GAVIN PUBLISHERS

OPEN ACCESS

Annals of Case Reports

Šteňová E, et al. Ann Case Report 6: 612.

DOI: 10.29011/2574-7754.100612

Case Report

Bilateral Mastoiditis and Pachymeningitis as the Manifestation of Granulomatosis with Polyangiitis

Emőke Šteňová[1*], Stela Kašperová[1], Soňa Kiňová[1], Alena Tuchyňová[2]

[1]1st Department of Internal Medicine, Faculty of Medicine, Comenius University in Bratislava and University Hospital Bratislava, Slovakia

[2]National Institute for Rheumatic Diseases, Piešťany, Slovakia

[*]**Corresponding author:** Emőke Šteňová, 1st Department of Internal Medicine, Faculty of Medicine, Comenius University University Hospital Bratislava, Staré Mesto, Mickiewiczova 13, 813 69 Bratislava, Slovakia

Citation: Šteňová E, Kašperová S, Kiňová S, Tuchyňová A (2021) Bilateral Mastoiditis and Pachymeningitis as the Manifestation of Granulomatosis with Polyangiitis. Ann Case Report 6: 612. DOI: 10.29011/2574-7754.100612

Received Date: 16 January, 2021; **Accepted Date:** 20 January, 2021; **Published Date:** 25 January, 2021

Abstract

Vasculitis is an inflammatory disorder of the blood vessels. Clinical symptoms are depend on the size of the involved vessels and affected organs. Upper airway disease is the most common manifestation of granulomatosis with polyangiitis (GPA). We report a 54-year-old woman with infrequent mastoiditis and meningeal involvement in GPA.

Keywords: Bilateral mastoiditis, Granulomatosis with polyangiitis, Otitis media, Pachymeningitis

Abbreviations: ANCA: Anti-Neutrophil Cytoplasmic Antibodies; BVAS: Birmingham Vasculitis Activity Score; CRP: C-Reactive Protein; ENT: Ear, Nose, Throat; ESR: Erythrocyte Sedimentation Rate; GPA: Granulomatosis with Polyangiitis; OM: Otitis Media

Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener´s granulomatosis, is a rare autoimmune systemic necrotising vasculitis characterised by damage of small- and medium-size blood vessels associated with significant morbidity and mortality. The disease usually presents with non-specific constitutional symptoms such as fever, night sweats, weight loss and fatigue. GPA shows a broad clinical manifestation ranging from limited disease to a severe generalised form with multiorgan involvement. Pulmonary and renal damage can become fatal in some cases due delayed diagnosis. The respiratory system is most commonly affected, i.e. upper and lower respiratory system with common ENT (ears, nose, throat) manifestations [1]. Chronic otitis media with painful discharge, decreased hearing, deafness, sinusitis, crusting rhinitis, saddle-nose deformity and septum perforation are frequent symptoms. Anti-neutrophil cytoplasmic antibodies (ANCA) are present in almost all patients with GPA.

Case

A 54-year-old Caucasian woman presented with complaints of left ear fullness and pain for one month. The patient´s medical history was insignificant. She had consulted an otolaryngologist; paracentesis and drainage of serous fluid of the left ear were performed. After this intervention, she developed a high-grade fever at 38-39ºC. Empirical antibiotic treatment was started (clarithromycin) with a partial effect; she continued to have low-grade fever at 37.2-37.6ºC. One month after paracentesis, a severe headache appeared in occipital lobe and in the region of the left ear, radiating to the left eye. She was admitted to hospital due to acute sever rotational dizziness and vomiting worsened with movement. The physical examination revealed bilateral conjunctivitis. Based on neurologic examination findings, peripheral vestibular syndrome was diagnosed. Initially, laboratory studies showed an elevated erythrocyte sedimentation rate (ESR) 80 mm/hour; C-reactive protein (CRP) was also high (29.93 mg/l) and the complete blood count (CBC) was without abnormalities. Hepatic transaminase activity was slightly elevated and kidney function tests were normal, but urine analysis showed mild erythrocyturia without proteinuria. Control CBC after one week showed mild hypochromic anaemia (113g/l), reactive thrombocytosis (619.1G/l) and leukocytosis (14.65G/l) with neutrophilia. Computed tomography (CT) scans of the head and temporal bones were performed. On the image, there was fluid in the mastoid air cells bilaterally (Figure 1). Cerebrospinal fluid analysis showed

monocyte-predominant pleocytosis, hyperproteinorachia and intrathecal synthesis of IgG, which are atypical for bacterial infection and thus reactive inflammation was suspected. Magnetic resonance imaging (MRI) showed bilateral otitis media and mastoiditis, diffuse dural thickening and meningitis in the posterior cranial fossa (Figure 2.a, b). There were no signs of paranasal sinus inflammation. CT did not reveal any pulmonary findings, and functional tests did not show abnormalities. Intravenous antibiotic therapy was administered with moxifloxacin. However, this therapy increased acute phase reactants (CRP 143 mg/l). We suspected a non-infective aetiology of chronic mastoiditis. The patient tested positive for c-ANCA by indirect fluorescence and proteinase-3 by enzyme-linked immunosorbent assay (normal range 0-20, the patients result was >200kIU/l). Based on these findings, she was diagnosed with granulomatosis with polyangiitis with multiorgan manifestation (ENT, CNS, nephritis, conjunctivitis). The Birmingham Vasculitis Activity Score (BVAS) classified the disease status as severe (BVAS 20).

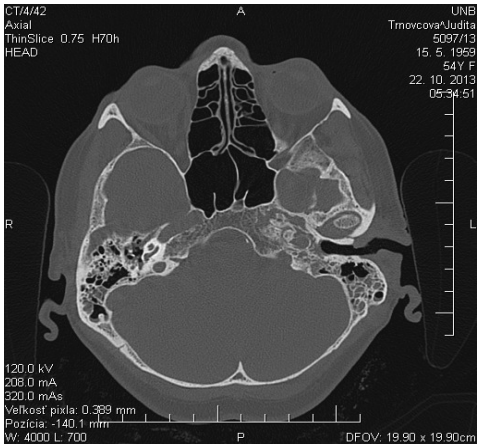
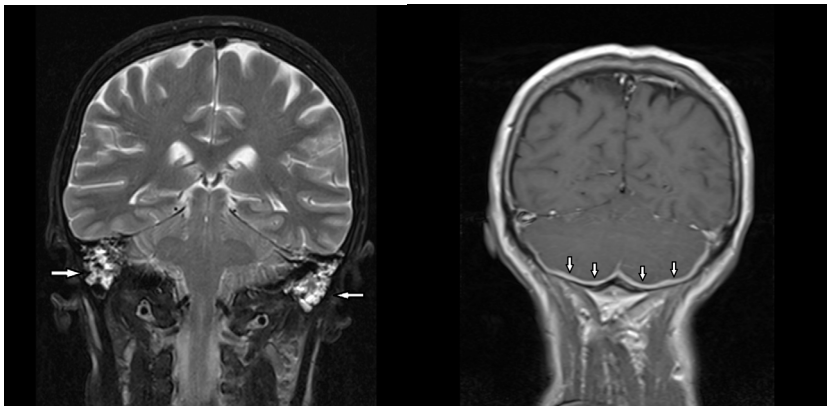


Figure 1: Bilateral mastoiditis (CT)- fluid in the mastoid air cells (white arrows).

Induction treatment included intravenous methylprednisolone 250 mg/day for 3 days followed by oral glucocorticoid therapy (prednisone 40 mg daily). Due to the renal involvement, monthly intravenous pulses of 1000 mg cyclophosphamide and 500 mg methylprednisolone were indicated. After one week of corticosteroid treatment, the patient had no complaints regarding ENT. This therapy led to the normalisation of ESR and CRP after one month and laboratory parameters of liver and renal function were also normal. We continued with this monthly intravenous treatment (cyclophosphamide and methylprednisolone) and slow oral corticosteroid tapering for 12 months. The control MRI scan after one year of treatment did not show inflammation of the processus mastoideus bilaterally. Corticoid-sparing immunomodulatory therapy with oral methotrexate 15 mg weekly was started. After 25 months of an established diagnosis, systemic steroid therapy was slowly tapered and stopped, and the patient has continued with methotrexate treatment without relapse.



(a) **(b)**

Figure 2: a, b: Diffuse dural thickening - MRI (white arrows).

Discussion

Otologic involvement occurs in 20-60% of GPA patients, and otitis media (OM) is the most common manifestation [2]. Destructive granulomatous inflammation may spread from the middle ear through the mastoid towards the petrous apex, accompanied by effusion, mastoiditis, facial palsy or hypertrophic pachymeningitis. In this case inflammation spread from middle ears through the inner ears to the mastoids and caused peripheral vestibular syndrome, which is an uncommon presentation of disease. Symptoms of OM are usually unilateral, and bilateral involvement with bilateral mastoiditis is extremely rare [3]. The diagnosis of GPA in oligosymptomatic cases with medial ear involvement is often delayed. Yoshida et al. reported three months on average required to reach the final diagnosis of ANCA-associated vasculitis presenting with OM [4]. The diagnostic criteria for ANCA- associated OM were designed for early diagnosis of disease [5].

Otomastoiditis may be associated with other neurological complications. Facial nerve palsy is seen in 5% of cases [6]. CNS manifestations include also vasculitis of small to medium-sized vessels of the brain or spinal cord and granulomatous masses that usually involve the orbit, optic nerve, meninges or brain [7]. Meningeal involvement, which can be diffuse or focal, is rarely presented. The mechanisms causing CNS disorders in GPA are contiguous invasion of the granuloma from extracranial sites, CNS vasculitis and remote intracranial granuloma [8]. Pachymeningitis as a complication of GPA should be suspected if patient complains of a severe headache. Early diagnosis of GPA with meningeal infiltration prevents other serious complications and the need for invasive treatment such as surgical decompression.

Conclusion

Patients presenting with recurrent otitis media, a slowly resolving ear inflammation or mastoiditis with peripheral vestibular syndrome unresponsive to conventional therapy should be investigated for granulomatosis with polyangiitis. The importance of early diagnosis and treatment must be emphasised in order to avoid progression to a severe form of disease, especially in oligosymptomatic cases.

The authors have no conflict of interest.

References

1. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC et al. (2013) 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 65:1-11.
2. Costa CF, Polanski JF (2015) Wegener Granulomatosis: Otologic Manifestation as First Symptom. Int Arch Otorhinolaryngol. 19: 266-268.
3. Preuss SF, Stenner M, Beutnet, Laudes M, Klussmann JP (2008) Fatal course of Wegener's granulomatosis with bilateral otomastoiditis and bilateral facial nerve palsy. Otolaryngol Head Neck Surg. 138: 799-800.
4. Yoshida N, Iino Y (2014) Pathogenesis and diagnosis of otitis media with ANCA-associated vasculitis. Allergol Int. 63: 523-532.
5. Harabuchi Y, Kishibe K, Tateyama K, Morita Y, Yoshida N et al. (2020) Clinical characteristics, the diagnostic criteria and management recommendation of otitis media with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (OMAAV) proposed by Japan otological society. Auris Nasus Larynx. 4: S0385-8146(20)30161-9.
6. Drinias V, Florentzson R (2004) Facial palsy and Wegener's granulomatosis. Am J Otolaryngol. 25: 208-212.
7. Holle JU, Gross WL (2011) Neurological involvement in Wegener's granulomatosis. Curr Opin Rheumatol. 23: 7-11.
8. Tasali N, Cubuk R, Celik L, Canbora B (2009) Atypical meningeal involvement in Wegener granulomatosis: MR imaging survey of a particular case. AJNR Am J Neuroradiol. 30: E116.