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Multiple system atrophy- An updated review for medical practitioners

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Introduction: Multiple system atrophy (MSA) is a rare group of neurodegenerative syndromes including olivopontocerebellar atrophy, striatonigral degeneration, and Shy-Drager syndrome. The syndrome is rare and not familiar to many clinicians.

Aim and Significance: To provide an updated review of the disease for clinicians to be aware of this rare, but important disease in their daily practice.

Methods: A systematic review was performed using the PUBMED and MEDLINE databases.

Review and Discussion: MSA is a group of neurodegenerative disorders, typically affecting patients >50 years, with an estimated incidence of 3/100,000. To date, no established environmental risk factors for MSA have been reported, although data are limited.

Clinically, the main features of MSA are akinetic-rigid parkinsonism, autonomic failure including urogenital dysfunction, cerebellar ataxia, and pyramidal signs in different combinations. Autonomic dysfunction such as severe variability in blood pressure and heart rate, postprandial hypotension, orthostatic hypotension, and syncope is frequently seen. There are two different, often overlapping, clinical subtypes of MSA: MSA with predominant parkinsonism (MSA-P) subtype and MSA with predominant cerebellar ataxia (MSA-C) subtype. The etiology of MSA is unknown.

The diagnosis of MSA is based upon the clinical features. No laboratory or imaging studies are diagnostic, particularly since findings are often normal or vague in early disease. However, to exclude other conditions, neuroimaging can be helpful and may show signs of putaminal atrophy, slit-like signal change at the posterolateral putaminal margin, and hypointensity of the putamen relative to the globus pallidus. The "hot cross bun sign", that is sometimes seen, refers to hyperintense T2 signal in the shape of a cross within the pons that arises from degeneration of transverse pontocerebellar fibers. Importantly, the lack of dramatic and sustained response to levodopa, is one of the features that distinguish MSA from idiopathic Parkinson disease.

Disease progression in MSA usually occurs over one to 18 years, and is often faster than that of idiopathic Parkinson disease. The median time from MSA onset to death is 6 to 10 years.

No treatment is available for MSA. Management is symptomatic, and although the autonomic symptoms may respond to pharmacotherapy, the motor symptoms in MSA are highly resistant. Physical, occupational, and speech therapy are helpful for the supportive care. Several medications are currently under investigation, including reducing alpha-synuclein aggregation using rifampicin and rasagiline and infusing autologous mesenchymal stem cells which theoretically may have a neuroprotective effect.

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