Years, and are common, with the literature reporting ENT involvement of EGPA often precede the diagnosis by several years. Ear, nose and throat (ENT) symptoms of EGPA are rare, primary, vasculitis that affects small blood vessels. Ear, nose and throat (ENT) manifestations are discussed in this series. Presenting symptoms, 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.

Discussion—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, 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 56 years. Twenty-eight of them 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.

Discussion.—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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