MRI disclosed serpentiform nerve fascicles surrounded and separated by fibrous and fatty tissue within the expanded nerve sheath as typical features of fibrolipomatous hamartoma.[2 4] Fibrolipomatous hamartoma can clearly be distinguished from lipomas within the nerve sheath, which are characteristic focal masses that dislocate and compress the normal nerve bundles,[6] and from segmental and plexiform neurofibromatosis, in which the neurofibroma has MRI signal characteristics of soft tissue and not of fat. Furthermore, in plexiform neurofibromatosis, the tortuous nerve is studded by small tumours.[6 7] The unique features of fibrolipomatous hamartoma as identified by MRI , allow the identification of this benign nerve tumour preoperatively. This facilitates the decision to decompress affected nerves at the preferential sites of nerve entrapment and helps to avoid diagnostic nerve biopsy or even resection of an ambiguous nerve tumour.

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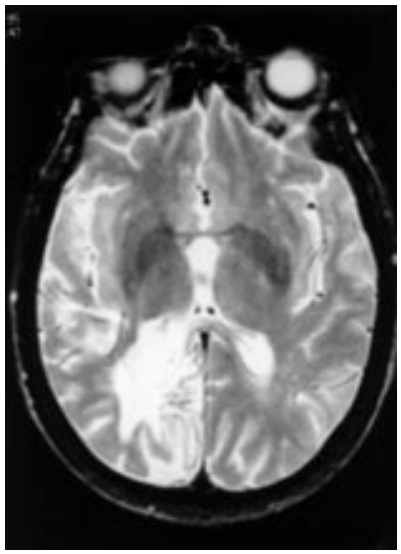
## Ipsilateral mydriasis in focal occipitotemporal seizures

Occipital epilepsy is characterised by seizures which usually begin with oculomotor or elementary visual symptoms and often spread to other cortical and subcortical regions. We think that the present case report is of particular interest because of the nature of partial status comprising positive (clonic jerks of eyes and left face) and negative motor components (left hemiplegia) with dilatation of the right pupil; these clinical features were shown to be related to a lesion in the right occipital lobe by MRI and EEG. To our knowledge, the association of these data has not been previously reported.

A 37 year old man had a history of chronic renal failure due to an idiopathic mesangiocapillary glomerulonephritis since he was 20 years old. He underwent a renal transplantation which was unsuccessful because of an arterial thrombosis. Two years later, a second kidney was transplanted to the patient. Shortly afterwards, while he was immunosuppressed with steroids and azathioprine, he had a *Listeria monocitogenes* meningoencephalitis. Treated promptly with ampicilline, he had an excellent outcome, except for recurrent generalised seizures due to a residual lesion in the right occipital lobe, shown by CT. The patient was treated with oral 100 mg phenytoin thrice daily and required haemodialysis because of the progressive worsening of his renal function owing to chronic organ rejection. One year later, he was admitted to our hospital because of fever of unknown origin. A few days after admission, he developed a focal status epilepticus characterised by stupor, tonic deviation of the head and clonic jerks of the eyes to the left, full dilatation of the right pupil with sluggish reaction to light, clonic movements of the left face, and left hemiplegia with hyperreflexia and extensor plantar response. No previous clonic movements were seen on plegic limbs. Meanwhile, left pupil responses remained normal. Laboratory studies showed evidence of chronic renal failure and the serum concentrations of phenytoin were 5 μg/ml (reference range 8-20 μg/ml). After intravenous administration of 1000 mg diphenylhydantoin, the seizures stopped and simultaneously the pupillary dilatation disappeared. Thereafter, pupillary assessment showed no abnormalities. A left hemiparesis and extensor plantar response were transient postictal findings. A long TR/long TE (2000/80) MRI showed a fairly well defined area of hyperintensity on the right occipital lobe, without any sign of a mass effect (figure). Lumbar puncture disclosed a clear CSF under normal opening pressure, with no pleocytosis and containing normal amounts of glucose and proteins; microbiological studies were negative. Several hours after the status had finished, an EEG disclosed sharp spikes and slow waves over the right temporo-occipital region. Fever responded to empirical antibiotic therapy. The clinical course was uncomplicated and the patient continued on phenytoin, being free from seizures after one year of follow up.



*A long TR/long TE (2000/80) MRI showing a fairly well defined area of hyperintensity on the right occipital lobe.*

Our patient showed a focal status epilepticus with right pupillary dilatation and tonic deviation of the eyes and head to the left; other associated clinical features were clonic movements of the left face and left hemiplegia. All these phenomena disappeared dramatically on phenytoin treatment, thus indicating their epileptic pathogenesis. Although an ictal EEG was not recorded, major although indirect arguments in favour of a right occipitotemporal onset are the interictal EEG spikes as well as the MRI lesion located on that area.

Hemiplegia may be a well known negative ictal phenomenon.[1] Its association with clonic movements at other levels, as seen in our patient, suggests its ictal mechanism. The absence of previous convulsions on plegic areas further supports that idea. The concomitance of clonic and atonic seizures has only been previously described in a few series and is a very uncommon clinical pattern. However, it is difficult to ascertain that such paresis in this case is directly due to neuronal discharges. Postictal (Todd's) paralysis may appear in the context of partial status epilepticus, probably due to local fluid changes and this could be another explanation for the hemiplegia in our patient.

Unilateral mydriasis during fits should arouse the suspiction of brain herniation and proper imaging studies should be performed to rule out such a possibility, and that was the case in our patient. Only eight cases with ictal mydriasis have been reported as far as we know. Five patients showed the pupil abnormality contralateral to the epileptic scalp EEG focus and three had ipsilateral mydriasis.[2]

The exact pathophysiology of pupil changes during seizures remains unclear; it has been stated that miosis would represent an excitatory component[3] whereas dilatation would be interpreted as a negative ictal phenomenon.[1] Descending inhibition of the Edinger-Westphal nucleus would result in pupillary dilatation and impairment of the pupillary light reflex; this could be mediated by leu-enkephalin fibres which may produce pronounced inhibition of this nucleus in experimental studies.[4] Animal experiments performed in macaques showed that electrical stimulation of the anterior occipital lobe produced contralateral ocular deviation with asymmetric dilatation of both pupils that was greater in the homolateral eye[5]; these features were found in our patient, although we did not see changes in the diameter of the left pupil and the reason for this disparity is unknown. This finding was by contrast with the contralateral mydriasis obtained when the frontal eye field was stimulated[5] and in patients with frontal epilepsy. The exact anatomical basis for the changes in pupil diameter during fits remains to be elucidated although the reports and experiments commented on herein, including our case, indicate that an ictal pupil dilatation found in a patient with contralateral epileptic movements suggest occipitotemporal pathology on the same side as the mydriasis; if all these changes are ipsilateral, a frontal lesion should be suspected on the opposite side.

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**Parkinson's disease and tumour in the supplementary motor area: a re-evaluation**

In 1988 Straube and Sigel[1] reported on a 56 year old patient with a bilateral Parkinson´s syndrome, including resting tremor, rigidity, bradykinesia, and a favourable response to levodopa medication, starting at the age of 51 years. This patient was discovered to have a tumour (low grade astrocytoma grade II-III) in the left hemisphere, mainly involving the supplementary motor area. Based on the low chance that both Parkinson's disease and astrocytoma occurred at the same time in this young patient (estimated probability 0.005%–0.1%), it was postulated that the tumour in the supplementary motor area induced the bilateral parkinsonian syndrome, although the levodopa responsiveness was unusual. Importantly, however, the supplementary motor area is considered to represent one of the critical motor areas which are establishing the motor circuit through the basal ganglia.[2 3] We now report that this patient has idiopathic Parkinson´s disease independently of astrocytoma grade II.

The patient is now 65 years of age. She has typical Parkinson's disease, featured by bradykinesia, rigidity, and resting tremor predominantly on the left side. She has responded favourably and continuously to levodopa over the past five years.

Her daily medication at the report of 1988 was 400 mg levodopa (plus decarboxylase inhibitor) and 12.5 mg bromocriptin. Her medication now consists of a combination therapy of 400 mg levodopa (plus decarboxylase inhibitor) and 1 mg pergolide per day. Currently, she has developed biphasic motor fluctuations, and biphasic and peak of dose dyskinesia on both sides (predominantly left) and nocturnal akinesia. Testing with apomorphine (3 mg subcutaneously) disclosed a positive dopaminergic response according to the criteria previously published.[4 5]

Her astrocytoma grade II was treated successfully with iodide-125 seeds in the tumour. She currently presents with a cyst in her supplementary motor area. Over the past two years there was regrowth of the tumour (anaplastic astrocytoma, grade WHO III). She was retreated with a cause trial of *x* rays and cortisone. Under this treatment, her parkinsonian syndrome (shuffling gait, resting tremor, on-off fluctuations, freezing episodes) deteriorated transiently for about 10 weeks. This deterioation seemed to distinctly exceed potential *x* ray/cortisone induced side effects, as for instance manifested by fatigue and equilibrium disturbances. After cessation of the treatment, the parkinsonism gradually improved to the previous stage over a period of six months in combination with increased levodopa/dopamine agonist dosages.

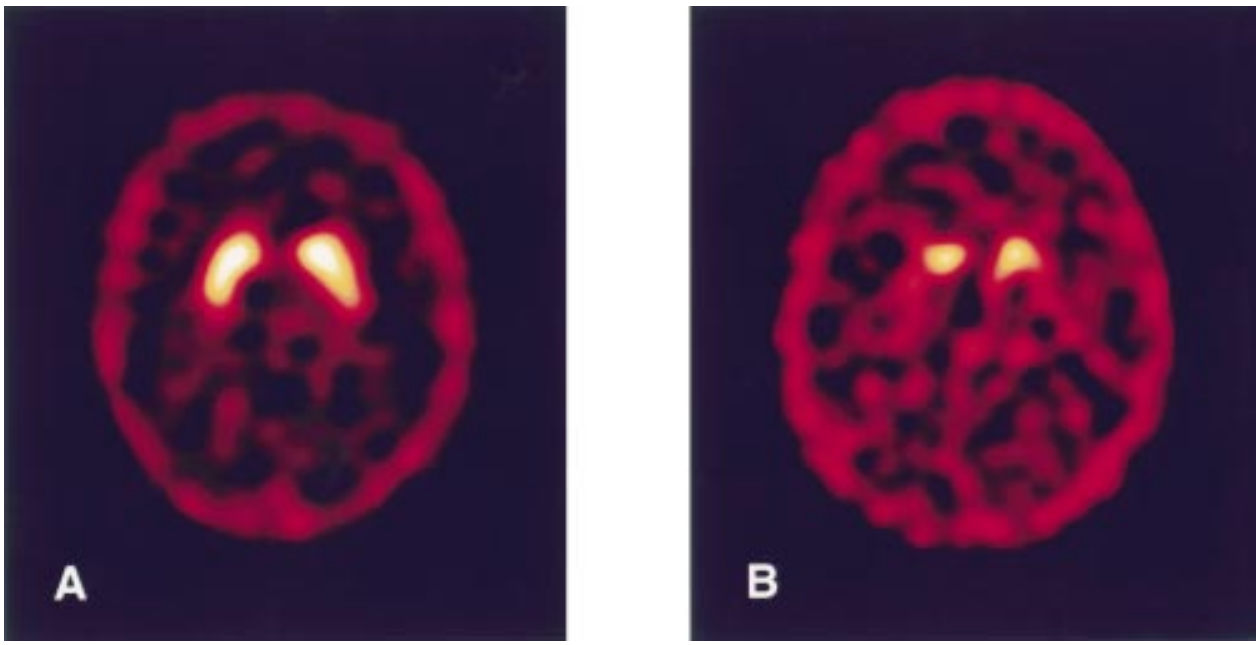
Single photon emission computed tomography (SPECT) investigations performed with [I-123]IPT (N-(3-iodopropen-2-yl)-2β-carbomethoxy-3β-(4-chlorophenyl) tropane, a cocaine analogue with high affinity for the presynaptic dopamine transporter,[6 7] displayed greatly reduced striatal binding of the radioligand. Compared with age matched controls specific [I-123]IPT binding was significantly reduced with more pronounced decrease of tracer accumulation in the putamen than in the caudate. Two SPECT scans were performed at an interval of 12 months. Both studies disclosed findings compatible with idiopathic Parkinson's disease (figure, table).[8] Multiple system atrophy, progressive supranuclear palsy, and other atypical Parkinson's disease syndromes were excluded by clinical and MRI criteria. Additionally, an iodobenzamide (IBZM) SPECT showed normal, striatal D2 receptor binding, compatible with the diagnosis of idiopathic Parkinson's disease.[9]

In conclusion, the proposal of the previous report[1] that a tumour in the supplementary motor area may cause a parkinsonian syndrome is withdrawn. At present we are not

*Spect-IPT values*

| | *March 1995* | *March 1996* | *Age matched controls* |
|---|---|---|---|
| Striatum left | 3.1 | 3.1 | |
| Striatum right | 3.0 | 2.6 | |
| Mean (SD) | 3.0 | 2.9 | 7.3 (1.3) |
| Caudate left | 4.5 | 3.9 | |
| Caudate right | 4.2 | 3.7 | |
| Mean (SD) | 4.4 | 3.8 | 8.6 (1.4) |
| Putamen left | 1.8 | 2.7 | |
| Putamen right | 2.2 | 1.7 | |
| Mean (SD) | 2.0 | 2.2 | 6.5 (1.4) |

SPECT was performed 90 to 120 minutes after injection of 150 MBq IPT. After reconstruction by filtered backprojection transverse slices corrected for attentuation were realigned parallel to the AC-PC line. For semiquantitative evaluation of specific (I-123)IPT binding ratios beween striatum, caudate, putamen, and background regions were calculated. Specific uptake ratios were defined as mean counts per pixel in the respective region of interest minus mean counts in a background region divided by the mean counts in the background region. The uptake ratios obtained in the patient studies and in age matched controls are listed in the table. The increased IPT binding in the left putamen in the 1996 study is most probably due to an artefact caused by a spot of high background activity accidentally located around the putamenal region of interest in this investigation.




*Compared with a representative control (A) the SPECT study of the patient showed profoundly reduced striatal IPT uptake (B). The decrease of specific IPT uptake was more pronounced in the putamen than in the caudate.*