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 multifactorial, including genetic and environmental factors. Toxic neuronal damage by free radicals is thought to be an important factor in the pathogenesis of these neurodegenerative disorders. The cytochrome *P*-450 monooxygenase enzymes detoxify toxic environmental compounds and we have previously reported that there is a highly significant excess of cytochrome *P*-450 debrisoquine hydroxylase (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 neurodegeneration, 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.[2,3]

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 clinically definite or probable cases of amyotrophic lateral sclerosis.[4] 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 amyotrophic 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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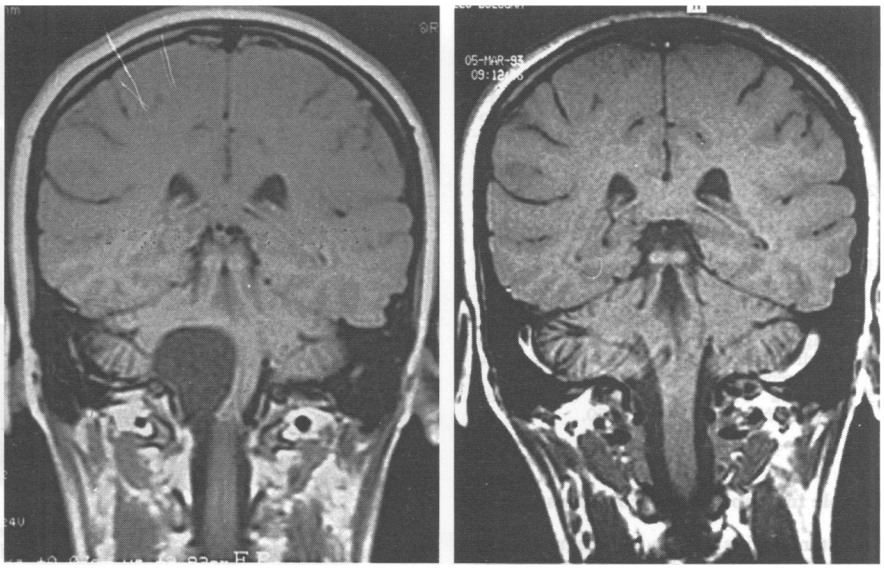
Debrisoquine hydroxylase (CYP2D6) and N-acetyl transferase (NAT) polymorphism in Parkinson's disease (PD), amyotrophic lateral sclerosis (ALS), and controls (Ctrls)

	CYP2D6			*NAT*		
	No	*PMs*	*MAF*	*No*	*Slow Ac*	*MAF*
PD	272	11·8%	0·259	272	50%	0·7
ALS	96	5·1%	0·250	96	60%	0·75
Controls	720	5·0%	0·206	96	63%	0·8

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

Glioependymal cyst of the cerebellopontine angle

Intracranial cysts may be intracerebral or extracerebral. Arachnoid and subarachnoid cysts are the most common extracerebral types. These can be of developmental, traumatic, or inflammatory origin. Extracerebral cysts lined by epithelial ependymal cells are reported under a variety of names—ependymal, glioependymal, neuroepithelial, choroidal epithelial, and epithelial cysts—and at a variety of sites with the location at the cerebellopontine angle being exceptional.[4] 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 examination showed a palsy of the right ninth cranial 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.

evidence of communication between cyst and fourth ventricle. The supratentorial ventricular system seemed to be normal. Computerised tomography and digital vertebral angiography were non-contributory.

A right retromastoid craniotomy was performed with the patient in the supine position and head turned to the left. After incision of the dura and cerebellar retraction, a large cystic mass with a thick wall appeared. The rostral pole reached the seventh and eighth cranial nerves. The caudal pole extended to the first cervical root. The ninth, 10th, and 11th cranial nerves were stretched downward. The cyst did not communicate with either the fourth ventricle or subarachnoid spaces. Incision of its wall immediately produced collapse of the cyst and relaxation of the nerves; the underlying brainstem seemed severely distorted. The cyst was opened into the subarachnoid space. The postoperative course was uneventful and CT within a few hours of operation showed significant reduction in the size of the cavity.

One year later, there was a significant improvement in the glossopharyngeal palsy and the disappearance of rhinolalia, and MRI showed resolution of the cyst (figure, right).

Histological examination showed the cyst wall to be composed of two distinct layers; the inner layer of neuroglial tissue and the outer of fibrous tissue. The luminal surface was lined by cubic cell epithelium of ependymal origin.

According to Morimura,[5] our patient's cyst should be described as a glioependymal cyst thereby emphasising its developmental and heterotopic origin and its neuroepithelial nature. Three previous reports have described glioependymal cysts of the cerebellopontine angle cistern, two of which were symptomatic and were treated successfully by surgical marsupialisation or fenestration, keeping the wall in situ, as we have done for our patient, with good long term relief of symptoms.

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Double intraspinal enterogenous cysts

An enterogenous cyst in an intramedullary or intradural extramedullary location in the ventral cervicodorsal spinal canal may be a cause of spinal cord compression. Embryologically, the cyst derives from endodermal tissue displaced dorsally into the spinal canal ventral to the neural plaque through the interposed mesodermal layer that forms the vertebral bodies.[1] Among 63 histologically verified cases of enterogenous cyst in the English literature, the case reported here is the only one that has been found with double cysts.

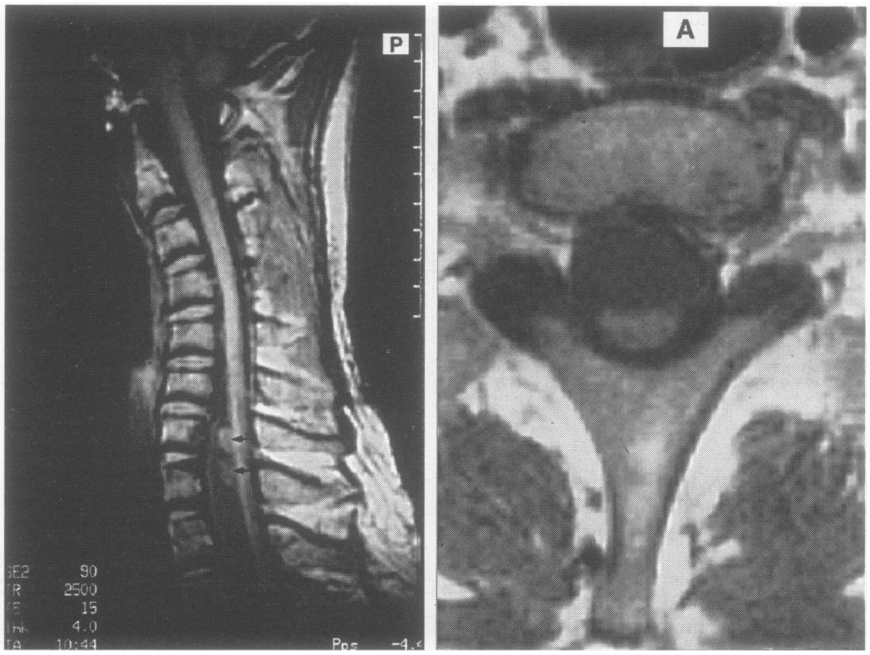

Figure 1 Composite photographs of two MR images in axial view (right) and sagittal view in T1 weighted sequence (left). The spinal cord has been displaced posteriorly by two separate enterogenous cysts (arrows).

A 30 year old man presented with a history of weakness and atrophy of intrinsic muscles confined to his right hand for 15 years. He had neither history suggestive of poliomyelitis nor relevant sensory dysfunction before developing the present illness. The symptoms showed no progression until one year before admission when atrophy of his left hand and paraesthesia over his chest and right leg insidiously developed.

Needle electromyography showed chronic denervation over muscles innervated by C7-T1 roots with absent volitional activity over C8-T1 roots. The compound muscle action potential was absent over the right abductor pollicis brevis and abductor digiti minimi whereas sensory nerve coordination in his right arm was normal.

Lateral radiographs of the cervical spine showed widening of the anteroposterior diameter of the cervicothoracic junction. Magnetic resonance imaging of the cervical

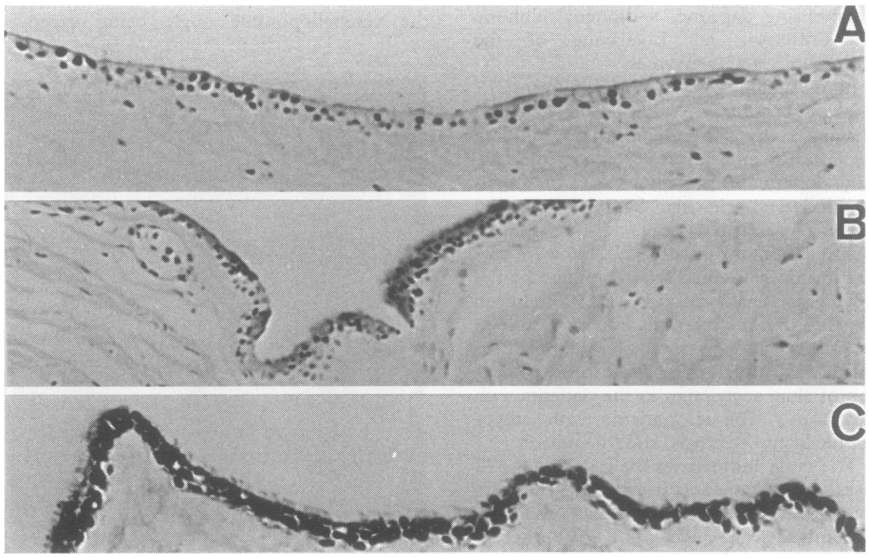

Figure 2 Composite photographs of sections from the cyst. (A) Histological appearance showing that the cyst wall is composed of connective tissue lined by a single layer of ciliated and non-ciliated columnar epithelium (haematoxylin and eosin, originally × 260). (B) Intracytoplasmic mucin can be identified in the epithelial cyst lining (diastase pretreated periodic acid Schiff reaction, originally × 165). (C) The cytoplasm responds positively to carcinoembryonic antigen (CEA) staining (anti-CEA, originally × 220).