A case of normal pressure hydrocephalus presenting as levodopa responsive Parkinsonism

Sir: It is well recognised that normal pressure hydrocephalus may rarely present with pure Parkinsonian signs and gait disturbance without other features of the triad, namely intellectual impairment and urinary incontinence.1-3 I report a case which presented with Parkinsonian findings only for 2 years and which initially responded to levodopa. This case highlights a pitfall of diagnosing idiopathic Parkinsonism on clinical grounds, when there is a positive response to levodopa, which is usually taken to indicate idiopathic disease.

A 64 year old woman presented with a 2 year history of difficulty in walking, slowness in her actions and stiff limbs. She had difficulty getting up from a chair and shuffled when walking. There was no other previous medical history of note. Examination revealed facial impassivity and a resting tremor in the left hand. There was a cog wheel rigidity in all four limbs, left greater than right. There was no sign of pyramidal involvement. Plantar responses were flexor and there was no intellectual or urinary disturbance. Her gait was shuffling with no arm swing.

The diagnosis of Parkinsonism, probably idiopathic, was made and treatment with Sinemet Plus (levodopa and carbidopa) was commenced. On three tablets daily her gait had substantially improved but dyskinesia had not occurred.

She was admitted 1 year later with further deterioration in function. She had great difficulty buttoning clothes and her walking was slow with occasional falls. Signs remained those of akinetic rigid Parkinsonism without intellectual impairment. A CT scan (figure) showed marked hydrocephalus. Again she showed marked improvement (PD rating scores improved by 50%) with increasing dose of Sinemet to eight tablets daily and so conservative management was pursued. Three months later, she again deteriorated swiftly so that she could not walk unaided. At this stage the other features of the hydrocephalic triad supervened with intellectual problems (slowness of thought and poor memory) and occasional urinary incontinence. A ventricular atrial shunt was inserted and she quickly improved so that all anti-Parkinson drugs could be withdrawn. Two years later she remains well, fully functional without any sign of Parkinsonism or intellectual impairment. She walks normally, her only abnormality being slight postural instability when pushed briskly at the shoulders and a mild titubation of the head (a new finding). This woman had signs and symptoms of pure Parkinsonism entirely attributable to idiopathic normal pressure hydrocephalus. Ventricular atrial shunting completely relieved all signs of Parkinsonism. This is an unusual case with a prolonged presentation of purely Parkinsonian findings. Similar cases have been reported,1-3 but none with a positive response to levodopa. This is usually taken to indicate that the disease is idiopathic but this case highlights the pitfall of relying on the levodopa test to confirm this. It is possible that the levodopa response noted was largely a placebo one along with the beneficial effect of a hospital admission. The absence of dyskinesia with a moderate dose of levodopa (250 × 4) should also raise suspicions as to the origins of Parkinsonism, although Lang et al4 has reported fluctuatory dyskinesia secondary to levodopa occurring in a child with Parkinsonism due to hydrocephalus. Other features of the Parkinsonism usually alert one to the possibility of underlying normal pressure hydrocephalus: symptoms of disease, absence of tremor, greater involvement of legs over arms and above all, postural instability. Certainly if Parkinsonism fails to respond to levodopa, or shows secondary failure as in this case, it may be worthwhile considering normal pressure hydrocephalus, as surgical treatment can prove highly effective.

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References

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