improvement in neurological symptoms, indicating that the cytomegalovirus infection was associated with her neurological disease. The aetiology of Bickerstaff's brainstem encephalitis is still unclear. A relation with herpes simplex virus infection has been noted,[4] 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.[1-4] In this patient, the presence of serum anti-GQ1b antibody, which is common in Fisher's syndrome,[5] indicated that humoral autoimmune mechanisms, common to Fisher's syndrome, function in the development of Bickerstaff's brainstem encephalitis.[1] 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.

AKIHIRO KANZAKI
SEISO YABUKI
Department of Neurology, Kochi Municipal Central Hospital, Kochi, Japan
NOBUHIRO YUKI
Department of Biochemistry, Tokyo Medical and Dental University, Tokyo, Japan

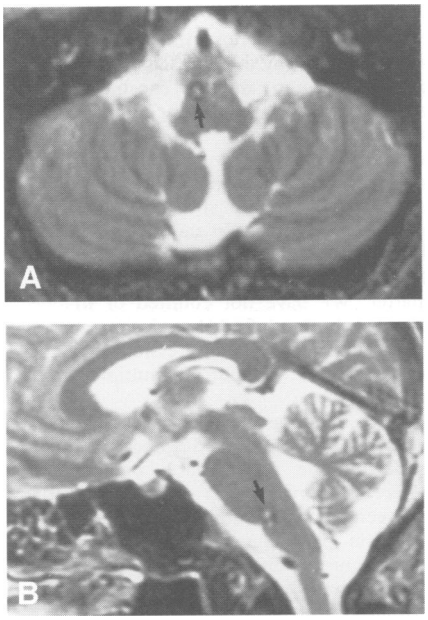
Correspondence to: Dr Akihiro Kanzaki, Department of Neurology, Kochi Municipal Central Hospital, 2-7-33 Sakurai-cho, Kochi 780, Japan.

1 Bickerstaff ER. Brain stem encephalitis (Bickerstaff's encephalitis). In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology*. Amsterdam: North Holland 1978;**34**: 605–9.
2 Yuki N, Sato S, Tsuji S, Hozumi I, Miyatake T. An immunologic abnormality common to Bickerstaff's brain stem encephalitis and Fisher's syndrome. *J Neurol Sci* 1993;**118**: 83–7.
3 Al-Din AN, Anderson M, Bickerstaff ER, Harvey I. Brainstem encephalitis and the syndrome of Miller Fisher: a clinical study. *Brain* 1982;**105**:481–95.
4 Maran AGD. Bickerstaff's encephalitis. *J Laryngol* 1966;**80**:520–3.
5 Chiba A, Kusunoki S, Shimizu T, Kanazawa I. Serum IgG antibody to ganglioside GQ1b is a possible marker of Miller Fisher syndrome. *Ann Neurol* 1992;**31**:677–9.

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

Raymond syndrome[1] 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 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 tibial 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 medial pontomedullary junction on both the T1 and T2 weighted images (figure). Vertebral angiography showed no abnormality. Thus the lesion was probably produced by a haemorrhage from a cavernous haemangioma at the pontomedullary junction.



(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).

Both Millard-Gubler syndrome (facial palsy and contralateral hemiplegia) and Raymond syndrome are well known to induce crossed paralysis due to a caudal pontine lesion. The pure form of either syndrome has, however, rarely been reported. The lesion producing the pure Millard-Gubler syndrome[2] is located more laterally than seen in our patient, whereas that producing isolated abducens nerve palsy[3] is located more dorsally. As the haemorrhage was restricted to the ventral and medial pons, our patient was considered to show pure Raymond syndrome.

MARIE SATAKE
JUN-ICHI KIRA
TAKESHI YAMADA
TAKURO KOBAYASHI
Department of Neurology, Neurological Institute, Faculty of Medicine, Kyushu University, Fukuoka 812, Japan

1 Toppel VL. Strabismus paralyticus als Teilerscheinung eines doppelseitigen Raymond-Syndroms. *Klin Monatsbl Augenheilkd* 1967; **151**:663–72.
2 Yasuda Y, Matsuda I, Sakagami T, Kobayashi H, Kameyama M. Pontine infarction with pure Millard-Gubler syndrome: precise localization with magnetic resonance imaging. *Eur Neurol* 1993;**33**:333–4.
3 Donaldson D, Rosenberg NL. Infarction of abducens nerve fascicle as cause of isolated sixth nerve palsy related to hypertension. *Neurology* 1988;**38**:1654.

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.[1] 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 shivering" in his head 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 time 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 hyperintense 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 confirmed the presence 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 hamartoma, possibly of a similar nature to the focal dysplasia of the cerebral cortex described by Taylor *et al*".[2]

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 the operation 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.

Constriction 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

J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.58.2.261-a on 1 February 1995. Downloaded from http://jnnp.bmj.com/ on September 23, 2021 by guest. Protected by copyright.

attributed to the excitation of a predominantly crossed occipitopretectal tract.[3] Cogan stated that "removal of the pupillo-constrictor zone in one occiput of the cat results in anisocoria with the larger pupil on the opposite side".[4] This finding presumably explains the unilateral pupillary dilatation reported here as a negative ictal phenomenon.

J W LANCE
Institute of Neurological Sciences, Prince of Wales Hospital, Sydney, Australia

Correspondence to: Professor J W Lance, Suite 5A, 66 High Street, Randwick, NSW 2031, Australia.

1 Gastaut H, Broughton R. *Epileptic seizures, clinical and electrographic features, diagnosis and treatment.* Springfield, IL: Charles C Thomas, 1972:109.
2 Taylor DC, Falconer MA, Bruton CJ, Corsellis JAN. Focal dysplasia of the cerebral cortex in epilepsy. *J Neurol Neurosurg Psychiatry* 1971;**34**:369–87.
3 Lance JW, Smee RI. Partial seizures with visual disturbance treated by radiotherapy of cavernous hemangioma. *Ann Neurol* 1989; **26**:782–5.
4 Cogan DG. *Neurology of the ocular muscles.* Springfield, IL: Charles C Thomas, 1948: 115.

Glial cytoplasmic inclusions are not exclusive to multiple system atrophy

In 1989 Papp *et al*[1] reported finding argyrophilic 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 confirmed.[2] The value of glial cytoplasmic inclusions as a diagnostic hallmark of multiple system atrophy has been emphasised by one of the authors[3] as well as by others.[2]

At the UK Parkinson's Disease Society Brain Bank in London, tissue is donated by patients with principally movement disorders. Glial cytoplasmic inclusions occurred in all brains from patients with multiple system 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 oligodendrocyte inclusions were identified. These were filamentous argyrophilic structures (figure) immunoreactive with tau and ubiquitin antiserum. In corticobasal degeneration 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 affected cerebral cortex and in the brain stem as well as in cerebellar hemispheric white matter, in the absence of any neuronal abnormalities. In the cases of Steele-Richardson-Olszewski syndrome inclusions were most prominent in cerebellar white matter. We have not counted or mapped the distribution of glial inclusions in our cases, but have the impression that they are less numerous than in multiple system atrophy.

Frontal hemispheric white matter; corticobasal degeneration with argyrophilic cytoplasmic inclusions in oligodendrocytes (some arrowed); modified Bielschowsky originally × 400.

These findings have important implications for histological diagnosis and our understanding of disease pathogenesis. There is increasing awareness of overlap between many neurodegenerative conditions, in particular those associated with parkinsonism; thus the Lewy body, Pick body, neurofibrillary tangle, or the glial cytoplasmic inclusion are not exclusive to any of the conditions in which they abound. One explanation may be that neurons and glia have a limited repertoire of responses to a variety of different stimuli, resulting in morphological similarities between clinically distinct neurodegenerative diseases. Alternatively, shared pathogenetic pathways may underlie the cytoskeletal abnormalities seen in these conditions, the exact pattern of pathology being dictated by host factors such as age of exposure and genotype.

S E DANIEL
J F GEDDES
T REVESZ
Parkinson's Disease Society Brain Bank, Institute of Neurology, 1 Wakefield St, London WC1N 1PJ, UK

Correspondence to: Dr S E Daniel.

1 Papp MI, Kahn JE, Lantos PL. Glial cytoplasmic inclusions in the CNS of patients with multiple system atrophy (striatonigral degeneration, olivopontocerebellar atrophy and Shy-Drager syndrome). *J Neurol Sci* 1989;**94**:79–100.
2 Lantos PL, Papp MI. Cellular pathology of multiple system atrophy: a review. *J Neurol Neurosurg Psychiatry* 1994;**57**:129–33.
3 Daniel SE. The neuropathology and neurochemistry of multiple system atrophy. In: Bannister R, Matthias CJ, eds. *Autonomic failure*, 3rd ed, Oxford: Oxford University Press, 1992.

Is visual neglect body-centric?

One theory of unilateral visual neglect proposes that it results from disruption of representations of space. But what exactly is the nature of the spatial map that is disrupted? Is it retinotopic, head-centric, body-centric, mapped with respect to gravity, 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 hemispace 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 measurements of saccadic 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 another 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 visual neglect 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 convince myself that all the patients reported here could see a moving hatpin in each quadrant of the visual field.) All eight patients had CT performed 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 target shape (a circle with eight spokes and a diagonal running through the circle) on a small piece of card. They were then presented 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 at 45° right of 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 table shows the results of the experiment. As expected, patients cancelled items only on the right half of the cancellation sheet (by contrast with patients with only visual field loss, who are able to cancel targets on both sides of the paper).

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 significant difference in performance between these two conditions (paired $t = 1·1$, $df = 14$, $p = 0·3$). Thus patients performed just

J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.58.2.261-a on 1 February 1995. Downloaded from http://jnnp.bmj.com/ on September 23, 2021 by guest. Protected by copyright.