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Limited otological eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) – a new disease subset

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Introduction. – Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare, primary vasculitis, characterized by asthma, eosinophilia and extravascular eosinophilic granulomas. Paranasal sinus involvement is common; however otological involvement in EGPA has been described as rare and occurring late in the disease process. We performed a prospective survey of EGPA patients whose primary disease manifestation was otological involvement, in order to determine the frequency and characteristics of this presentation.

Methods. – Otological involvement was defined as the presence of myringitis, otitis media with effusion (OME), chronic otorrhoea, and conductive or sensorineural hearing loss (SNHL) attributable to vasculitis after exclusion of other causes. Sixty patients from a total of 73 with suspected or confirmed EGPA were identified with ENT involvement, and were reviewed prospectively over 36 months. Of these, seven had limited otological disease and are discussed in this series. Presenting features, audiometry, laboratory, histology and radiology results, and disease course were documented.

Results. – Seven patients were identified. All were female and the median age was 50 years. In all cases, otological involvement occurred in the early stages of the disease and led to the diagnosis of EGPA. They presented with copious middle ear and mastoid granulation tissue with conductive hearing loss, tympanic perforations and thick, tenacious otorrhoea. All patients responded to glucocorticoids, with near complete resolution of their symptoms.

Conclusion. – We have described a series of patients with limited EGPA which has predominant otological involvement, which we propose is a novel disease subset. Evidence from their presentations supports this categorisation. It is likely that there is under diagnosis of this condition. Greater awareness of limited EGPA in the otolaryngological community would permit earlier diagnosis and treatment, better hearing outcome, and the prevention of progression to permanent sequelae.

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The ear, nose and throat manifestations of eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome)

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Introduction. – Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare, primary, vasculitis that affects small blood vessels. Ear, nose and throat (ENT) symptoms of EGPA often precede the diagnosis by several years, and are common, with the literature reporting ENT involvement at 70–85%. Thus, EGPA patients are often first referred to an ENT surgeon who has a role in recognising and facilitating the diagnosis, treatment and disease monitoring. There has been little focus on ENT involvement in previous prospective EGPA studies. Therefore, our objective was to establish a database of symptoms affecting the ENT system in patients with EGPA.

Methods. – A prospective review of patients with known or suspected EGPA over 18 months was carried out. For each patient, a full ENT history and examination, including otoscopy, anterior rhinoscopy and flexible nasendoscopy was obtained, and pure tone audiometry and tympanometry performed (where indicated).

Results. – Thirty patients were reviewed at a total of 57 visits. The male to female ratio was 1:1, with a median age of 56 years. Twenty-eight of the 30 EGPA patients (93.3%) had ENT involvement. Of these, all had involvement of the nose (100%), 17 of the ear (60.7%), while none had involvement of the throat.

Discussion. – A database of ENT manifestations of EGPA has been accumulated. Otological manifestations of the disease have been found to occur more frequently and earlier than previously reported. Patients with ear involvement of EGPA either represent an under-diagnosed group, or a new subset of EGPA, which features aggressive otological symptoms. Nasal and laryngeal disease was found to occur with frequencies in keeping with the current literature.

Conclusion. – ENT involvement is common in EGPA, and therefore a clearly defined role for the otolaryngologist is required. We have found otological manifestations to occur both more frequently and earlier than previously reported, and to occur independently of systemic features of the disease. This possibly represents a disease subset.

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ANCA-positive and ANCA-negative phenotypes of eosinophilic granulomatosis and polyangiitis (EGPA): Outcome and long-term follow-up of 46 patients from a single Polish centre

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Introduction. – Eosinophilic granulomatosis and polyangiitis (EGPA) is a rare, systemic, necrotizing, small-vessel vasculitis. Recently attention is drawn to the fact that there are two clinically relevant phenotypes of EGPA: ANCA-positive and ANCA-negative. The aim of this study was to evaluate the outcome and follow-up of EGPA patients from one Polish centre according to their ANCA status.

Methods. – A retrospective cohort study of EGPA patients diagnosed in our hospital between 1998 and 2012 with available follow-up data was conducted. Only patients who fulfilled at least four of six criteria of the American College of Rheumatology were enrolled. According to the ANCA status the course of the disease, organ involvement during disease, methods of treatment and outcome were evaluated.

Results. – A total of 46 EGPA patients (30 females, 16 males) were enrolled, of whom 15 were ANCA-positive (32.6%). Exact characteristics and results of the study are presented in Supplementary data.

Discussion. – In our study, organ involvement at the time of diagnosis in both phenotypes was similar to other studies. Both FFS and BVAS at the time of diagnosis did not differ between the two groups and in both patients underwent similar treatment strategies. Final outcomes were similar in both groups. The ANCA-negative patients had higher mean maximal eosinophil count and less frequent kidney involvement and neuropathy. Interestingly, neuropathy in ANCA-positive patients was more resistant to treatment. It can be suspected that in ANCA-negative patients the main mechanism of organ involvement could be eosinophil tissue infiltration and in ANCA-positive a vascular damage.

Conclusion. – If properly treated, EGPA is a disease with good prognosis. Although it is suspected that different patomechanisms are responsible for organ involvement in ANCA-positive and ANCA-negative phenotypes, in both of them, prognosis and outcomes are similar.