P158 A CASE OF REFRACTORY PAEDIATRIC COGAN’S SYNDROME TREATED WITH TOCILIZUMAB

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Background/Aims
Cogan’s syndrome is a rare vasculitis, characterised by progressive sensorineural bilateral hearing loss, vestibular symptoms and non-syphilitic interstitial keratitis. We present a paediatric case which was refractory to initial treatment and subsequently successfully treated with Tocilizumab.

Methods
Case report.

Results
A previously well 14-year-old boy presented with sudden onset hearing loss, tinnitus and vertigo. There was no history of new medication, trauma or tick bite. There was no family history of autoimmune disease or genetic hearing losses. On examination his cranial nerves except for vestibulocochlear were grossly intact. He had a steady gait with no cerebellar signs. Otoscopy was unremarkable. Audiology showed a moderate to severe sensorineural hearing loss bilaterally, worse on the right. He had normal type A tympanograms. An autoimmune screen was carried out which showed normal FBC, inflammatory markers, ACE and Chitotriosidase. ANCA, Rheumatoid factor and lyme serology were negative. MRI of the internal auditory meati was normal. He subsequently developed visual disturbance and was diagnosed with bilateral interstitial keratitis. A unifying diagnosis of Cogan’s syndrome was made. An MRI scan of his head, neck and upper thorax looking for evidence of large vessel vasculitis was normal. Genetics looking for evidence of a monogenic autoinflammatory disorder and primary immunodeficiency were negative. He was initially treated with a weaning course of oral prednisolone with improvement in symptoms. Unfortunately, 4 months later had clinical and audiological deterioration in hearing. He therefore received pulsed IV methylprednisolone and commenced subcutaneous methotrexate and adalimumab. 3 months later, there was both clinical and audiological improvement and he started to wean prednisolone. 7 months later, he presented with a 24-hour history of reduced hearing on the right, confirmed on audiogram. He received a further pulse of IV methylprednisolone and a short course of high-dose oral prednisolone, followed by a slowly weaning course. 3 months later, again he felt his hearing had deteriorated and this was confirmed on audiogram, with a 40-decibel loss in his previously good ear. He received two doses of IV methylprednisolone and background steroids were increased to 20mg daily. Due to frequent relapses, adalimumab was changed to IV tocilizumab at 10mg/kg 2-weekly, alongside methotrexate. IV tocilizumab was changed to the subcutaneous route during the COVID-19 pandemic and was tolerated well. His hearing subsequently improved and tocilizumab interval was extended to 3-weekly in Feb 2021. At last review he was stable and successfully transitioned to adult services.

Conclusion
Evidence regarding treatment options in paediatric patients is lacking due to the rarity of the condition and consequent difficulty in arranging high-quality trials. This is the first case report of use of tocilizumab for Cogan’s syndrome in children, highlighting it as a well-tolerated and successful treatment modality.

Disclosure
C. Anderson: None. K. McAllister: None. J. Walsh: None.