnormal enhancement from the tympanic segment to the second genu, and it was difficult to delineate the facial nerve on the left side due to post operative changes in the mastoid. On the repeat MRI, there was progression of the involvement of the central skull base, particularly basioocciput on the left side centered at the margin of the hypoglossal nerve canal, with left prevertebral enhancing soft tissue and also involving longus capitis. There were postoperative changes in the mastoid and the facial nerves remained unchanged in appearance. The immunology team was satisfied that alternative diagnoses had been excluded, and the patient was diagnosed with WG.

He was commenced on his first of six four-weekly cycles of cyclophosphamide for treatment of WG, and high-dose prednisolone was continued. During the 12-day admission, his pain improved, and he had some improvement in left facial nerve function (to HB grade IV from HB grade V) post cyclophosphamide.

He required yet another admission three weeks later with worsening frontotemporal headaches. His second cycle of cyclophosphamide was administered one week early without complications, notably improving his pain. He was also commenced on IV immunoglobulin therapy 3 × 45 g on consecutive days post cyclophosphamide, and his regimen was altered to three weekly cycles, and prednisolone was continued.

The patient has completed his third cycle of cyclophosphamide. Clinically, his left facial nerve had improved to HB grade III, and his right remains at grade V. He no longer experiences headaches and has returned to work. A facial nerve electromyography (EMG) was performed, which showed that facial nerve responses were absent. Needle examination revealed increased insertional activity and fibrillation potentials, in facial nerve innervated muscles with no activation of motor unit potentials on the right and reduced on the left. Incidentally, the right peroneal motor amplitude was reduced with slowing of the motor conduction velocity. There was no electrophysiological evidence of a large fiber peripheral neuropathy. A follow-up
EMG is planned to further elucidate prognosis of the right facial nerve. We hope that delayed treatment with cyclophosphamide has not compromised his recovery.

3. Discussion

Confirming the diagnosis of WG can be problematic in many cases, as it tends to rely on laboratory and histological findings [4,9,12]. An elevated cytoplasmic pattern antineutrophil cytoplasmic antibody (c-ANCA) level is highly specific for active WG, with a specificity of 98% [4,9,12]. Other diseases that can very occasionally result in raised serological c-ANCA include microscopic polyangitis, idiopathic glomerulonephritis, Churg-Strauss syndrome, and ulcerative colitis. Positive c-ANCA test results have additionally been reported in patients with tuberculosis, Hodgkin’s lymphoma, human immunodeficiency virus infection, nasal septal perforation, and monoclonal gammopathies. The serological ANCA test can be useful as a marker of disease activity and remission following treatment. Although biopsies of suspicious lesions are able to confirm the diagnosis, they are often unrewarding and show only non-specific granulomatous disease, particularly when taken from the middle ear, as occurred in this case with the nasal mucosal biopsies returning normal results [4,9,12].

As WG rarely presents with exclusively otologic symptoms, the clinical presentation can be misleading, as it was in this case. Since the differential diagnosis of this presentation of WG includes tuberculosis, sarcoidosis, polyarteritis nodosa, and malignancy of the temporal bone, nasopharynx or parotid gland, a more aggressive investigative course should be undertaken [4]. Additionally, the likely failure of an antibiotic course should lead the treating team to investigations including chest X-rays, mantoux tests, p- and c-ANCA titers, and biopsies of the middle ear mucosa [4,9]. Importantly, though elevated c-ANCA levels are well reported indicators of WG, the sensitivity has been shown to be only 66%, rising to 90% in active cases [9].

The survival rates of WG have improved dramatically over the last decades, contradicting the poor and often fatal prognosis associated with the disease, due to the widespread administration of early therapy with corticosteroids and cyclophosphamide [2,9,10]. Remission rates of 70–85% have been quoted in the literature, depending on the extent and severity of the disease, with progressive renal disease being the main determinant of survival [4]. The steroid agent causes a reduction in the number and size of lymphocytes present in the lymph nodes and spleen, but does not produce cytolytic effects on the stem cells of the bone marrow [4]. The cyclophosphamide used in conjunction acts as a steroid sparing agent and increases the immunosuppressive action of the steroids, destroying the lymphocytes in proliferation [4].

In the literature, combined use of steroid and immunosuppressive drugs is recommended when there is middle ear and inner ear involvement, and when there is acute onset [4,13].

The prognosis of WG-induced neuropathies is dependent on early diagnosis and prompt initiation of appropriate treatment. Surgical decompression of the facial nerve is not advised, as it may worsen the damage of the nerve [4].

In conclusion, the early diagnosis and initiation of appropriate treatment of WG is essential to prevent progression of the disease to a severe and irreversible phase. We report the first case of WG confined to the head and neck, without typical systemic manifestations, emphasizing that an awareness of this rare presentation, and high index of clinical suspicion is required to prevent the unnecessary delay of appropriate treatment, as occurred in this case, and to optimize the patient’s prognosis, particularly when classic therapy for otitis or facial palsy is ineffective.

4. Summary

(i) WG is an uncommon immunologically mediated necrotizing vasculitis characterized by the development of granulomatous lesions.

(ii) This case describes the first documented case of WG confined solely to the head and neck.

(iii) Diagnosis of WG is often problematic, though an elevated c-ANCA level is 98% specific for WG. Presentations confined to otologic symptoms can be misleading.

(iv) Diagnosis has often been guided by biopsies of lesions confirming WG, and the lack of such lesions able to be readily biopsied can slow diagnosis, as in this case. Additionally, these biopsies can often be unrewarding showing only non-specific granulomatous disease and should not be relied on in order for a diagnosis to be made.

(v) Prognosis of WG-induced neuropathies is dependent on early diagnosis and prompt initiation of treatment.

References