CASE STUDY

Wegener’s Granulomatosis Causing Bilateral Facial Paralysis and Deafness

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KEYWORDS

Wegener’s granulomatosis; Otitis; Bilateral facial paralysis

Abstract Bilateral facial paralysis (BFP) is an uncommon condition that typically occurs as a manifestation of systemic disease. We present a female patient with Wegener’s granulomatosis (WG), particularly upper respiratory and ear impairment who develops hypoacusis and BFP, resistant to immunosuppressive therapy and steroid boluses. Her imaging tests showed no involvement of the facial nerve as it passed through the ear structures. The patient finally improved the BFP; however, deafness is permanent and she has entered into a cochlear implant programme. Published papers on BFP are rare and they make no reference to WG as a possible aetiology.

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PALABRAS CLAVE

Enfermedad de Wegener; Otitis; Parálisis facial bilateral

Granulomatosis de Wegener causante de parálisis facial y cofosis bilaterales

Resumen La parálisis facial bilateral (PFB) es una entidad infrecuente, que habitualmente se presenta como manifestación de una enfermedad sistémica. Presentamos el caso de una mujer afecta de granulomatosis de Wegener (GW), con especial afectación de las vías respiratorias altas y ótica, que desarrolló hipoacusia y PFB resistente a tratamientos inmunosupresores y bolos de corticoides, con pruebas de imagen que no muestran afectación del nervio facial en las estructuras óticas. Finalmente, la paciente mejoró de la PFB, pero la cofosis es permanente y se ha realizado un implante coclear. Las series publicadas sobre PFB son escasas, y no hacen referencia a la GW como posible etiología.

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Introduction

Wegener’s granulomatosis (WG) is a multisystemic disease characterised by the formation of granulomas and necrotising vasculitis. Locoregional involvement is common and may include otological manifestations. Otolaryngologists represent an essential part of the multidisciplinary teams...
involved in the diagnosis and treatment of this disease. Facial nerve involvement has been described during the course of the disease, although it is very rare for it to appear as an initial symptom or in the form of bilateral facial paralysis (BFP).²

BFP is a rare entity which usually appears as a manifestation of a systemic disease, with the most common causes being Lyme disease, Guillain–Barre syndrome, sarcoidosis, bacterial meningitis and cranioencephalic lesions.³

Case Report

We present the case of 33-year-old female suffering WG, primarily affecting the upper respiratory and aural (repeated otitis) pathways. Since June 2009, she presented persistent otic inflammatory activity, prompting repeated consultations at the Otorhinolaryngology and Collagenosis Services. The patient presented chronic dizziness, unsteadiness, headaches and frequent episodes of nausea and vomiting.

In late August 2009, she was diagnosed with otitis media with effusion (OME) and underwent myringotomy with placement of bilateral ventilation tubes. Following this intervention, the headaches worsened, hearing loss increased and, 2 weeks later, she developed right, peripheral facial palsy of grade III in the House–Brackmann classification, for which she was admitted to hospital. During admission we initially requested the following tests: (1) blood count and biochemical analysis, which were normal; (2) ESR, which was 74; (3) otic exudate cultures, with normal flora; (4) lumbar puncture and CSF analysis, which was normal, and (5) chest radiography and Mantoux test, which were negative. Being an immunosuppressed patient, we initially stopped glucocorticoids and cytostatics until we obtained the first microbiological data. We took into account the possibility of bacterial, tuberculous and fungal meningitis, and the patient was treated with meropenem and ampicillin.

We requested a magnetic resonance imaging (MRI) scan, which showed bilateral, chronic otomastoiditis with an acute inflammatory appearance but without intracranial repercussion. During admission, the paralysis became bilateral and we observed the loss of the ventilation tube in the left ear, so a new one was placed. We requested a computed tomography (CT) scan, which revealed occupation of the right ear mastoid, without signs of erosion of bone structures, as well as an intact facial nerve, without dehiscence. Finally, the patient developed peripheral dizziness symptoms. She received a total of 8 boluses of 1 g methylprednisolone and the previous immunosuppressants were replaced by mycophenolate sodium. This treatment achieved a partial improvement and she was discharged with outpatient follow-up (Figs. 1 and 2).

At present, dizziness, OME and BFP symptoms have improved. However, cophosis is permanent and the patient has been included in a cochlear implant programme at another hospital.

Discussion

Involvement of the ENT region represents a classic symptom of patients with WG and it appears in over 70% of patients as an initial symptom. Almost all patients present some

Figure 1  Cerebral MRI scan showing bilateral otomastoiditis with an acute inflammatory appearance, with no evidence of intracranial repercussion.

ENT manifestation during the course of the disease.⁴ Otic involvement in WG may include OME, chronic otitis media, sensorineural hearing loss, facial paralysis and vertigo. OME is the most common type of otic involvement, with tubal obstruction or nasopharyngeal inflammation being the most common causes.⁵ Sensorineural hearing loss is also common among these patients.⁶

Figure 2  Cerebral CT scan showing no abnormalities in facial nerve paths.
Facial paralysis in patients with WG is infrequent and is rarely the first sign of disease. It is normally secondary to compression of the nerve in the middle ear, especially in its path through the Fallopian canal, when it is dehiscent or due to the existence of vasculitis affecting the microcirculation.\(^7\)

BFP is a rare condition, which usually indicates a more severe alteration than unilateral paralysis.\(^2\) There are few case series of BFP in the literature. In the series of Keane with 43 patients, Bell’s palsy was noted as the most common cause, followed by Guillain–Barre syndrome.\(^3\,\(^8\)\) The series of Wormald was smaller, with only 24 patients, but also noted Bell’s palsy as the most common cause.\(^3\,\(^9\)\) Other series are more limited and limited to several cases of BFP. These series do not make reference to WG as the cause of bilateral paralysis. We have found only 1 reference in the literature of a patient suffering WG who developed BFP.\(^10\)

Other systemic diseases affecting the middle ear and temporal bone which should be included in the differential diagnosis of otic involvement of WG are tuberculosis, fungi, syphilis, Lyme disease, sarcoidosis, polyarteritis nodosa, Churg–Strauss and tumours in neighbouring structures.\(^10\) Early diagnosis and prompt initiation of treatment are essential to improve the prognosis of facial nerve involvement.\(^11\,\(^7\)\)

In the present case, glucocorticoids were discontinued and treatment with broad spectrum antibiotics was initiated upon suspicion of infection. After conducting complementary tests which did not reveal the existence of infection, treatment was replaced by boluses of methylprednisolone and mycophenolate sodium. Placement of ventilation tubes in the ears was performed upon suspicion of facial nerve compression. Following improvement, the patient was discharged, with immunosuppressive therapy and outpatient controls. Although BFP and dizziness symptoms remitted, unfortunately ophiasis was irreversible and the patient was eventually included within a cochlear implant program.

The treatment of vasculitis with ANCA (anti-neutrophil cytoplasmic antibodies) has been well defined, but must conform to the characteristics of each patient and the severity of the granulomatosis. Furthermore, the risk of side effects inherent to the treatment should also be considered. Relapses are still common among patients with WG. These patients typically receive a combination of corticosteroids and immunosuppressants. After achieving remission, cyclophosphamide is often replaced by a less toxic immunosuppressant. Biological drugs should be initiated with caution, since their adverse effects are few, but potentially severe.\(^13\) Improvement in the prognosis of these patients is due to an adequate treatment, correctly adjusted prophylaxis and careful management of complications.\(^13\)

**Conflict of Interests**

The authors have no conflict of interests to declare.

**References**