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Otorrinolaringological manifestations of granulomatosis with multiple Mononeuropathy and Poliangeítis: A diagnostic challenge

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Abstract
Granulomatosis with Polyangiitis (GPA), previously known as Wegener's Granulomatosis, is an entity with great potential to trigger multiple manifestations in the head and neck region. The initial presentation form and its non-specific characteristics is usually misdiagnosed and confused with different entities. Within the manifestations of peripheral neuropathy, multiple mononeuritis is more common, while central nervous system involvement is more rare. We address an interesting case of granulomatosis with polyangiitis that manifested itself with multiple cranial nerve involvement mononeuritis.

Keywords: granulomatosis with Poliangeítis, multiple mononeuritis, manifestations in ear, nose and throat

Introduction

In 2011, the American College of Rheumatology, the American Society of Nephrology and the European League against Rheumatism recommended that the name "Wegener's granulomatosis" be changed to "granulomatosis with polyangiitis (GPA)", abbreviated as Weneger (GW) is a systemic vasculitis of small vessels, necrotizing and granulomatous GPA11 Granulomatosis with Polyangiitis is a condition with great potential to trigger multiple manifestations in the head and neck region, in most cases being the initial presentation form and the nonspecific characteristics of its symptoms it is usually under-diagnosed and confused with different pathologies, such as allergic and / or infectious, which leads to a delay in diagnosis and treatment [1, 2]. Involvement of the upper respiratory tract is frequent during the course of the disease1, 3. Neutrophil anti-cytoplasmic antibodies (ANCA), which produce a cytoplasmic pattern (c-ANCA) are highly specific during the active phase [2, 3]. The positivity of proteinase 3-ANCA (PR3-ANCA) is normally associated with the progression of the disease, from its localized to the systemic state.2 It is a vascular systemic disease characterized by vasculitis of arterioles and venules, most patients have pathology nasal or sinus, the otological condition varies from 19 to 61% [3].

Neurological manifestations occur in 22% to 54% of patients. The most common is peripheral neuropathy, especially multiple mononeuritis, while central nervous system (CNS) involvement occurs only in 2% to 8% of cases [6, 7, 8, 9].

In 2017, Fragoulis and collaborators conducted a retrospective study in Greece evaluating the CNS commitment in the GPA. They presented 77 patients of which 9 (11.7%) developed neurological manifestations. The manifestations found were: sensory-motor symptoms (33.3%), severe headache and hearing loss (33.3%), delusions and convulsions (22.2%), diplopia (11.1%) and cerebellar symptoms (11.1%). It was seen that in these patients the ENT and the disease activity levels were frequently lower; as well as they had less lung involvement [10].

The American Rheumatology (ACR) criteria for GPA include the following 12

- Nasal or oral inflammation (painful or painless oral ulcers, or purulent or bloody nasal discharge)
- Abnormal chest x-ray showing nodules, fixed infiltrates or cavities.
- Abnormal urinary sediment (microscopic hematuria with or without red blood cell molds)
- Granulomatous inflammation in the biopsy of an artery or perivascular area.

We present a case of otological manifestation and involvement of more than two cranial nerves, approached as an infectious process erroneously, until the final diagnosis is found.

Case Presentation

A 27-year-old female patient, previously healthy, who begins a current condition 7 months prior to hospitalization with bilateral nasal obstruction and persistent hyaline rhinorrhea, managed with nasal steroid and antihistamine with partial improvement; Four months later, he presented with progressive bilateral otalgia and hearing loss, as well as intermittent acute bilateral tone tinnitus, left peripheral facial paralysis and unquantified fever, beginning medical management of complicated acute otitis media, placing bilateral ventilation tubes, showing clinical improvement and complete recovery of facial paralysis. At 2 weeks later, peripheral

facial paralysis of the right side, bilateral yellowish otorrhea and a greater degree of hearing loss are added, as regards this third level unit.

Upon physical examination, a patient with peripheral facial paralysis on the right side is observed (stage III / VI on the House-Brackmann scale, bilateral yellowish otorrhea through the tympanostomy tubes, the audiometry reveals a severe sensorineural hearing loss Figure 1, was performed Simple and contrasted tomography of the ear and temporal bone, revealing only occupation of mastoid air cells and middle ear due to liquid density, without abscess formation or bone destruction, Figure 2, during his hospital stay he presents dysphonia, dysphagia to solids and liquids, hemoptysis and Bilateral parotid volume increase, laryngoscopy was performed evidencing left chordal paralysis Figure 3, in the same way, mechanic swallowing in oropharyngeal phase with water-soluble contrast material was performed, demonstrating data of glotic insufficiency with data of aspiration by twelfth cranial nerve paralysis When re-evaluating the case involving affection of the VII, VIII and XII cranial pair, magnetic resonance imaging of the brain is requested in the search for central pathology, which was normal.

A bronchoscopy is performed where the left chordal paralysis and bleeding traces from the right middle lobe are confirmed. Figure 4. Subsequently, a simple and contrasted chest CT scan is observed where pulmonary nodules of defined edges are observed in the right lung. Figure 5, Antibodies to fungi and Negative bacteriological study. Biopsy of nasal septum with data of nonspecific chronic inflammation Figure 6. Bronchial lavage and brushing negative to malignancy. Laboratories general urine test with glycosuria> 500 mg / dl, leukos 8-10, yeasts, hyphae. Glucose 268. Creatinine 0.75, Cholesterol tola 312 mg / dl Triglycerides 227 mg / dl ALT 926 mg / dl AST 436 mg / dl, alkaline phosphatase 226 mg / dl, DHL 372 mg / dl, PCR <0.7 mg / dl Antibodies c- Positive ANCAS Management with glucocorticoids and cyclophosphamide was initiated by the rheumatology department, presenting improvement of the inflammatory process, recovering the motor function of the VII and XII cranial nerve, without recovering the sensory function of the bilateral bilateral cranial nerve, resulting in severe bilateral hearing loss in the 6 months of follow-up.

Discussion

There are few studies in the literature that document bilateral facial paralysis (PFB) and multiple mononeuritis (MNM), which involved cranial nerves VII, VIII, XII within the spectrum of manifestations of Granulomatosis with Polyangiitis (GPA).

GPA is a systemic vasculitic disease, although some forms are limited to the respiratory tract [1, 2]. If left untreated, GPA constitutes a disease with rapid progression, with 82% mortality within one year [1, 3].

Facial nerve paralysis has been reported during the course of the disease, although it is a characteristic that occurs in extremely rare cases.5 Pulmonary involvement is common, including nodular lesions [5].

ANCAs are detectable in almost 100% of patients with generalized GPA disease, although only 60% in their localized form. The ELISA test for highly sensitive PR3-ANCA yields a sensitivity of 96% and a specificity of 98.5% 2.5. The combination of glucocorticoids and cyclophosphamide is the standard treatment of GPA. It is important to start treatment before irreversible changes occur5.

Other systemic diseases with which the differential diagnosis of the otic involvement of GPA should be established are tuberculosis, fungi, syphilis, Lyme disease, sarcoidosis, other vasculitis and tumors. Patients usually present with constitutional symptoms that include fever, malaise, anorexia and weight loss. Prodromal symptoms can last for weeks or months without evidence of specific organ involvement.14,15 Keep in mind its most common symptoms such as atypical ones, especially multiple mononeuritis and bilateral facial paralysis could guide the diagnosis, for early treatment, improving neurological prognosis. The otolaryngologist plays an important role in the multidisciplinary team involved in the diagnosis, monitoring and treatment of these patients. Within the multiple classifications, we could consider our case as vasculitis associated with anti-neutrophil cytoplasmic autoantibodies (ANCA), since neuropathy is a common characteristic of most ANCA-associated vasculitis and is associated with a more serious disease.13

In conclusion, GPA is difficult to diagnose, considering in the refractory inflammatory states of the ear, associated with multiple mononeuritis that involves more than two cranial nerves. This case revealed an aggressive progression that began with otological manifestations, being diagnosed based on serum and clinical characteristics.

Anexo

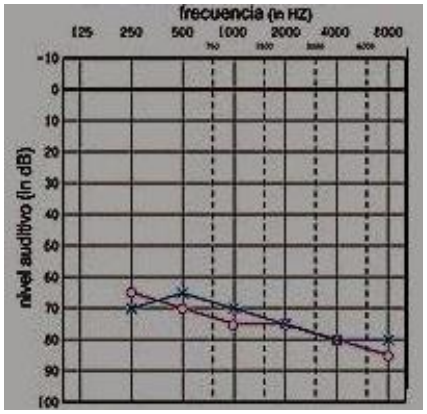

Fig 1: Tonal audiogram, showing a bilateral sensorineural hearing loss schematized.

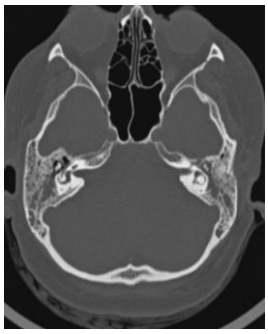

Fig 2: Simple tomography of the middle ear and mastoids showing occupation of mastoid air cells and middle ear by liquid density, without abscess formation or bone destruction

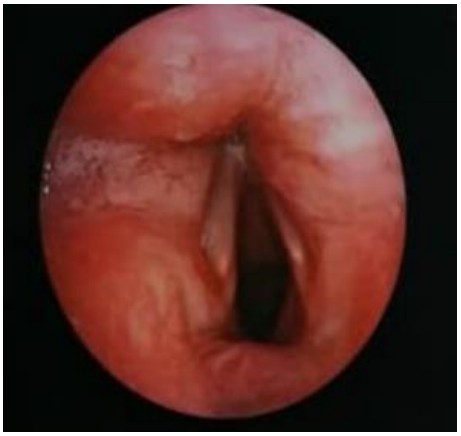

Fig 3: Laryngoscopy evidencing left chordal paralysis in position for true left vocal cord mean.

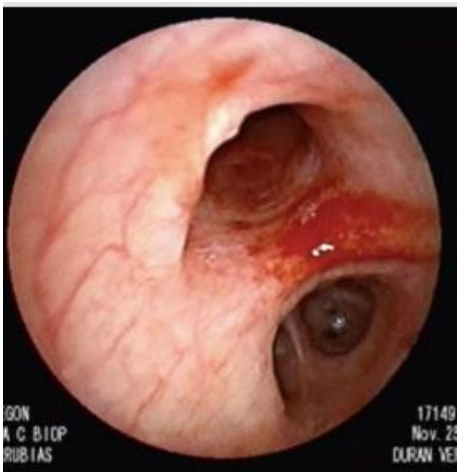

Fig 4: Bronchoscopy confirming left chordaeal paralysis and traces of bleeding from the right middle lobe. Bronchial brushing was taken with a negative result for fungi, bacteria and malignancy.

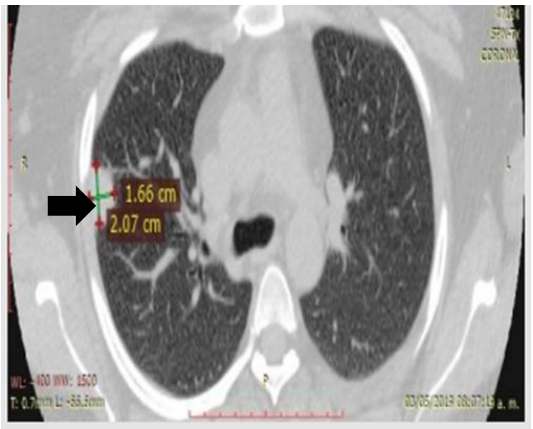

Fig 5: CT scan of the chest with a lung window showing pulmonary nodules with defined edges in the right lung

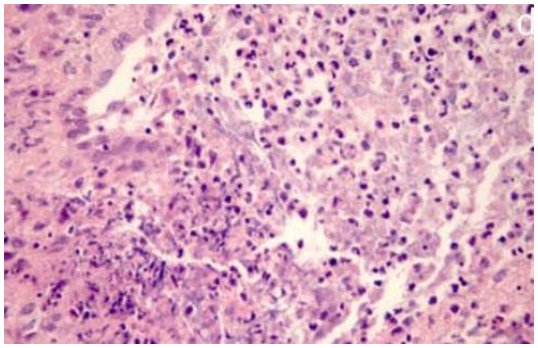

Fig 6: Biopsy of the nasal septum with data of nonspecific chronic inflammation

Interest conflict

All authors declare that there are no conflicts of interest in this work.

Authorship

Each author contributed substantially to the design, analysis, revision and approval of the writing, assuming responsibility for all aspects of the work.

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Ethical issues and informed consent

This work keeps the identity of the patient and respects the ethical principles for medical research in human beings with adherence to the declaration of Helsinkide the World Medical Association

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