

Section 1

Diagnosis

Case

1

First seizure: is it epilepsy?

Christophe Rauch and Hermann Stefan

History

A 27-year-old man went to a supermarket in the afternoon when he remarked on blurred vision and then lost consciousness. Motoric relinquishings were observed by other customers. The young man regained consciousness upon the arrival of the emergency physician. The physician detected a tongue bite and measured a low blood glucosis level of 45 mg/dl. The patient was referred to the emergency room of the neurological department.

The patient told that this was the first seizure he had suffered. Furthermore, he told that he had not eaten much that day. There were no other diseases known in this patient, no familiar disposition.

Actual treatment

None

What to do?

Further investigations

- EEG: Alpha-EEG, no epileptiform activity
- **Blood samples:** Blood glucosis 62 mg/dl, no further noticeable values
- MRI: normal finding

Diagnosis

Hypoglycemia with seizure.

Treatment

Treatment of the hypoglycemia and research for reasons for the hypoglycemia. Further investigations for Diabetes mellitus, insulinoma, and further differential diagnosis.

General remarks

According to the consensus definition of seizure and epilepsy by The International League Against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE), an epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.

Special remarks

Hypoglycemia can be one cause for seizures especially if a seizure occurs in the morning before breakfast or after exercise. Therefore, the reason for hypoglycemia needs to be resolved. One misdiagnosis can be an insulinoma which is one of the most common hormone-secreting tumors of the gastrointestinal tract. It causes hypoglycemic phases with symptoms concerning the autonomic system such as sweating, tremor, anxiety, etc. and symptoms concerning the central nerve system such as confusion, lethargy, bizarre behavior, and cognitive troubles, etc. These symptoms can be quite similar to epileptic seizures. Epilepsy can be already diagnosed if only one spontaneous seizure occurs, if signs for an enduring predisposition such as spike-wave activity in the EEG coincide. This was not the case in our patient.

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Section 1: Diagnosis

Suggested reading

Fisher RS, van Emde Boas W, Blume W, <i>et al.</i> Epileptic seizures and epilepsy: definitions proposed by the International League Against Epilepsy (ILAE) and the International Bureau for	Epilepsy (IBE). <i>Epilepsia</i> 2005; 46 : 470–2.	<i>J Neurol Neurosurg Psychiatry</i> 2004; 75 : 1091–2.
	Graves TD, Gandhi S, Smith SJM, <i>et al.</i> Misdiagnosis of seizures: insulinoma presenting as adult onset seizure disorder.	Pohlmann-Eden B, Beghi E, Camfield C, Camfield P. The first seizure and its management in adults and children. <i>BMJ</i> 2006; 332 : 339–42.

Section 1

Diagnosis

Case

2

Intractable epilepsy and epilepsia partialis continua associated with respiratory chain deficiency

Francesco Mari and Renzo Guerrini

Clinical history

A 5-year-old boy with a history of severe developmental delay and dyskinetic quadriplegia was admitted to our ward after onset of a cluster of convulsive seizures associated with epilepsia partialis continua involving the right facial and shoulder muscles (Fig. 1).

General history

There was no relevant family history of neurological or systemic disorders.

Examination

Dyskinetic quadriplegia. Severe developmental delay. No dysmorphic features.

Image findings

Several brain MRI scans were performed during follow-up. The first scan, 3 days after the admission, showed a left temporo-parieto-occipital ischemic lesion (DWI positive) as well as atrophy brain changes, involving both the cortical and subcortical structures (Fig. 2). A repeat MRI scan, 10 days later, revealed additional ischemic lesions in the frontal lobes.

Follow-up

Epilepsia partialis continua persisted despite treatment with multiple antiepileptic drugs in the intensive care unit. As clinical and brain MRI findings were consistent with a mitochondrial disorder, we performed muscle biopsy.

Special studies

Muscle biopsy with biochemical study of the respiratory chain revealed a deficit of complex I and III activity.

Diagnosis

Mitochondrial encephalopathy (complex I–III deficit) with epilepsia partialis continua.

General remarks

Clinical presentation of respiratory chain defects in childhood causes a large variety of clinical symptoms. In general, a diagnosis of respiratory chain defect is difficult to consider when the initial symptoms occur. Epilepsy is frequently reported with different clinical characteristics including neonatal onset of status epilepticus, Ohtahara syndrome, West syndrome, epilepsia partialis continua, and Alpers syndrome. Appearance of a worsening epileptic syndrome often correlates with a clinically relevant overall worsening of mitochondrial pathology.

Special remarks

Epilepsia partialis continua, i.e., continuous focal jerking involving a body part, is related to a fixed or progressive lesion involving the motor cortex. Several studies have highlighted the relevance of this peculiar epileptic pattern in children with vascular lesions secondary to mitochondrial disorders.

Future perspectives

Despite the increasing knowledge of the molecular bases of mitochondrial encephalomyopathies, no specific treatment for these disorders is available yet. Palliative/supportive measures and metabolic therapies are useful in ameliorating specific problems and quality of life of patients. Gene therapy is a promising approach, but the clinical relevance of its use remains elusive.

Section 1: Diagnosis

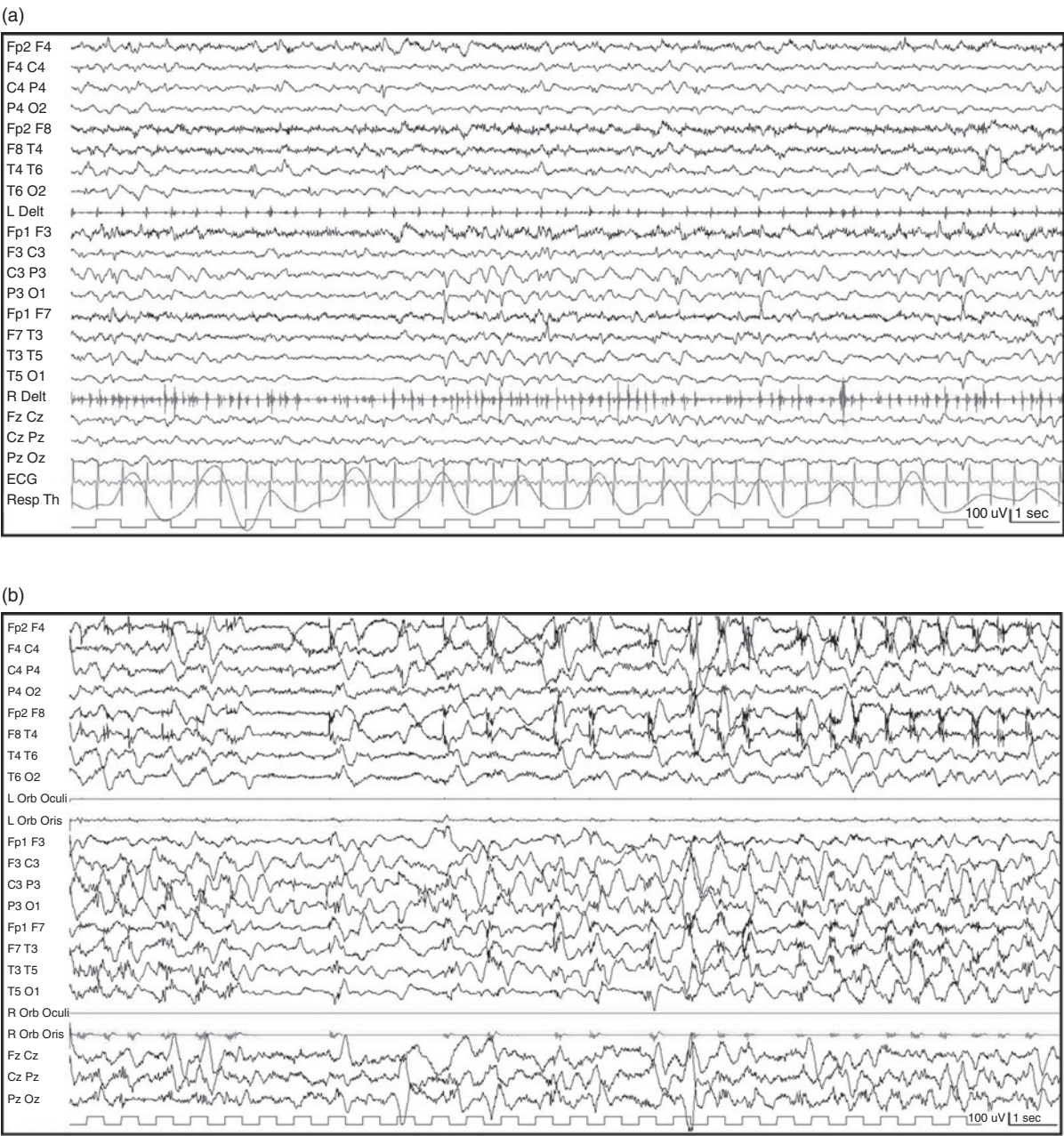


Fig. 1(a), (b). Polygraphic video-EEG recordings: diffusely slow background activity with superimposed rhythmic spikes and slow-waves over the left hemisphere, with centro-parietal predominance. Continuous myoclonic potentials are recorded over the right deltoid and also involve the right orbicularis oris.

Case 2: Intractable epilepsy and epilepsia partialis continua associated with respiratory chain deficiency

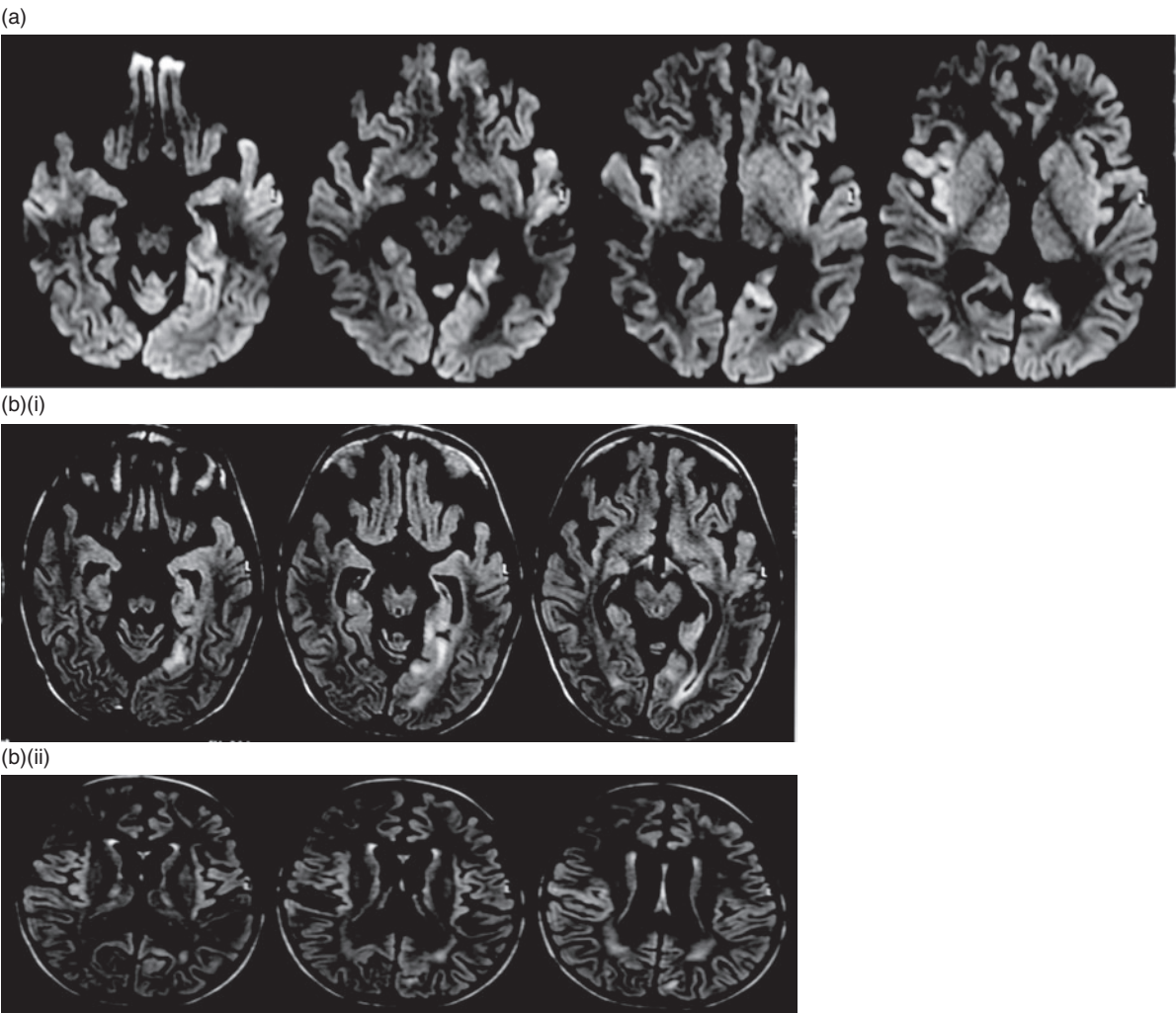


Fig. 2. Axial brain MRI images (a) DWI and (b) FLAIR: subacute left temporo-occipital vascular lesion and mild diffuse cortical and subcortical atrophy.

Suggested reading

Cockerell OC, Rothwell J, Thompson PD, Marsden CD, Shorvon SD. Clinical and physiological features of epilepsia partialis continua. Cases ascertained in the UK. *Brain* 1996; **119**: 393–407.

DiMauro S, Schon EA. Mitochondrial respiratory-chain

diseases. *N Engl J Med* 2003; **348**: 2656–68.

Riquet A, Auvin S, Cuisset JM, *et al.* Epilepsia partialis continua and defects in the mitochondrial respiratory chain. *Epilepsy Res* 2008; **78**: 1–6.

Sabbagh SE, Lebre A, Bahi-Buisson N, *et al.* Epileptic phenotypes in

children with respiratory chain disorders. *Epilepsia* 2010; **51**: 1225–35.

Veggiotti P, Colamaria V, Dalla Bernardina B, *et al.* Epilepsia partialis continua in a case of MELAS: clinical and neurophysiological study. *Neurophysiol Clin* 1995; **25**: 158–66.

Section 1

Diagnosis

Case

3

Reasons for violent behavior – when a man strangles his wife

Frank Kerling and Hermann Stefan

Clinical history

A 44-year-old patient was suffering from epilepsy with complex partial seizures since the age of 12 and he was being treated with ethosuximide and phenytoin. Under this drug regimen the patient was seizure free during day-time, but there were nocturnal episodes of walking around, opening/closing doors and afterwards falling asleep again. The patient had no memory of the event and his wife reported that her husband was not reacting when she talked to him. One night she tried to hold him, because he wanted to leave the house. In this situation he tried to strangle her and she and her daughter were extremely scared. Therefore she thought about divorce and the patient had to sleep in the cellar with the door locked at night-time. He was very desperate and remorseful because of the situation and was admitted to our hospital for diagnostic reasons.

Examination

Neurological and psychiatric examinations were without any pathological findings except for a somehow depressed mood because of the familial situation.

Special studies

In video-EEG monitoring one nocturnal complex-partial seizure was detected. The patient showed nose wiping, oral automatisms, rocking body movements, and an increased muscle tone in the right face and arm. Initially, there was a right frontotemporal 6 Hz pattern and after 34 seconds the pattern propagated to the left temporal lobe. The seizure was followed by a 15-minute state of confusion, wandering, and aggressiveness. Afterwards, the patient was not able to remember the seizure or

the confusion. MRI was normal and interictal SPECT showed hypoperfusion left temporal.

Follow-up

We informed the patient and his wife that he was suffering from nocturnal seizures and that the violence was due to postictal confusion. Therefore, we optimized pharmacotherapy and started a new combination with oxcarbazepine and levetiracetam. Ethosuximide and phenytoin therapy were stopped or tapered down, respectively. The patient was almost seizure free with one seizure every 6 months and the patient and his family were satisfied with this result.

Diagnosis

Postictal confusion with aggressive behavior.

General remarks

Postictal aggression and violence against others are pretty rare with a percentage from 5% to 10% of all postictal confusional states. There is clinical heterogeneity among patients with postictal violence with respect to etiology of epilepsy, age of onset, laterality, and memory of adverse behaviors. Several clinical features, including male gender, are common. Typically, the episodes of postictal aggression are not isolated events, but recur repeatedly often with stereotyped behaviors. Subacute postictal aggression is even more likely after a cluster of seizures than after a single ictus. Aggression occurs especially when relatives or hospital staff try to restrain the patient. Most of the patients have medically intractable epilepsy and are remorseful in the interictal period like our patient was.

Case 3: Reasons for violent behavior – when a man strangles his wife

Special remarks

Our patient showed abnormal violence during a typical state of nocturnal confusion, when his wife tried to restrain him. He had been suffering from epilepsy for decades and was seizure free during the day-time. A postictal aggression was more probable than somnambulism. There is evidence that nocturnal

seizures, especially without tonic–clonic movements, can be missed by the partner and only the confusion is noticed. The results of the video–EEG helped the patient and his family to understand the context between seizures and confusion and the familial situation was stabilized. A modified drug therapy led to sufficient seizure control.

Suggested reading

Gerard ME, Spitz MC, Towbin JA, Shantz D. Subacute postictal aggression. *Neurology* 1998; **50**: 384–8.

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Section 1

Diagnosis

Case

4

Repetitive monocular eye adduction

Adam Strzelczyk, Sebastian Bauer, and Felix Rosenow

Clinical history

A 24-year-old student presented with recurring episodes of involuntary eye movements without disturbance of consciousness for the last 3 weeks. He reported suffering from 5 to 8 attacks per day with an initial feeling of slightly impaired vision followed by adduction of the left eye only lasting for up to 10 seconds. For the last 5 days he had also noticed hypoesthesia of the left hand during these episodes. For that reason, he presented to the neurological emergency department.

Examination

At the physical, neurological, and ophthalmologic examination including visual fields, no abnormalities were detected. He had no history of febrile convulsions, seizures, or any other risk factors for epilepsy. An episode as described above was observed by the treating physician and an EEG with video recording was recommended.

Special studies

During a 2-hour video-EEG monitoring two right frontal EEG seizures (max. F4/C4) of up to 17 seconds duration were recorded, all of which were accompanied by an adduction of the left eye and symmetrical head nodding. No other ictal or postictal symptoms were observed. Visual evoked potentials were within normal limits (Fig. 1a,b).

Image findings

3T MRI sagittal FLAIR images showed a right frontal focal cortical dysplasia. No other focal lesions were detected (Fig. 2).

Follow-up

Initial treatment with levetiracetam up to 4000 mg daily led to a decrease in seizure frequency by half. With a change of anticonvulsive treatment to carbamazepine 400 mg daily, a sustained seizure freedom of more than 2 years could be achieved.

Diagnosis

Symptomatic focal right frontal epilepsy.

Seizures:	simple motor seizures -> somatosensory auras
Cause:	right frontal cortical dysplasia
Related medical conditions:	none

General remarks

Monocular nystagmus is a rare and heterogenous neurological phenomenon. It is usually not of epileptic origin and has been reported in cases of anterior visual pathway lesions, monocular blindness, spasmus nutans, and brainstem lesions associated with stroke or multiple sclerosis. Epileptic binocular nystagmus remains also an infrequent semiological finding and should not be confused with the epileptic gaze deviation. The physiology of binocular epileptic nystagmus is incompletely understood. Kaplan and colleagues postulated that epileptic nystagmus is mediated by a seizure focus in a cortical saccade region contraversive to both quick and slow phases of nystagmus.

Grant and colleagues reported a case of epileptic monocular nystagmus with seizures originating in the

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Case 4: Repetitive monocular eye adduction

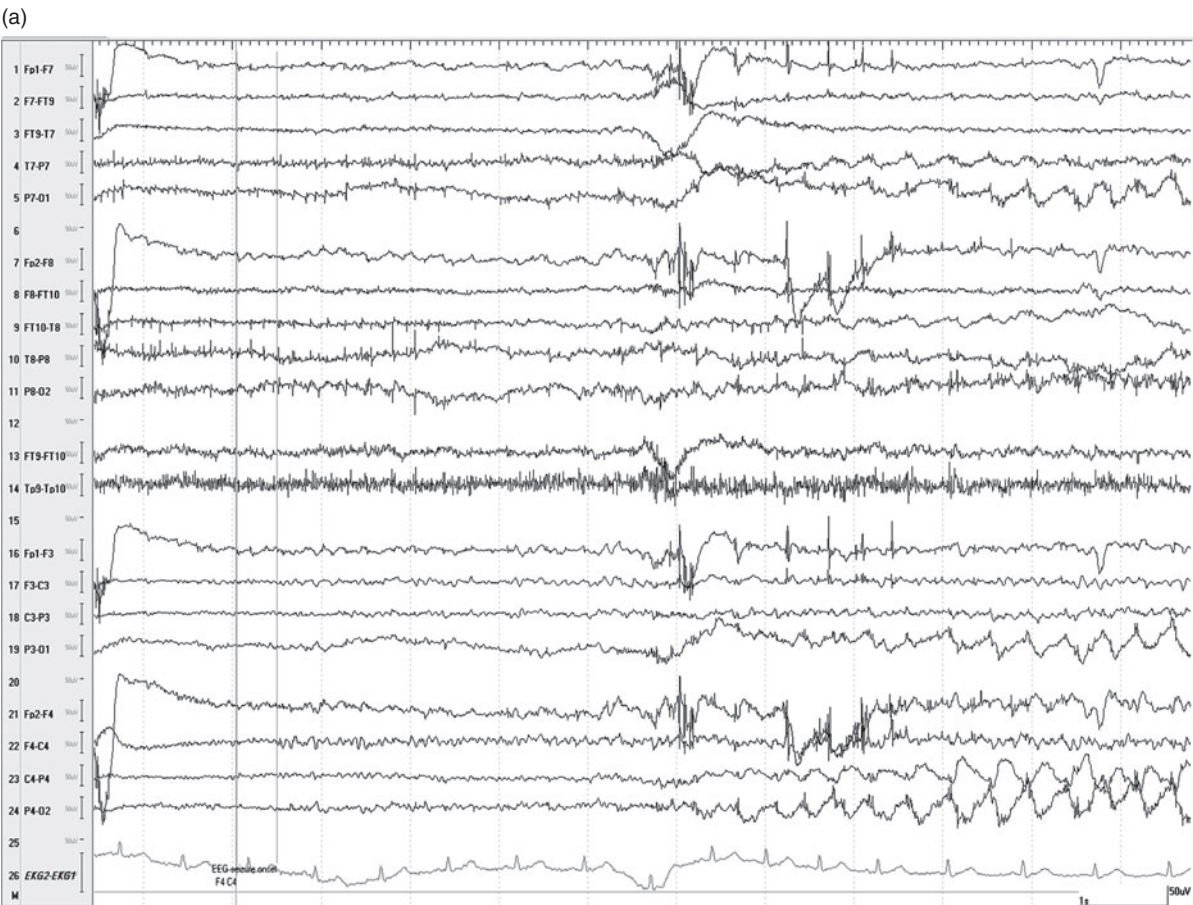


Fig. 1(a). EEG seizure pattern with right frontal seizure onset (max. F4/C4).

occipital lobe, pointing towards an activated cortical saccade region, which caused simultaneous supranuclear inhibition of ipsilateral eye movement or triggered monocular eye movement commands. Both our case and the one reported by Grant are at variance with Hering’s law, which postulated that premotor eye movement commands are always binocular, and that these commands consist of separate components for the control of saccades and accommodation.

Special remarks

Recent data from invasive video–EEG monitoring with subdural grids reported by Thurtell and

colleagues give evidence for a three-dimensional cortical control of gaze mediated by the frontal eye fields. Based on earlier findings from lesion, imaging, and stimulation studies the human frontal eye fields are thought to be located at the caudal end of the middle frontal gyrus known to be in control of contralateral versive eye movements, saccades, and smooth pursuit.

Both described patients developed disconjugated contraversive horizontal eye movements in response to electrical stimulation of the frontal cortex or during focal seizures close to the frontal eye field.

Suggested reading

Grant AC, Jain V, Bose S, *et al.*
‘Epileptic monocular nystagmus’. *Neurology* 2002; **59**: 1438–41.

Kaplan PW, Tusa RJ.
‘Neurophysiologic and clinical correlations of epileptic nystagmus’. *Neurology* 1993; **43**: 2508–14.

Lobel E, Kahane P, Leonardis U, *et al.*
‘Localization of human frontal eye fields: anatomical and functional findings of functional magnetic resonance imaging and intracerebral

Section 1: Diagnosis



Fig. 1(b). Right frontal EEG seizure pattern; please note the occipital artifacts due to symmetrical head nodding.

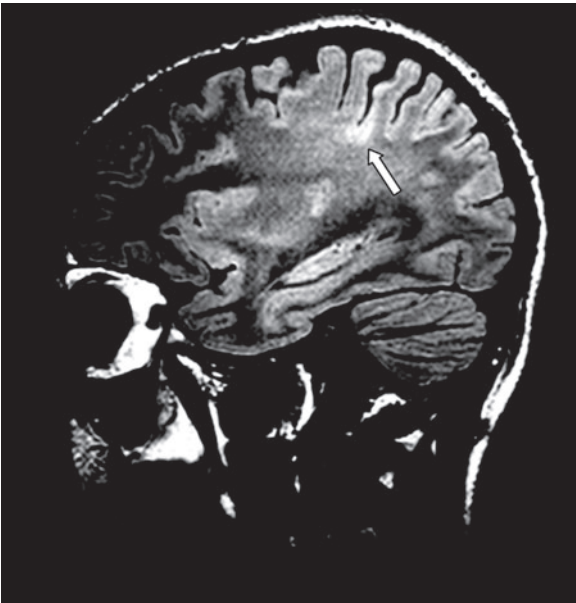


Fig. 2. Sagittal FLAIR image with a right frontal focal cortical dysplasia (arrow).