

Abstract EFAS/DGA 2007

Clinical findings and imaging in large endolymphatic duct and sac syndrome

Bartel-Friedrich, S., Amaya, B., Rasinski, C., Kösling, S.

Martin-Luther-University Halle-Wittenberg, Halle/S.

Objective: Large endolymphatic duct and sac syndrome (LEDS) is known the most common kind of radiologically detectable inner ear malformations (IEM). However, LEDS is relatively unknown among general radiologists and audiologists. Therefore, we evaluated the incidence of LEDS in the own patient population and aimed to present our experiences regarding imaging findings, clinical presentation and follow-up.

Methods: Based on complete recordings of all patients, indicated and sent from ENT department to radiology between 1994 and 2002, we identified all radiologically diagnosed cases of IEM including LEDS and all patients, in whom an IEM was clinically suspected. The retrospective study included clinical data, HR-CT and MRI.

Results: Among 169 patients, 17 patients (median age: 12 years, 12 females) and 28 ears, respectively, had LEDS. In 10 patients (6%; 15 ears), no other IEM were detected, called isolated LEDS, seven patients showed additional IEM. Audiometric data at time of imaging revealed sensorineural hearing loss (HL) in 21 ears (eight ears with additional conductive component), deafness in 6 ears and normal hearing in 1 ear of 28 ears. In 26 ears with LEDS (two ears with unknown course excluded) seventeen ears showed progressive (P) or fluctuating progressive (FP) HL (ten -71%- of 14 ears with isolated LEDS) and 9 ears showed constant HL (four ears with isolated LEDS). Seven of 11 patients with P-HL or FP-HL had partly repeated sudden HL (eleven ears of 17 ears; seven with isolated LEDS). A trigger for worsening of hearing was found in 6 patients. A correlation between the severity of morphological changes on imaging and the degree of HL could not be detected. Only five young patients underwent imaging within two years after onset of HL. Three patients received a CI.

Conclusion: LEDS might be an underestimated cause of P-HL or FP-HL and repeated sudden HL in children and young adults. Early imaging plays an important role in ensuring the diagnosis

