Progressive supranuclear palsy: the ugly cousin of Parkinson's disease

MEDDUMA B. KAPPAGODA MB BS, DO, FRCS(Ed), FRACS, FRACO, FRCOphth

The clinical features of progressive supranuclear palsy can be subtle, particularly in the early stages, and provide opportunities for misdiagnosis.

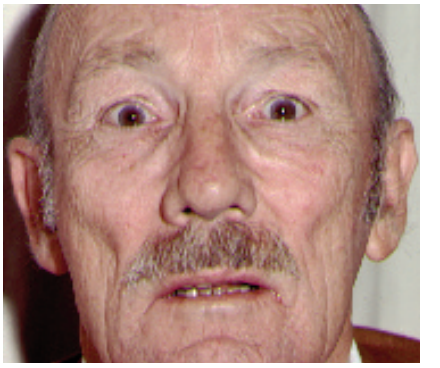

Figure 1. The 'startled' facial expression of a patient with progressive supranuclear palsy.

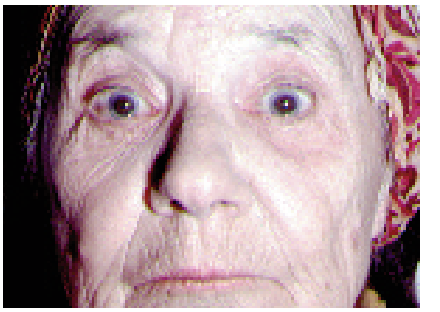

Figure 2. The straight ahead stare.

Progressive supranuclear palsy (or Steele–Richardson–Olszewski syndrome) is a rare neurological disorder. It is sometimes misdiagnosed as Parkinson's disease: about 6% of patients initially suspected of Parkinson's disease may actually have progressive supranuclear palsy.

Clinical features

The clinical features of progressive supranuclear palsy are similar to those seen in degenerative diseases of the central nervous system. There is a superficial resemblance to Parkinson's disease, from which it should be differentiated, but it is an important cause of parkinsonism.

In addition to the common symptoms of stiffness, slow movement and clumsiness, the following features are seen in both progressive supranuclear palsy and Parkinson's disease:

- walking difficulty, postural imbalance and instability
- dementia
- emotional instability
- an immobile face with infrequent blinking
- pseudobulbar palsy, with difficulties in speech and swallowing.

Although progressive supranuclear palsy has many features of Parkinson's disease, significant differences do exist (see Table).

One of the earliest symptoms in progressive supranuclear palsy is loss of balance while walking. Unexplained falls are often described by the patient as being caused by attacks of dizziness, which can raise suspicions of inner ear disease or vertebrobasilar insufficiency and lead to investigations directed at inner ear or vascular pathology.

A change in personality (an early feature) and difficulty with speech are often explained away as signs of 'senility'. Compared with Parkinsons disease, problems with vision, speech and swallowing are more common and severe, and patients may show certain facial expressions (Figure 1). Poor eye contact during conversation may give the impression of tunnel vision (Figure 2).

Mild mental changes in progressive supranuclear palsy can be misinterpreted as manifestations of depression; however, the mental change is one of slowed thought processing. On the other hand, in Alzheimer's disease, memory functions are greatly impaired and there are additional difficulties with language function and visuospatial orientation.

Patients may present with visual difficulties that relate to visual tasks or activities (see the box on page 132). Damage to neural cells in the brainstem cause an early inability to co-ordinate eye movements and aim the eyes accurately in the vertical plane. The effect of progressive supranuclear palsy on eye movement is shown in Figures 3a to d. Tasks involving

Key points

- Progressive supranuclear palsy is a clinical diagnosis. The disease is rare and delays in diagnosis are common. It is easily misdiagnosed as Parkinson's disease.
- Visual and postural problems occur early. Vertical gaze is impaired; downgaze is affected first.
- The disease is progressive and there is no effective cure.
- Mental and personality changes in progressive supranuclear palsy are mild, yet the patient can be demanding. Management is supportive and the burden on the carer should be recognised.

Dr Kappagoda is former Head of the Department of Ophthalmology, Concord Hospital, Sydney, NSW.

The man who couldn't play his cards straight

A 50-year-old man complained about his reading glasses that had been prescribed recently. He had recently retired from active work as a carpenter because he had 'trouble' with his sight. His wife thought he had become rather lazy, even though he was actually mentally alert.

Clinical examination revealed 6/6 vision in each eye with spectacle correction for slight hypermetropia.

He could easily read N5 with his bifocal addition at one-third of a metre. His ocular muscle balance was normal and he had no difficulty with convergence.

When it was pointed out that his bifocal prescription was appropriate and that he could read small print with his glasses, he mentioned that his difficulty occurred when playing cards. He explained that he could not clearly see the figures and numbers, adding that one does, of course, keep one's cards close to one's chest.

Further ocular examination revealed:

- absent voluntary downgaze
- reduced voluntary upgaze
- intact doll's eye reflex movements (oculocephalic reflex).

An examination of the patient's motor system showed rigidity of the neck muscles. His speech was slurred and he walked with a slightly broad-based gait. There was no tremor.

His visual difficulty was thus seen to be related to the defect in down gaze.

Table. Progressive supranuclear palsy and Parkinson's disease

Feature	Progressive supranuclear palsy	Parkinson's disease
Onset	Earlier (fifth decade)	Later (sixth decade)
Initial symptoms	Postural symptoms, gait disorder, personality change	Bradykinesia, tremor at rest
Gait	Broad-based, stiff, unsteady	Short, slow, shuffling
Tremor	Not present	Present
Posture	Erect, with head tilted back and a tendency to fall backwards	Stooping, bent forward
Speech	Irregular, explosive in quality	Soft in volume, a rapid succession of words
First gaze affected	Downgaze	Upgaze
Muscle rigidity	Present in axial musculature (particularly neck muscles), resulting in backward arching of the neck and difficulty controlling sitting (a tendency to 'fall into a chair')	More generalised, with stiffness in the limbs and more difficulty using the hands
Facial expression	Grimacing, anxious or startled	'Ironed-out' or mask-like in appearance (hypomimia)
Cognition	More severely affected (mental slowing and deficits in attention); general loss of interest or increased irritability	Less severely affected

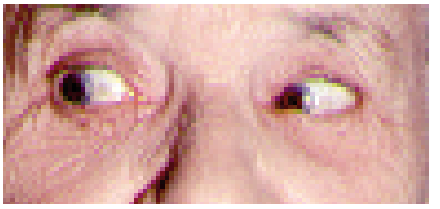

Figure 3a. Normal right gaze.

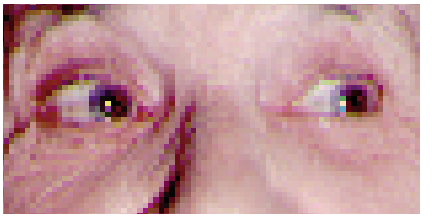

Figure 3b. Normal left gaze.

downgaze (such as reading) are affected first, and patients have difficulties with stairs and driving. They tend to be 'messy eaters' at the table and may show the 'dirty tie' sign.

Ocular examination

An analysis of eye movements will show saccadic anomalies (loss of voluntary vertical saccades and small slow vertical saccades) and continuous square wave jerks. Smooth pursuit movements of progressive supranuclear palsy are relatively preserved. On the other hand, in Parkinson's disease, stuttering eye movements are seen during smooth pursuit movement (caused by catch up saccades), and the saccades themselves are hypometric with an increased intersaccadic interval, and there is also associated convergence weakness and abnormal head movements.

Management

Progressive supranuclear palsy is a clinical diagnosis – blood tests, CT and MRI are usually not helpful. Recognising that the visual problem is caused by progressive supranuclear palsy will prevent unnecessary investigations or change of glasses. The task or activity affected should be considered in any visual problem experienced by the patient.

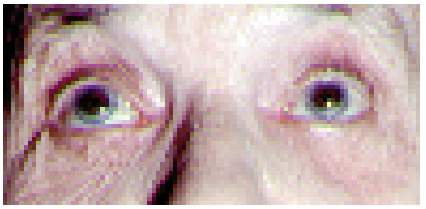

Figure 3c. Normal up-gaze.

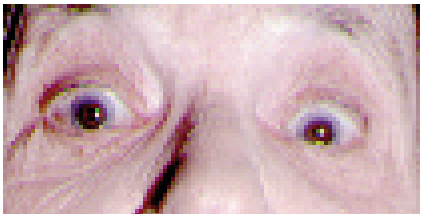

Figure 3d. Defective down-gaze.

Unfortunately, there is no known effective treatment for progressive supranuclear palsy. The neuropathology overlaps that of Parkinson's disease, but there is no benefit from drugs such as dopamine agonists and levodopa. As the disease follows a progressive course, management becomes essentially supportive. General health and nutrition need to be maintained. Some of the patient's difficulties can be overcome with suitably designed walking aids and hand rails installed in the home. Much depends on the quality of care.

Progression

Progressive supranuclear palsy is slowly but relentlessly progressive – early symptoms get worse and new symptoms develop. Eventually walking becomes difficult and the patient may become confined to a wheelchair. Associated pseudobulbar palsy affects speech and swallowing. Doll's eye movements are intact at first, but Bell's phenomenon is commonly absent. A deficit of eye movement can progress to complete ophthalmoplegia.

There are abnormalities of eyelid movements and patients experience difficulty opening or closing the eyes. The resulting eyelid freezing and stare may be misdiagnosed as blepharospasm or apraxia of lid opening. A coexisting ocular or orbital condition can mislead and cause a delay in diagnosis (see the box above).

Final comments

The clinical features of progressive supranuclear palsy can be subtle – particularly in the early stages – giving an opportunity for misdiagnosis. The disease is mistakenly diagnosed as Parkinson's disease, and is known as its 'ugly cousin'. Visual problems develop in all cases and the impairment of vertical gaze is easy to detect.

There is a clear burden on the carer. The patient can be very demanding and may be seen to be difficult and irritable. The dementia may escape recognition because it is mild and only slowly progressive. It should be understood that the personality changes are part of the illness. Patients usually need full-time care in the later stages, and the plight of the carer at home should not be ignored. MT

A case of malingering?

A 60-year-old man was referred for an ophthalmic assessment. He had been treated for an overactive thyroid 10 years previously. Following treatment, his eyes had became prominent and he was now incapable of moving them. A diagnosis of total ophthalmoplegia from thyroid eye disease (orbitopathy) had been made, but it had not been a firm diagnosis and there was an impression that he was malingering. He was aware that doctors did not quite believe his story.

On examination, he was pleasant and co-operative. His wife answered most questions about his present complaint and medical history. His answers were brief and given in a hoarse monotone.

His visual acuity was 6/18 in each eye. There was no correctable refractive error, and his reduced vision could be explained by macular degenerative changes seen on ocular fundoscopy. He had bilateral symmetrical proptosis and lid retraction, but no congestive signs. There was increased resistance to passive retropulsion of the globe (often elicited in thyroid orbitopathy).

The patient had a fixed vacant stare, and looked straight ahead. He did not move his eyes on command and had a complete absence of voluntary and involuntary eye movements. Doll's eye reflexes could not be elicited. He was able to look to the sides only by turning his body. The resistance to passive head turns during the test came from the marked nuchal rigidity. The optokinetic nystagmus response could not be obtained by rotation of the optokinetic drum, which verified the patient's story and eliminated any possibility of malingering.

This case illustrates that a coexisting ocular or orbital condition or a previous diagnostic label can mislead and cause a delay in diagnosis.

Further reading

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