deteriorating neurological state accompanying the extensive venous obstruction and cortical vein thrombosis. It is likely that the risk of haemorrhagic complications in this condition was reduced by using local rather than systemic thrombolysis. It is interesting to note that streptokinase seemed to be more effective than urokinase at the dosages employed. Moreover, it may be that other cases can be treated successfully by clearing the straight and transverse sinuses without directing treatment to the sagittal sinus.

The association of ulcerative colitis and dural sinus thrombosis has been previously documented, but typically during a flare up of bowel disease in the context of severe illness. In our patient the dural sinus thrombosis occurred while the ulcerative colitis was relatively quiescent, and she continued to develop areas of thrombophlebitis despite anticoagulation suggesting that thrombotic complications need not be confined to aggressive bowel disease. Our experience suggests that in fulminant dural venous sinus thrombosis local endovascular infusion of streptokinase as described should be considered as an early therapeutic option.

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Cervical dystonia as an isolated sign of a basal ganglia tumour

We report a patient who had cervical dystonia for two years before a tumour involving the basal ganglia and surrounding white matter was detected. This case indicates that symptomatic focal dystonia may be the only early sign of a basal ganglia tumour. This male patient with an unremarkable medical history felt pain in his right neck and shoulder at the age of 40. When referred to a neurologist some months later, a laterocollis with inclination of the head to the right side was diagnosed. Trihexiphenidyl (50 mg daily) was effective in reducing the patient’s complaints but had to be discontinued because of dizziness. The improvement of laterocollis outlasted the period of medication. A first injection of botulinum toxin was given two years after the onset of symptoms, which again was effective in reducing the feeling of muscle tension. At that time the neurological state was otherwise normal.

One month later, the patient developed a headache and numbness in the right temporoparietal region. There was hypaesthesia in the first right branch of the trigeminal nerve. Brain MRI showed an extended tumour involving the right basal ganglia and frontoparietal white matter with a large surrounding oedema. A stereotactic brain biopsy gave evidence of an anaplastic glioma consisting mainly of glial fibrillary acidic protein negative tumour cells. As the tumour was too extensive to be surgically removed, radiotherapy and steroid medication were started. In the course of the next nine months, a left hemiparesis developed, whereas the laterocollis almost disappeared. The patient died from a pulmonary embolism less than three years after the first appearance of cervical dystonia. A mixed grade III glioma was confirmed at necropsy. Aside from this case describing a glioma involving the basal ganglia associated with laterocollis, cervical dystonia has been reported as a consequence of arteriovenous malformations located at the head of the caudate nucleus in two cases, of infratentorial tumours, and of traumatic lesions.

Cases of symptomatic contralateral hemidystonia have been described after basal ganglia infarction and after trauma. Although there was no familial history of dystonia in our patient, a single case cannot rule out the possibility that his dystonia was idiopathic and unrelated to the tumour. If, however, the tumour is not merely coincidental with laterocollis, the question arises as to whether brain imaging should be performed also in patients presenting with cervical dystonia to exclude a structural pathology.

Laterocollis in this case improved in parallel with tumour growth and almost disappeared when hemipaesis developed. After infarcts and head trauma, dystonia may develop when hemipaesis improves. This clinical finding as well as pathological investigations have led to the hypothesis that dystonia may arise when striatal lesions leave the pyramidal tract relatively intact. In the case reported here, tumour growth may have presented as cervical dystonia only when basal ganglia began to be involved. With tumour extension into the internal capsule and white matter surrounding the basal ganglia, dystonia improved and paresis developed. The reversed

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sequence of symptoms in this case may give additional support to the notion that the degree of dystonia and paresis may be inversely related.

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Debrisoquine hydroxylase gene polymorphism in Parkinson's disease and amyotrophic lateral sclerosis

The aetiopathogenesis of Parkinson's disease and amyotrophic lateral sclerosis is considered to be multifactoral, including genetic and environmental factors. Toxic neuronal damage by free radicals is thought to be an important factor in the pathogene-

sis of these neurodegenerative disorders. The cytochrome P-450 monooxygenase enzymes detoxify toxic environmental com-

pounds and we have previously reported that there is a highly significant excess of cytochrome P-450 debrisoquine hydroxy-

lase (CYP2D6) gene mutation in patients with Parkinson's disease compared with controls.1 The mutation leads to loss of the normal enzyme and the phenotype known as the poor metaboliser status and confers susceptibility to Parkinson's disease. To see whether the association between the poor metaboliser genotype and Parkinson's disease is selective for this type of neurodegen-

eration, we have now compared the frequency of poor metaboliser mutations in patients with Parkinson's disease, patients with amyotrophic lateral sclerosis, and controls. We have also studied the frequency of polymorphism of another gene, N-acetyl transferase, which is responsible for metabolising dapsone, a diphenylsulphone via CYP3A4, an isoenzyme of the cytochrome P-450 enzymes.1

Blood samples were obtained from 272 cases with idiopathic Parkinson's disease (diagnosed by a neurologist and fulfilling the UPDRS criteria) and 96 cases of clini-

cally definite or probable cases of amyotrophic lateral sclerosis.1 Samples from 720 healthy controls were also studied. Identification of mutant CYP2D6 were carried out on genomic DNA amplified by the polymerase chain reaction (PCR) followed by restriction fragment analysis as described previously.1 N-Acetyl transferase polymorphism was studied using PCR for identification of slow acetylators in the patients with Parkinson's disease, patients with amyotrophic lateral sclerosis, and 96 controls.

The table summarises the results. Of 272 cases of Parkinson's disease, 11-8% were poor metabolisers by CYP2D6 genotype (mutant allele frequency = 0.259) whereas only 5-1% and 5% of patients with amyo-

trophic lateral sclerosis and controls (each p < 0.05) respectively were poor metabolisers.

Changes in N-acetyl transferase polymorphism were, however, not significant between patients with amyotrophic lateral sclerosis and those with Parkinson's disease.

We conclude that CYP2D6 polymorphism leading to poor metaboliser status is significantly more common in Parkinson's disease compared with amyotrophic lateral sclerosis. This finding further strengthens the initial observation that CYP2D6 polymorphism confers increased susceptibility to Parkinson's disease. Further studies on the functions of CYP2D6 are required to identify those at risk for developing Parkinson's disease as well as the various factors leading to development of Parkinson's disease.

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Debrisoquine hydroxylase (CYP2D6) and N-acetyl transferase (NAT) polymorphism in Parkinson's disease (PD), amyotrophic lateral sclerosis (ALS), and controls (Crtl)

<table>
<thead>
<tr>
<th>CYP2D6</th>
<th>NAT</th>
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<tbody>
<tr>
<td>No</td>
<td>PMs</td>
</tr>
<tr>
<td>PD</td>
<td>272</td>
</tr>
<tr>
<td>ALS</td>
<td>96</td>
</tr>
<tr>
<td>Controls</td>
<td>720</td>
</tr>
</tbody>
</table>

PMs = poor metabolisers; MAF = mutant allele frequency; Slow Ac = slow acetylators; ALS = amyotrophic lateral sclerosis.

Glioependymal cyst of the cerebello-

pontine angle

Intracranial cysts may be intracerebral or extracerebral. Arachnoid and subarachnoid cysts are the most common extracerebral types. These can be of developmental, trauma-

tic, or inflammatory origin. Extracerebral cysts lined by epithelial ependymal cells are reported under a variety of names—ependymal, glioependymal, neuroepithelial, choroidal epithelial, and ependymal cysts—and at a variety of sites with the location at the cerebellopontine angle being excep-
tional.1 We report the case of a huge cyst located near the right bulbopontine junction. A 21 year old woman complained of a nasal tone to her voice—rhinolalia—for one year. There was no head trauma, infection, or other CNS disorder. Neurological exami-

nation showed a palsy of the right ninth cran-

nial nerve. Routine laboratory profiles were normal. Brain MRI found a cystic lesion in the posterior cranial fossa that filled the right cerebellopontine angle cistern and compressed the brainstem (figure, left). The fourth ventricle, medulla oblongata, and pons were shifted to the left. There was no

MRI showing a glioependymal cyst of the right cerebellopontine angle. Left, preoperative view; showing a severe brainstem compression; right, one year after operation, with disappearance of the lesion.

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