SHORT REPORT

Parkinsonism associated with obstructive hydrocephalus due to idiopathic aqueductal stenosis

M Zeidler, P J Dorman, I T Ferguson, D E Bateman

Abstract

Two cases of parkinsonism after recurrent obstructive hydrocephalus due to idiopathic aqueductal stenosis are reported. In both patients an extrapyramidal syndrome was noted in the absence of contemporaneous evidence of hydrocephalus or shunt failure. One of the patients underwent a shunt operation, but showed no clinical improvement. However, both patients improved after the administration of dopaminergic therapy. The seven previously reported cases of this syndrome were reviewed and it is concluded that the prognosis of the parkinsonism is good, usually with total, or near total, resolution. It is recommended that if a patient with idiopathic aqueduct stenosis develops hydrocephalus or evidence of shunt malfunction in association with acute parkinsonism their shunt should be replaced. If there is no evidence of hydrocephalus or shunt malfunction they should initially be treated with domaminergic medication.

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Department of Neurology, Royal United Hospital, Bath, UK
M Zeidler
D E Bateman

Department of Neurology, Southmead Hospital, Bristol, UK
P J Dorman
I T Ferguson

Correspondence to:
Dr David Bateman, Department of Neurology, Royal United Hospital, Bath, UK.

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Parkinsonism is a rare complication of obstructive hydrocephalus.[1–8] There are only seven patients reported with an extrapyramidal syndrome secondary to non-neoplastic aqueductal stenosis.[1–4 8 9] Lack of awareness of this association may cause diagnostic difficulty and unnecessary shunt revision. We report two further cases to emphasise the association of parkinsonism and recurrent obstructive hydrocephalus.

Case histories

CASE 1

A 57 year old racehorse trainer presented with a one year history of unsteadiness of gait associated with general lethargy. He had a lifelong history of asthma, requiring intermittent courses of oral steroids in addition to regular inhaled steroid and bronchodilator therapy. He admitted heavy and long standing alcohol consumption. Neurological examination showed a generalised hyperreflexia and equivocal planters. Sensory testing disclosed reduced appreciation of vibration sense at both ankles.

Brain CT showed dilatation of the lateral and third ventricles. Subsequent MRI confirmed this, and showed a normal sized fourth ventricle with aqueductal stenosis. Routine blood tests were normal.

Gait and cognitive function deteriorated over the next three months, necessitating insertion of a ventriculoperitoneal shunt. This operation was successful allowing him to return to his previous occupation. Sixteen months later he gradually deteriorated, developing blurred vision, problems with upgaze, and loss of drive. Over the next eight months he continued to deteriorate with increasing unsteadiness of gait. Repeat neurological assessment disclosed complete absence of upgaze to command, although vertical movements on the doll's head manoeuvre were preserved. Despite the clinical deterioration MRI showed that the ventricles were of normal size. On neurosurgical assessment the shunt reservoir failed to empty and refill satisfactorily. This was presumed to account for his deterioration. Subsequent shunt replacement was initially followed by clinical improvement. Acute shunt failure (confirmed radiologically) recurred and an emergency procedure was required to resite it. On this occasion the recovery was marred by an exacerbation of his longstanding asthma. His subsequent neurological recovery was slow with parkinsonian features noted for the first time. Brain CT at this point showed normal ventricular size excluding recurrent hydrocephalus. He complained of double vision, dribbling of saliva, shaking, and difficulties in turning over in bed. His voice had become soft, monotonous, and slow. A Torkildsen operation was performed as further shunt dysfunction was thought to possibly underlie this deterioration. Again his postoperative course was complicated by both seizures and respiratory distress requiring ventilation. On extubation he was found to have a severe extrapyramidal disturbance with: an immobile facies characterised by an absence of spontaneous movement, soft voice, bradykinesia, rigidity, and

Cases of extrapyramidal syndrome associated with obstructive hydrocephalus due to idiopathic aqueductal stenosis

Case	Age at onset of parkinsonism	Time to onset of parkinsonism after hydrocephalus first diagnosed	Previous shunts before onset of parkinsonism (n)	Parinaud's syndrome	Hydrocephalus present at onset of parkinsonism?	Response to shunting	Medication prescribed	Drug withdrawal possible
Case 1	57	2 y	3	Yes	No. But clinical assessment suggested shunt blockage	No immediate improvement	Bromocriptine 3mg bd Sinemet 250 qds	No
Case 2	22	14 months	2	Yes	No	Not shunted	Madopar 125 tds increased to 250 tds	Successfully after 2 y
Case 3[1]	17	1 y	Many	Yes	Yes : mild	No immediate improvement	Sinemet 125 tds	Unknown
Case 4[2]	21	9 months	3	Yes	Yes	Initially slow improvement	Benztropine 2mg tds Prolopa 100/25 bd	Unknown
Case 5[3]	14	At presentation	Nil	Yes	Yes	Initially responded to shunting alone	Sinemet 125 tds	Successful 6 months after last shunt revision
Case 6[4]	7	1 y	4	No	Yes	Yes in combination with levodopa	Increasing dosage up to Sinemet 110 qds	Successful after a few months
Case 7[8]	16	1 y	2	Yes	No. But nuclear shunt scan showed blockage	Slow improvement	Sinemet 25/100 ½ tab tds	No
Case 8[8]	26	17 y	1	Yes	No. Functional status of shunt unknown	None	Sinemet and intravenous lisuride - no response	—
Case 9[9]	28	24 y	1	No	Yes	Parkinsonism promptly disappeared	No antiparkinsonian drugs given	—

bd=twice daily; tds=three times daily; qds=four times daily.

cogwheeling affecting all four limbs and trunk, but a tremor was not noted. The previously noted abnormality of vertical gaze persisted and bilateral internuclear opthalmoplegia was present.

In view of the above extrapyramidal signs he was started empirically on bromocriptine and Sinemet Plus. The dosages were gradually increased to 3 mg twice daily and two tablets four times a day respectively. On this treatment he made a gradual recovery, eventually becoming independently mobile without aid and self caring. At outpatient review after discharge his improvement continued with the extrapyramidal signs becoming less prominent, allowing a reduction in the dosages of the dopaminergic agents prescribed. He has since died from an acute deterioration of his longstanding obstructive airways disease.

CASE 2

A 21 year old previously fit and well woman presented with a nine day history of progressive bifrontal headache, vomiting, and deteriorating conscious level. On examination she had no eye opening to pain or verbal responses and her best motor response was flexion to pain. Bilateral papilloedema was present. Brain CT showed severe and dramatic supratentorial hydrocephalus with a normal fourth ventricle and posterior fossa, consistent with aqueduct stenosis. Skull radiography showed changes due to chronic raised intracranial pressure. A ventriculoperitoneal shunt was inserted and she made a rapid recovery. Repeat brain CT showed resolution of the severe hydrocephalus.

Ten months later she was admitted after a two week history of headache, vomiting, blurred vision, and progressive drowsiness. Neurological examination disclosed dysarthria and failure of upgaze. Brain CT showed recurrent hydrocephalus and her shunt was successfully revised.

Three months later she developed increasing headaches and sleepiness. She was readmitted and MRI excluded hydrocephalus. She improved and was discharged, but three weeks later she was again admitted with increasing blurred vision, excessive drowsiness, immobility, and increased dependency. On examination she was drowsy but awoke to verbal stimuli. She responded to simple commands but was mute. There was pronounced bradykinesia, rigidity, an impassive face, and postural tremor of the left arm. She had a brisk jaw jerk, pout reflex, and grasp reflexes. Tendon reflexes were symmetrically very brisk with bilateral extensor planter responses. Power was normal. There was bilateral lid retraction, reduced convergence, limited upgaze, hypometric horizontal saccades, and convergence retraction nystagmus. Pupillary light reactions were normal. Fundoscopy showed clear disc margins but absent venous pulsation.

Brain CT showed generalised atrophy, most prominent in the region of the basal ganglia, temporal lobes, and brain stem. The ventricles were prominent with a shunt in situ. These appearances were confirmed by MRI which additionally showed that the aqueduct was thin but not obstructed. Routine tests excluded both metabolic and infective causes of the extrapyramidal syndrome.

She was started on 125 mg Madopar thrice daily and within two days there was evidence of improvement in the rigidity. The dose of Madopar was slowly increased to 250 mg three times daily and over the next fortnight she gained spontaneous speech and greater mobility. On discharge she was still clearly parkinsonian but by now was self caring and fully mobile.

Over the subsequent years she has required six shunt revisions after episodes of blockage, but without relapse of her extrapyramidal syndrome. This has completely resolved, allowing a successful withdrawal of dopaminergic medication without its recurrence after nine months of follow up. The pretectal signs have diminished but she still has some limited upgaze.

Discussion

Seven other cases of an extrapyramidal syndrome associated with obstructive hydrocephalus due to idiopathic aqueductal stenosis have been described.[1–4 8 9] The table summarises these with the current cases. The clinical picture was remarkably uniform: in five of the seven previous cases the extrapyramidal syndrome was associated with, and often preceded by Parinaud's syndrome and in all but one case was symmetric. The parkinsonism usually occurred many months or years after the initial presentation of hydrocephalus and often followed repeated episodes of shunt failure. Hydrocephalus was clearly present at onset of the parkinsonian syndrome in five of the seven cases; another patient had radiological evidence of shunt obstruction and presumed raised intracranial pressure but no hydrocephalus. CSF shunting was performed in all cases and dopaminergic therapy was given in all but case 9 (shunt revision was effective alone). It is difficult to ascertain the individual therapeutic benefit of these two interventions in combination. However most patients improved after the administration of dopamine agonists, usually after a poor initial response to shunting, although one patient responded to shunting alone. Those cases with clear hydrocephalus at onset responded best to shunting. The prognosis for the syndrome was good in all patients with only minimal or no parkinsonian features at follow up.

In our first patient parkinsonism occurred without radiological hydrocephalus but with possible clinical evidence of shunt malfunction and there was initial clinical improvement after shunt revision. However, he then subsequently became severely parkinsonian without hydrocephalus or evidence of shunt failure. His shunt was resited without benefit and he was prescribed dopaminergic therapy with subsequent slow improvement. In our second case the parkinsonism developed after symptoms suggestive of episodic shunt malfunction but without evidence of hydrocephalus. Her shunt was therefore not replaced and she responded rapidly to levodopa.

These two patients, and those previously described, suggest that there may be two contributory causes to the parkinsonian syndrome. In patient 1 the parkinsonism initially responded to shunting and among the previous cases those with radiological hydrocephalus responded best to shunting suggesting a component of direct immediately reversible compression of the basal ganglia pathways. In patient 2 however, and subsequently in patient 1, radiological hydrocephalus was absent, although there was clinical evidence of shunt malfunction. Resiting of the shunt was ineffective in patient 1 on this occasion, but he subsequently responded to levodopa as did patient 2 implying that chronic intermittent mild hydrocephalus may cause temporary levodopa deficiency reversible over a period of weeks or months.

Recurrent basal ganglia and midbrain compression due to hydrocephalus can therefore cause an extrapyramidal and pretectal syndrome either by direct pressure or more chronically by impairment of blood flow to or neuronal transport in the striatum resulting in dopamine deficiency which is gradually reversible with time.[1–3 5 8] Single photon emission computed tomography of the brain in this syndrome has shown decreased cerebral blood flow in the regions of the caudate and putamen. An improvement in flow was shown after ventriculo-peritoneal shunting.[1] The therapeutic effect of dopamine may not only be due to the replacement of dopamine deficiency secondary to damage to the nigrostrial pathways. It has been postulated that dopamine has a vasodilatory effect on the striatal vessels, increasing local blood flow and hence function.[10] Case 8 failed to respond to levodopa and had normal ^{18}F-dopa PET implying that the syndrome may also occur due to impairment of more distal basal ganglia connections.[8]

This rarely described clinical association of an extrapyramidal disorder with obstructive hydrocephalus may be underrecognised as the clinical picture is often dominated by impairment of consciousness. We recommend that hydrocephalus should be considered in the differential diagnosis of parkinsonism and particularly in patients with suspected progressive supranuclear palsy. If patients with idiopathic aqueduct stenosis develop hydrocephalus or evidence of shunt malfunction in association with acute parkinsonism their shunt should be replaced. If there is no evidence of hydrocephalus or shunt malfunction they should initially be treated with domaminergic medication. These recommendations should help prevent misdiagnosis, avoid unnecessary shunt revision, and obviate long standing dependency on dopamine agonist drugs. Aqueductal stenosis should be added to the ever increasing list of causes of secondary parkinsonism. Importantly, by contrast with the other causes of secondary parkinsonism, the prognosis for the extrapyramidal syndrome is good.

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