Wegener's Granulomatosis: An Unusual PresentationCase Report and Review of Literature

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Abstract

Wegener's granulomatosis is a rare vasculitic disorder affecting multiple systems and typically involves the kidneys, lungs and the nose. Its presentation as an isolated involvement of the ear is very uncommon and only a few cases presenting as an isolated involvement of the ear with unilateral facial palsy and deafness have been reported in world literature. We report a case of isolated otological involvement presenting as facial palsy and bilateral profound deafness. The renal and pulmonary systems and the nose were unaffected.

INTRODUCTION

Wegener's granulomatosis (WG) is a multi-system disease of unknown aetiology and is characterized by granulomatous lesions involving both arterioles and venules with widespread necrotizing vasculitis. It classically presents as a triad affecting the upper and lower respiratory tracts and the kidneys. However, any organ can be the site of granulomatous disease or vasculitis or both[1].

Otologic manifestations have been reported during the course of the disease but they are rare as initial presenting features[2-6]. Facial palsy and deafness as presenting features is even less common.[7-11]. Anticytoplasmic antibodies against neutrophil polymorphonucleate granules (c-ANCA)which are positive in 97% of the cases, and Anti Proteinase 3 (anti PR3), positive in 100% of cases, are highly specific for the diagnosis[13]. Early diagnosis and timely medical treatment lead to high rates of remission of an otherwise lethal disease. We report a case of WG presenting only with facial paralysis and the unusual occurrence of bilateral profound deafness. This combination as a presenting feature is unique and is worth noting in an entity Otolaryngologists typically see only in the nose.

CASE REPORT

PRESENTATION

A 47 year old female presented to the Emergency services of the Department of Otolaryngology, Head & Neck Surgery of INSCOL Hospital, Chandigarh, India with the sudden onset of complete Lower Motor Neuron (LMN) type Facial palsy of the Right side. There was no history of trauma.

HISTORY

One month prior to presentation, the patient developed fullness in the right ear and mild hearing loss and tinnitus following an episode of Upper Respiratory Tract Infection. The right tympanic membrane showed a Grade 2 Retraction of the Pars Tensa and the facial nerve was normal.

A Pure –Tone Audiogram confirmed a mild conductive hearing loss. A tentative diagnosis of Secretory Otitis Media was made and a course of topical and systemic nasal decongestants was prescribed with advice on ear autoinflation. These measures did little to alleviate her symptoms. A High Resolution Computed Tomography (HRCT) of the temporal bone was performed but this was not contributory and medical treatment was extended. About a month after her initial symptoms, the patient suddenly developed unilateral Right Sided LMN facial palsy and bilateral deafness and was referred to us.

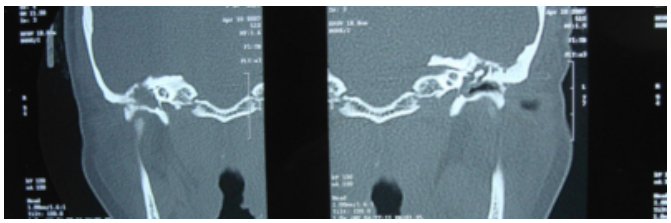
EXAMINATION AT PRESENTATION

Examination revealed bilateral congestion of both tympanic membranes with a bulge in the posterosuperior and posteroinferior quadrants. Tuning fork tests led us to suspect bilateral profound sensorineural deafness and this was confirmed on Pure Tone Audiometry. Electrophysiological tests (Nerve Stimulation testing) of the facial nerve confirmed significant loss of function on the Right in all the extra-temporal branches. HRCT of the temporal bone was repeated which showed a soft tissue density filling the

middle ear cleft on both sides with erosion of the facial canal on the right side (Figure 1). Based on the patient's previous history and current findings, we made a tentative diagnosis of Acute Suppurative Otitis Media with Complications.

Figure 1

Figure 1: High Resolution CT of Temporal Bones showing soft tissue densities in both mastoid cavities



MANAGEMENT

After a written informed consent, a Right myringotomy was performed but instead of pus or fluid, we were surprised to find only bleeding granulations. The procedure was abandoned and a fresh consent was obtained for Right Mastoid Exploration. On exploration, there were exuberant granulations in the mastoid antrum and the underlying bone was osteitic. The ossicles though intact were embedded in the granulations. We proceeded to remove these granulations as much as safely possible for histopathology and we also proceeded to decompress the facial nerve throughout its course. A nasal endoscopy was performed but was normal. The suspicion of WG was considered due to presence of exuberant granulations but histopathology was not confirmatory. However, samples sent for c-ANCA and anti-PR3 were strongly positive. An HRCT of the chest (Figure 2) was normal as was a Renal Ultrasound and urinalysis.

Based on the operative findings and positive c-ANCA and PR3 levels, we revised our diagnosis to Wegener's Granulomatosis and the patient was started post operatively on Prednisolone at 1mg/kg/day and Cyclophosphamide at 3mg/kg/day. There was significant improvement of the facial nerve status and the hearing. The hearing of the non-operated ear also recovered significantly.

The steroids were tapered over 6 months and discontinued. However the patient developed a relapse in the form of increase in the tinnitus and increased value of c-ANCA and low-dose steroid therapy was restarted at 0.3mg/kg/day. The patient is symptom free at one year of follow-up.

DISCUSSION

WG, first reported by Friedrich Wegener[14] in 1936, is considered to be of autoimmune origin, with a peak age of presentation in the fifth and sixth decades and with a slight male preponderance. (1.5:1).

One third of the patients present with a locoregional form. Described in 1953 by Fienberg[15] and subsequently supported by Cassan[16] in 1970, this ''limited'' form of WG is a restricted illness in which clinical and histologic findings are initially present only locoregionally, without renal involvment. An otologic involvement of WG is not uncommon. Overall, 38% of cases of WG manifest with otological involvement at some point in the course of the disease with the middle ear being the commonest site of involvement[2-6]. Unilateral serous otitis media is the most common manifestation seen in 90% of the cases with bilateral otitis in 33% of the cases. Chronic otitis media with mastoid disease has been seen in as high as 24% of the cases. The next site in the ear affected is cochlea and auditory nerve presenting as sensorineural hearing loss in 43% of the patients. Vertigo is uncommon and probably results from central nervous system involvment with a polyneuritis or because of the immunocomplex deposition in the vestibular portion of inner ear. Peripheral facial nerve palsy due to temporal bone involvement is seen in 8% of the cases of WG and is secondary to the concomitant otitis media[7-12]

Facial palsy as a presenting sign of WG is extremely uncommon with very few cases reported in the literature[7-12]. Ipsilateral hearing impairment may be seen as well. However our case had bilateral profound sensorineural deafness and unilateral facial palsy, which is a very unusual combination as a presenting feature. The early onset and the non specificity of the symptomatology precluded a correct diagnosis. Under these circumstances, the diagnosis can be suggested by failure of recovery following the usual principles of management chronic otitis media and facial palsy, or with the appearance of the symptoms or signs of other organ system involvement

Intra operatively our patient showed exuberant granulations in the mastoid and middle ear cavity that was previously well pneumatized. This feature is rarely seen in chronic otitis media and moreover, its development had been too quick and dramatic to be interpreted as acute otitis media facilitating the diagnostic suspicion.

A histopathologic identification of WG is a difficult finding, especially if the disease presents as a local manifestation. Moreover, it may often show non-specific granulomatous disease, particulary when it has been taken from the middle

ear, as seen in our case. [3,4,5]

An early diagnosis can be made by serologic studies of specific markers such as c-ANCA & anti-PR3[17]. The sensitivity of c-ANCA test varies in relation with the disease's activity. In the generalized phase it is 97%, whereas in case of locoregional involvment it is 60%[12]. The elevation of anti PR3 is highly specific even in the locoregional variant. It is an ideal test for an early diagnosis in all suspected cases.

It is recommended to use a combination of steroids and immunosuppressive drugs especially when there is middle ear and inner ear involvement and when the onset is acute. This treatment is the standard therapy for WG[18]. We initiated combined treatment with cyclophosphamide and steroids after positive results with c-ANCA and anti PR3. Surgical decompression of the facial nerve is usually unrewarding and may aggravate the damage of the nerve due to non identification of the nerve during surgery. However, at the time of surgery, we were unsure of the diagnosis and went ahead with decompression but fortunately there was no deterioration of the palsy. Untreated, the disease runs a rapidly fatal course and 82% of patients die within 1 year. Progressive renal disease is the major cause of death. ESR and c-ANCA levels are used for monitoring disease activity and for an early diagnosis of relapse.

In conclusion, the early diagnosis of WG and the appropriate timely treatment are important to prevent the progression of this disease to a generalized and irreversible phase. We report a case of unilateral LMN facial nerve paralysis with bilateral profound sensorineural deafness as presenting features of WG, emphasizing again that a high level of suspicion of this disease is needed to the Otorlaryngologist. This treatment protocol provides a high incidence of recovery (95% of the cases).

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