A CROSS-SECTIONAL STUDY OF VESTIBULAR CHARACTERISTICS OF INDIVIDUALS WITH INFRATEORTAL (CLASSICAL) SUPERFICIAL SIDEROSIS

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INTRODUCTION

Infratentorial superficial siderosis (iSS) is a rare neuro-otological disorder. It results 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 (commonly: cerebellum, and brainstem, but also the 8th cranial nerves).1-2 Susceptibility-weighted MRI is the reference standard diagnostic modality (Figure 1). Slowly progressive impairment of hearing and balance are the most common features of iSS. Vestibular dysfunction is described as mixed, of central (cerebellar or brainstem) and peripheral origin. It is difficult to ascertain the site of vestibular involvement in iSS due to small cohort numbers or limited test battery used, as well as paucity of published reports.

METHODS

Permission for the study was obtained from the departmental clinical governance team (as part of a 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 vestibular assessments of patients with radiologically confirmed diagnosis of iSS between 30/6/2004 and 01/09/2023. Vestibular assessments were undertaken at the UCLH NHS Foundation Trust, in line with the BSA guidelines; the results were compared to departmental or published or equipment manufacturers’ norms.8-11 Relevant anatomy, tests performed, equipment used and the departmental norms are described elsewhere.11 Data were anonymised at extraction; each case was reviewed separately for evidence of central, or peripheral, or mixed involvement where:

AIM

To characterise vestibular function in a large cohort of individuals with iSS and attempt to localise the affected segment of vestibular pathway

RESULTS

N = 31 patients had vestibular assessments: 22 (71%) males. Bedside findings were included in 2 cases

<table>
<thead>
<tr>
<th>Vestibular tests</th>
<th>Mean</th>
<th>Median</th>
<th>Standard deviation</th>
<th>Interquartile range</th>
</tr>
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<tbody>
<tr>
<td>Oculomotor*</td>
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<td>4</td>
<td>9</td>
</tr>
<tr>
<td>vHIT</td>
<td>29</td>
<td>15</td>
<td>4</td>
<td>14</td>
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<td>4</td>
</tr>
<tr>
<td>oVEMP</td>
<td>15</td>
<td>10</td>
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<td>8</td>
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<tr>
<td>cVEMP</td>
<td>12</td>
<td>7</td>
<td>3</td>
<td>5</td>
</tr>
</tbody>
</table>

Vestibular findings

- Central+peripheral b/l
- Central-only
- Peripheral-only
- Normal/peripheral b/l

Figure 2. Vestibular tests performed in the cohort. *Abnormal central vestibular oculomotor findings (abnormal findings in at least one: gaze, saccades, smooth pursuit; opsokinetik nystagmus or vestibulo-ocular reflex suppression). Legend: vHIT video head impulse test, o/cVEMP ocular/cervical vestibular evoked myogenic potentials.

Figure 3. Oculomotor findings. *Abnormal gaze findings suggestive of (c) central and (p) peripheral vestibular involvement.

CONCLUSION

First study to analyse vestibular tests from a large cohort of iSS patients.

We demonstrate predominantly central vestibular involvement, in keeping with the imaging findings.

Presence of additional peripheral involvement may suggest antegrade progression of vestibular dysfunction in iSS.

Normal findings suggest the onset of vestibular dysfunction may be preceded by the radiological manifestations of iSS however longitudinal studies are needed.

REFERENCES