AUDITORY PHENOTYPE OF INDIVIDUALS WITH INFRATENTORIAL (CLASSICAL) SUPERFICIAL SIDEROSES: A CROSS-SECTIONAL STUDY

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INTRODUCTION

Infratentorial (classical) superficial siderosis (ISS) is a rare neuro-otological disorder resulting from chronic extravasation of blood into cerebrospinal fluid (often due to dural defects) and deposition of iron-degradation product haemosiderin on the surfaces of CNS structures.

Susceptibility-weighted MRI is the reference standard diagnostic modality. Infratentorial structures (cerebellum and brainstem) are most commonly involved, as well as the 8th cranial nerves. Supratentorially, Sylvian fissures can be involved.2,2 (Figure 1).

ISS-related hearing impairment is predominantly downsloping, resembling age-related changes, and of mixed (sensory-end organ/cochlear or neural) origin.3 Central auditory (brainstem and beyond) involvement was described in a case report.4 It is difficult to ascertain the affected segment of the auditory pathway in individuals with ISS due to often small cohort numbers or limited test battery.5

AIM

To phenotype auditory function in a large cohort of ISS and identify the likely involved auditory structures.

METHODS

Permission for the study was obtained from the departmental clinical governance team (as part of clinical audit).

Patients were also invited to participate in a dedicated research study; permission from the NHS Research Ethics Committee was granted (REC 19/LO/1162/AM01).

We reviewed results of auditory assessments of patients with radiologically confirmed diagnosis of ISS between 30/6/2004 and 01/09/2023. Auditory testing took place at the UCLH NHS Foundation Trust, in line with the BSA guidelines.6,7; the results were compared to departmental or published or equipment manufacturers’ norms.6,11 Relevant anatomy, tests procedures and equipment are described elsewhere.8,12 Data were anonymised at extraction.

RESULTS

N=39, 27 (69%) males

• Of 96 ISS patients, 46 (48%) had auditory tests

• 7/46 (15%) cases were excluded due to unilateral assessments (due to cochlear implantation or previous surgery for vestibular schwannoma in 8 cases) or notes not retrieved (2 cases)

• In 5/39 (13%) only pure tone audiometry (PTA) was available for analysis

• In 4/39 (10%) information was retrieved from clinical letters

Each case was reviewed separately for evidence of end-organ sensory, neuronal or central involvement.

Statistical analysis performed using SPSS (v26-28, IBM, Armonk, NY). We tested for association between hearing levels and disease duration (time interval from causative event to test) using Spearman correlation.

Figure 1. Axial susceptibility-weighted MR images with signal loss (consistent with hemosiderin deposits) involving: A: cerebellum (asterisks), 8th cranial nerves (arrows) and brainstem (arrowheads). B: Sylvian fissures (asterisks).

Figure 2. LI/SN-S pattern suggesting of spatial processing difficulties. Abnormal (below 2 standard deviations) values were worst in High Cue and Spatial Advantage domains.

Figure 3. Auditory test findings (n=34). Retrocochlear (neural or beyond) loss was present in n=16, with confirmed cochlear involvement (n=6) or when cochlear loss could not be excluded (n=10). Three patients had bilaterally normal hearing (PTA and at least one other test). Cochlear loss was recorded (n=6) bilaterally, without unilateral retrocochlear involvement (n=2) or inconclusive (n=3) or with no retrocochlear involvement (n=3).

CONCLUSION

Our study included the largest (to date) cohort of ISS patients with auditory assessments.

We demonstrate predominantly retrocochlear origin of ISS-related hearing impairment, with evidence of central auditory dysfunction.

Hearing impairment in ISS may extend proximally beyond the brainstem but further studies are needed to correlate clinical findings with imaging.

References

1. Feamly JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. Brain. 1995;118 (Pt 4):1051-68


