insonism has been reported; that patient also had pyramidal signs. A further patient exhibited dopa-responsive tremor and facial impassivity during recovery from a more typical presentation of pontine myelolysis with supratentorial eye movements and tetraparesis. Pathological changes in the basal ganglia have been well documented in typical cases of pontine myelolysis, and it has been suggested that the pontine lesion masks the extra-pontine clinical features. In our case the large lesion in the pons was clinically silent. MRI is clearly the investigation of choice in patients presenting with neurological syndromes associated with myelolysis; subclinical or clinically atypical pontine myelolysis may be more common than is currently realised.

We are grateful to Dr S P Kane for permission to report a patient under his care.

R TINKER, M G ANDERSON, P ANNAND, A KERMOIDE, A E H HARDING* West Middlesex University Hospital, Isleworth, Middlesex.

*University Department of Clinical Neurology, National Hospital for Nervous Diseases, Queen Square, London.

Correspondence to: Dr All Harding, University Department of Clinical Neurology, National Hospital for Nervous Diseases, Queen Square, London WC1N 3BG, United Kingdom.


Visual hallucinations and the choliner-gic system in dementia

Cholinergic deficits are associated with various forms of dementia, including Alzheimer’s and Parkinson’s disease, in which they relate to the degree of cognitive impairment.1 New findings on the chemical pathology of senile dementia of Lewy body type (SDLT) suggest that particularly extensive cholinergic abnormalities in certain neocortical regions are associated with visual hallucinations. SDLT may be the second most common form of dementia in the elderly (after Alzheimer’s disease).2 It is characterised, clinically, by acute presentation with confusion frequently accompanied by hallucinations and, neuropathologically, by the presence of Lewy bodies, particularly in archicortical areas. The relative absence of neocortical neurofibrillary tangles in SDLT also suggests such patients might be more amenable to transmitter replacement therapy.

In 12 SDLT cases with frozen tissue available for biochemical analyses, visual hallucinations had been noted in six at presentation and continued throughout the course of the disease. It is therefore uncertain whether this symptom was not recorded. Choline acetyltransferase (ChAT) activities were reduced in both SDLT subgroups compared with normal levels but were significantly lower in those with hallucinations compared with those without for two of the three cortical areas examined (see Table). Thus in hallucinating cases ChAT activities in parietal and temporal cortex were reduced by 80–85% compared with 50–55% in the non-hallucinating cases. No other neurochemical or neuropathological parameter so far examined (including dopamine, serotonin, cholinergic receptors, Lewy bodies or plaques) divided the two groups.

The suggestion that extensive ChAT loss in SDLT cases with hallucinations may be related to this clinical feature is supported by psychopharmacological data. Thus, although no patient was receiving any anticholinergic medication at the time they presented, drugs such as scopolamine are known in certain cultural3 or medical4 situations to induce hallucinations. Severely degenerate cholinergic neurons innervating certain cortical areas in dementia disorders such as SDLT may similarly give rise to this symptom. If confirmed, this observation is important in relation to the function of cholinergic innervation of human neocortex (innervation of other areas such as hippocampus being involved in memory) and in relation to selecting suitable cases of dementia for cholinergic therapy (those with hallucinations having a more profound cholinergic defect).

Table. Cortical choline acetyltransferase activities* in senile dementia of Lewy body type, mean, SD

<table>
<thead>
<tr>
<th></th>
<th>Parietal</th>
<th>Temporal</th>
<th>Occipital</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>10.8±5.2</td>
<td>8.7±5.6</td>
<td>11.6±3.4</td>
</tr>
<tr>
<td>SDLT</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Without hallucinations</td>
<td>3.1±2.7</td>
<td>1.1±1.7</td>
<td>2.4±2.0</td>
</tr>
<tr>
<td>With hallucinations</td>
<td>1.6±1.4</td>
<td>1.5±1.6</td>
<td>6.0±4.2</td>
</tr>
</tbody>
</table>

*ChAT expressed as nmol/mg protein in Brodmann areas 39 or 40 (parietal), 21 or 22 (temporal) and 17, 18 or 19 (occipital). Case numbers in parentheses.


This case illustrates how Chiari malformations may present at any age and should be considered even in septuagenarians. The onset of symptoms in the eighth decade is extremely unusual, but presentation at the age of 74 has been reported once. Symptoms may precede diagnosis by several years, as in this patient. However, it is unclear why a congenital malformation should suddenly become symptomatic. Several reports have highlighted the role of trauma, degenerative changes in the cervical spine and cerebrovascular disease that may also contribute to neurological deterioration. Palatal paresis, dysphagia, dyspnea and ataxia are well recognised symptoms of type 1 Arnold-Chiari malformations, and recent reports have highlighted the respiratory problems that these patients may encounter. The presence of downbeat nystagmus strongly implied an abnormality at the cranio-cervical junction and this was confirmed. When not, imaging is required. Despite their ages both our patient and the one reported by Hosford and Spector[1] are very active following surgical decompression showing that age alone should not be regarded as a barrier to active management.

DMG HALPIN*, P TRENT, L SYMON*, AE HARDING
University Department of Clinical Neurology and the Ghugh Cooper Department of Neurological Surgery,* National Hospital for Nervous Diseases, Queen Square, London WC1N 3BG United Kingdom


MATTERS ARISING

Ramsay Hunt syndrome: to bury or to praise

"If there is one disease in the neurological literature that is difficult to define and diagnose, it is the cerebellar dysgenesis of Ramsay Hunt."

Radermacher, 1974.

Our recent suggestion that the Ramsay Hunt syndrome (dysmyelination cerebellar ataxia) is a nosologically useful diagnostic category[3] has provoked considerable controversy. We reached this conclusion following a study of 84 cases of progressive myoclonus epilepsy (PME), of which 13 were previously regarded as Ramsay Hunt syndrome. Review and restudy of this material established the diagnosis of mitochondrial encephalomyopathy (MERRF) in 11 of the 13 cases; the remaining two cases were not available for reexamination. Those patients who wish to preserve the Ramsay Hunt syndrome differ widely in their concept of the disorder,[4] which only reinforces our view that the term should be buried.

Tassinari et al recently reported a series of 13 patients with "Ramsay Hunt syndrome" who had onset of myoclonic or tonic-clonic seizures at ages 6 to 15 years with a mild cerebellar syndrome. Family studies suggested autosomal dominant inheritance and muscle biopsies failed to show evidence of mitochondrial disease. Tassinari's patients are different from the cases that we reclassified as MERRF and we agree that they do not have mitochondrial disease. The clinical, electroencephalographic and genetic features of their patients are, however, identical to those of Unverricht-Lundborg disease (Baltic myoclonus) as described by the original authors and the recent definitive studies of Koskiniemi.[5-7] There is no doubt that this disorder occurs outside the Baltic region.[8] Although there is as yet no diagnostic laboratory or clinical criterion for Unverricht-Lundborg disease, the clinical picture is distinctive and a clinical diagnosis can be made with considerable certainty.[9,10] We find the use of the term "Ramsay Hunt syndrome" for such patients is historically inaccurate and diagnostically misleading.

Tassinari et al have emphasised the electroencephalographic features of their patients with normal or mildly slow waking background activity, fast spike-wave discharges, photosensitivity and lack of activation during slow wave sleep.[4] These findings are not different from those of Unverricht-Lundborg disease.[11,12] Indeed, critical study of the EEG patterns in all the PMEs, including MERRF and Unverricht-Lundborg disease, reveals more similarities than differences.[13] Tassinari et al have also described vertex spikes during REM sleep.[14] Unfortunately, REM studies of Unverricht-Lundborg disease have not been reported. These spikes are, however, also seen in MERRF[15] and we suspect they may be common to all the PMEs, much like giant and slow potentials, which they may in fact represent.

Unlike the situation previously, the vast majority of patients with PME can now be accurately diagnosed during life.Whilst occasional undiagnosed patients remain, there is no residual homogeneous group of cases for which the term Ramsay Hunt syndrome is appropriate. Radermacher's frustration at attempting to define the Ramsay Hunt syndrome[16] can at last be put to rest. In retrospect it was a true symptom, with many causes, although we suspect that most reported cases were probably examples of MERRF. Its view as a nosological clinical category has been overtaken by clinical, genetic, biochemical and pathological advances in specific diagnosis.[17]

(We) come to bury, not to praise[18]

SAMUEL F BERKOVIC*, FREDERICK J ANGELMANN
*Department of Neurology, Austin Hospital, Melbourne, Victoria 3084, Australia.
†Montreal Neurological Institute and Hospital, Montreal, Quebec H3A 2B4, Canada.


Tassinari et al reply: We thank Drs Berkovic and Andermann for the kind comments they made on our recent paper. We are glad to know that the patients previously referred to as Ramsay Hunt syndrome (RHS) by these authors and subsequently found to have a mitochondrial encephalopathy (MERRF) were significantly different from those we have described under the eponym of RHS. This fact supports our statement that RHS and MERRF are different clinical, EEG and evolutive features.

Drs Berkovic and Andermann however criticise the term RHS applied to our cases. In these patients the main clinical complaint was action myoclonus combined with rare generalised epileptic seizures: indeed this association was described by Ramsay Hunt and later by the "Dysmyelination Cerebellar Ataxia". Ramsay Hunt also emphasised the coexistence of "cerebellar ataxia" but, in such cases, the cerebellar component is difficult to define because of the presence of severe intention myoclonus, as recently pointed out by Harding. Thus, in our opinion, the use of the term RHS for our patients is justified,