improvement in neurological symptoms, indicating that the cytomegalovirus infection was associated with her neurological disease. The serology of Bickerstaff's brainstem encephalitis is still unclear. A relation with herpes simplex virus infection has been noted, but no patients with Bickerstaff's brainstem encephalitis associated with cytomegalovirus infection have been reported.

With regard to the pathogenesis of Bickerstaff's brainstem encephalitis, an immune mechanism has been considered. In one patient, the presence of serum anti-GQ1b antibody, which is common in Fisher's syndrome, indicated that humoral autoimmune mechanisms, common to Fisher's syndrome, function in the development of Bickerstaff's brainstem encephalitis. The typical signs of meningoencephalitis—namely, fever at the onset of neuritic symptoms, meningeal irritation and CSF pleocytosis—and detection of cytomegalovirus DNA in the CSF may indicate the involvement of cytomegalovirus infection. Both cytomegalovirus infection and a post-infection autoimmune mechanism may have caused clinical symptoms in this patient.

This research was supported in part by grants in aid from the Uehara Memorial Foundation.

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Raymond syndrome (alternating abducens hemiplegia) caused by a small haematoma at the medial pontomedullary junction

Raymond syndrome is characterised by ipsilateral abducens nerve palsy and contralateral hemiplegia. Pure Raymond syndrome is extremely rare, as many nuclei and fibres exist near the root fibres of the abducens nerve. This is the first report in which the precise localisation of a pure form of Raymond syndrome was determined by MRI.

A 39 year old man awoke with horizontal diplopia, especially on right lateral gaze. Five days later, a Hess chart examination performed by an ophthalmologist showed a paresis of the right lateral rectus muscle. On admission 19 days after onset, the patient showed a mild paresis of the right abducens nerve and a subtle weakness of his left leg with moderate hyper-reflexia in the left upper and lower limbs. The Babinski reflex was positive and the abdominal reflex was absent on the left side and the Babinski reflex was negative and the abdominal reflex was positive on the right side. No facial weakness or deviation of the tongue on protrusion was found. All other general and neurological examinations were normal. Routine blood and urine examinations were normal. Evaluations of short latency somatosensory evoked potentials to posterior thalamic nerve stimulation, brainstem auditory evoked potentials, and blink reflex proved normal. Head CT was normal, but a brain MRI done 31 days after onset showed two punctate high signal intensity spots surrounded by low signal intensity areas at the right pontomedullary junction (arrows). 

(A) Axial (TR = 2500 ms, TE = 110 ms) and (B) sagittal (TR = 2500 ms, TE = 100 ms) MRI sections. There are two small high signal intensity spots surrounded by low signal intensity areas at the right pontomedullary junction (arrows).

Pupillary dilatation and arm weakness as negative ictal phenomena

Transient ictal hemiplegia is an uncommon feature of epileptic attacks that were classified by Gastaut and Broughton as unilateral atonic seizures. The present case was of particular interest because hemiplegia was accompanied by dilatation of the pupil on the side of the hemiplegia. A boy aged 9 years had a history of episodic weakness of his left upper and lower limbs, sometimes preceded by a sensation like a dog paws pinching the ictal side since the age of 5. His mother said that he would stare and his left arm then dropped limply to his side while his left leg became weak for about 10 to 40 seconds. During this period his left pupil dilated. In some episodes his left eyelid fluttered and the left side of his mouth turned up and his left arm and leg remained weak. The attacks increased in frequency until he was having two to eight each day, but subsided to once daily when carbamazepine treatment was started. There was no history of head injury or other relevant illness and no family history of epilepsy. His EEG showed an almost continuous sharp and slow wave discharge arising in the right parietal region. Brain CT was normal but MRI four years later showed a hypointense area involving both grey and white matter in the right parietal lobe; there was no mass effect or evidence of blood products surrounding the lesion.

At the age of 13 he underwent craniotomy and electrocorticography in his right hemisphere and was certain of an epileptic focus in the area surrounding an atrophic gyrus in his right parietal cortex. The abnormal area was then excised. The histology report (Dr W A Evans) concluded that “I find this lesion hard to classify. It is most likely a hamatoma, possibly of a similar nature to the focal dysplasia of the cerebral cortex described by Taylor et al.”

There was no postoperative neurological deficit and he was free of seizures until eight months later when his carbamazepine dosage was reduced from 1000 mg to 400 mg daily. Three years after stopping his carbamazepine dose was again reduced, when he had a recurrence of daily attacks of fluttering of his left eyelid and weakness of his left arm, but not the left leg, lasting 20 seconds. His EEG showed focal right parietal slow activity without epileptogenic features. Since then he has been subject to episodes about every 10 days with dilatation of the left pupil, weakness of the left arm, and some twitching of the left side of his face lasting about 10-20 seconds. He has never had any jerking or involuntary movement of his left arm.

Construction of the left pupil in association with hallucinations projected into the left visual field was reported as an ictal phenomenon by Lance and Smee and
attributed to the excitation of a predomi-
nantly crossed occipitopretectal tract.1 Cogan stated that *removal of the pupillo-
constrictor zone in one occiput of the cat
results in anosmia with the larger pupil on
the opposite side*.1 This finding presum-
ably explains the unilateral pupillary dilata-
tion reported here as a negative ictal
phenomenon.

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Giall cytoplasmic inclusions are not exclusive to multiple system atrophy

In 1989 Papp et al1 reported finding argy-
ophilic inclusions in the cytoplasm of
oligodendrocytes in cases of multiple system
atrophy, and their presence in the sporadic
form of this condition has since been con-
firmed.2 The value of giall cytoplasmic inclusions as a diagnostic hallmark of mul-
tiple system atrophy has been emphasised
by one of the authors3 as well as by others.7

At the UK Parkinson’s Disease Society
Brain Bank in London, tissue is donated by
patients with principally movement disor-
ders. Giall cytoplasmic inclusions occurred in
all brains from patients with multiple sys-
tem atrophy (total 56); however, in three of
seven cases with a pathological diagnosis of
corticobasal degeneration and two of 18 cases
with Steele-Richardson-Olszewski syndrome similar intracytoplasmic oligo-
dendrocyte inclusions were identified. These were filamentous argyrophilic struc-
tures (figure) immunoreactive with tau and
ubiquitin antiserum. In corticobasal degen-
eration they were most numerous in white
matter underlying the affected cortex, in the
corpus callosum, internal capsule, and in one
case, the basis pedunculi; occasional similar
inclusions were also identified in the af-
fected cerebral cortex and in the brain stem
as well as in cerebellar hemispheric white
matter, in the absence of any neu-
ronal abnormalities. In the cases of Steele-
Richardson-Olszewski syndrome inclusions
were most prominent in the cerebellar white
matter. We have not counted or mapped
the distribution of giall inclusions in our
cases, but have the impression that they are
less numerous than in multiple system atrophy.

These findings have important implica-
tions for histological diagnosis and our under-
standing of disease pathogenesis. There
is increasing awareness of overlap
between many neurodegenerative condi-
tions, in particular those associated
with parkinsonism; thus the Lewy body, Pick
body, neurofibrillary tangle, or the giall
cytoplasmic inclusion are not exclusive
to any of the conditions in which they abound. One explanation may be that neurons and

Floral neglect body-centric?

One theory of unilateral visual neglect pro-
poses that it results from disruption of represen-
tations of space. But what exactly is the nature
of the spatial map that is dis-
rupted? Is it retinotopic, head-centric,
body-centric, mapped with respect to
gy or even possibly object centred?
Many of those who have been attracted
by representational hypotheses have suggested
that it may be body-centric. In other words,
the hemisphere that patients with left sided
visual neglect fail to attend to is that to
the left of the body sagittal midline.

Evidence in favour of a disruption of body-centric (or socalled egocentric) spatial
representation has been presented from
our measurements of sacadic latency to briefly
illuminated targets with the head turned at
various angles with respect to the trunk.1

Furthermore, Heilman and Valenstein have
shown that line bisection is more accurate
when the task is presented to the right of
the body midline.2 Cancellation tasks are anoth-
er way of assessing neglect. If left sided
visual neglect is body-centric there should
be amelioration, or even complete absence,
of neglect when the task is performed in the
hemispace right of the body midline.

Eight right handed patients presenting
acutely with visu 45° right were examined.
All of them had left sided visual neglect on
the day of presentation; some also had left
sided hemiplegia or somatosensory loss.
None of the patients were considered to
have a substantial visual field loss on clinical
examination at the bedside. (Assessment of
the left half of the visual field was aided by
cueing attention, but not gaze, to the left.
Patients were asked to fix their gaze on
the author and simultaneously encouraged to
say whether relatively large objects—for
example, flowers—on the left were being
moved. Once patients were accustomed to
this task, the flowers were held stationary at
the edge of the left visual field and patients
were asked to keep attending towards the
flowers. A hatpin was then used to map
visual fields, but patients needed to be
reminded constantly to attend to the left. I
was eventually able to do up to 60 such
trials for each patient. All the patients
reported here could see a moving hatpin in
each quadrant of the visual field. All eight patients had CT per-
fomed within five days of presentation.
Evidence of cortical infarction involving the
right parietal cortex, or frontal cortex, or both
was found on all of the tomograms (table).

Each patient was asked to perform the
 cancellation task devised by Weintraub and
Mesulam.3 Patients were first shown the tar-
get shape (a circle with eight spokes and a
diagonal running through the circle) on
a small piece of card. They were then pre-
sented with the task, which has 60 such
targets disposed among 318 distractor shapes
on an A4 sheet of paper. Thirty targets
are present in each half of the sheet, so
the maximum score is 60. Patients were asked
to ring all the targets visible to them. No
time limit was imposed.

The task was first presented on a table
directly in front of the patients so that in
this condition the head and body midlines
were aligned. Patients were instructed to let
the experimenter know when they thought
that they had completed the task. After a
short break, the task was presented on the
table in the body midline. Patients performed
the second trial with the head turned 45° to the right and the trunk
held still in the original position. The

The mean cancellation score when head
and trunk were aligned was 6-7 (SD 4-7)
items; when the head was turned to the right
it was 5-9 items (SD 5-2). There was no sig-
ificant difference in performance between
these two conditions (paired t = 1-1, df =
14, p = 0.3). Thus patients performed just
Pupillary dilatation and arm weakness as negative ictal phenomena.

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*J Neural Neurosurg Psychiatry* 1995 58: 261-262
doi: 10.1136/jnnp.58.2.261-a

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