Case report/Kazuistyka

ENT manifestations of Wegeners granulomatosis

Otologiczne objawy ziarniniaka Wegenera

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ABSTRACT

Wegeners granulomatosis is a necrotizing granulomatous vasculitis with multisystemic involvement. We present two cases of Wegener's presenting with otological manifestations as the first symptom. These symptoms are subtle and diagnosis may be easily overlooked. Hence a high index of suspicion is required. Early diagnosis and treatment goes a long way in improving the outcomes and in preventing further complications.

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- Wegeners granulomatosis
- Facial palsy
- Hearing loss

Słowa kluczowe:
- ziarniniak Wegenera
- porażenie nerwu twarzowego
- utrata słuchu

Introduction

Wegener’s granulomatosis was first described by Heinz Klinger in 1931 and subsequently by Frederich Wegener in 1936 [1]. It is a rare autoimmune vasculitis characterized by necrotizing granuloma of the respiratory tract, focal necrotizing glomerulonephritis, and systemic vasculitis.

Several head and neck regions may be affected by the disease, producing clinical manifestations as follows [2].
- Nose: nasal congestion, epistaxis, discharge, crusting, saddle-nose deformity, septal perforation
- Larynx: dyspnea, stridor, hoarseness, or other alterations of the voice
- Ear: hearing loss, conductive hearing loss, sensorineural hearing loss, facial nerve paralysis, vertigo
- Oral cavity: ulcerations, gingivitis
- Orbit: swelling, proptosis, redness, pain, visual disorders

More than 80% of patients with this diagnosis experience rhinological morbidity and 20–40% experience otological morbidity at some point during their life [3].

We present a series of cases presenting with otological manifestations as presenting symptoms of Wegener’s.

Case report 1

A 35-year-old male presented with nasal obstruction, headache, occasional epistaxis and left sided facial
weakness (House Brackmann grade V) since two months. Just five days before presenting to us this patient developed a right sided facial weakness, associated with bilateral decreased hearing and otorrhea. Otoscopy revealed bilateral central perforation with bilateral grade V facial palsy (Figs. 1 and 2). Nose examination was normal. An audiogram showed right profound sensorineural and left moderately severe conductive hearing loss. HRCT temporal bone showed bilateral chronic mastoiditis. CT PNS showed left maxillary, ethmoidal and frontal sinusitis with enhancement in the region of left nasopharynx involving Eustachian tube, torus tubarius and fossa of rosenmuller. Laboratory tests showed a high ESR (105 mm at one hour). Urinanalysis was negative. Routine Xray chest demonstrated bilateral hilar prominence following which a CT chest was done. It showed multiple nodular opacities all over the lung parenchyma predominantly in lung bases and also subpleurally. A biopsy from nasopharynx was suggestive of granulomatous lesion with vasculitis. At this stage a diagnosis of Wegeners granulomatosis was suspected and a pANCA (pANCA dilution 1:10+) was done which was positive. Serum angiotensin converting enzyme levels were normal (12 U/l). A CT guided biopsy from the lung parenchymal lesion was taken which was suggestive of necrotising granulomatous lesion with epithelial cells and langhans giant cells. Thus a diagnosis of Wegeners was made and patient was started on systemic steroid therapy in the form of injectable methylprednisolone (1 g) and cyclophosphamide. Patient showed a significant improvement within two months of initiating treatment. ESR titres decreased (50 mm at 1 h), hearing improved and bilateral facial palsy recovered completely.

Case report 2

A 56-year-old female presented with right sided facial palsy since one year and decreased hearing since one month. She was a known diabetic since three years on treatment. Otoscopy revealed a right intact but retracted tympanic membrane and left tympanic membrane with a central perforation. She had a right grade II facial palsy (House Brackman grading). An audiogram revealed bilateral severe to profound mixed hearing loss. HRCT temporal bone showed bilateral sclerosing mastoiditis (Fig. 3). MR scan brain which had been obtained at an outside institution for facial weakness revealed ill defined patchy hypointense lesion in the central and posterior skull base adjoining the clivus and jugular foramen bilaterally. Laboratory tests revealed a positive pseudomonas growth on culture of the ear swab and an elevated ESR (65 mm at 1 h) and elevated CRP (55.23 mg/l). Urinanalysis and X ray chest were normal. CT chest showed evidence of interstitial lung disease with upper zone predominance. Further investigations revealed a positive serum ANCA antibodies. In view of positive ANCA antibodies a diagnosis of Wegener’s granulomatosis was reached. Patient was commenced on systemic steroid therapy in the form of prednisolone at 60 mg per day. Dramatically over the next two weeks patients general condition improved, ESR fell, hearing in both the ears improved from bilateral severe to profound mixed to bilateral mild to moderate conductive tending to sensorineural hearing loss and facial palsy completely recovered.

Discussion

Wegener’s granulomatosis is a necrotising granulomatous vasculitis with multisystemic involvement. It classically presents with involvement of the upper and lower respiratory tracts and the renal system. The prevalence of ear involvement varies from 19% to 45% of all the cases [4]. Otological involvement may be in the form of serous otitis media, chronic otitis media, sensorineural hearing loss, vertigo and facial palsy [5]. Conductive hearing loss may be secondary to granulation tissue in the middle ear cleft or...
Wegener's granulomatosis is a necrotizing granulomatous vasculitis demonstrated by the classical histopathology. A biopsy specimen from the ear is often small and it is frequently difficult to make a definitive histological diagnosis on this alone. More recently it has been said that a typical or a complete histopathological picture is not always essential for diagnosis of Wegener's. If the clinical picture is suggestive of the diagnosis, a less typical histological picture can be acceptable [6].

Two forms of Wegener's have been described (1) the classical generalised systemic or diffuse form that always involves the kidney and causes necrotising glomerulonephritis (2) the localised or the limited form without the involvement of upper respiratory tract or kidney [9].

Prognosis of Wegener's granulomatosis-induced neuropathies depends mainly on early diagnosis and prompt initiation of medical treatment [6, 12, 13]. In patients with locoregional otological or classical Wegener's granulomatosis who subsequently develop facial nerve palsy, surgical intervention is unrewarding [14]. In addition, biopsy of material from the tympanic membrane or middle ear often shows granulation tissue with chronic unspecific inflammation, and is therefore not diagnostic. Biopsies taken from pulmonary lesions are more specifically diagnostic of the disease [12, 15].

An early diagnosis leads to an early initiation of treatment in the form of steroids and cyclophosphamide which goes a long way in reducing the morbidity and mortality associated with the disease.

Conclusion

Otological manifestations may be the first presenting feature of Wegener's granulomatosis and the patient often presents to an otolaryngologist. These symptoms may often be subtle and diagnosis may be easily overlooked. A high index of suspicion is required to reach to a correct diagnosis. A middle ear disease failing to respond to conventional therapeutic measures, facial nerve paralysis in setting of multisystemic involvement, a consistently raised ESR should alert the clinician to the possibility of Wegener's. Once Wegener's is suspected ANCA levels should be done. Early diagnosis and treatment may delay or prevent the onset of the classical form of the disease and also improve the otological manifestations including facial nerve palsy. It also prevents unnecessary, difficult and potentially hazardous surgical intervention.

Authors' contributions/Wkład autorów

AS – study design, data interpretation, literature search, SD – data collection, literature search, JD – acceptance of final manuscript version.

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Conflict of interest/Konflikt interest

None declared.

Ethics/Etyka

The work described in this article have been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

The own research were conducted according to the Good Clinical Practice guidelines and accepted by local Bioethics Committee, all patients agreed in writing to participation and these researches.

REFERENCES/PIŚMIENNICTWO