PARKINSONIAN DISORDERS **MEDICAL APPENDIX**

(Including Parkinsonism, Parkinson's disease, Parkinson's syndrome, paralysis agitans)

DEFINITION

1. The Parkinsonian disorders are diseases of the extrapyramidal system or basal ganglia. The six paired nuclei that constitute the basal ganglia are the caudate nucleus, the putamen, globus pallidus, nucleus accumbens, subthalamic nucleus and substantia nigra. The term corpus striatum refers to the caudate nucleus and putamen, while the putamen and globus pallidus combined are termed the lenticular nucleus.

2. The relation of the basal ganglia and associated structures is complex. The putamen is concerned with motor function and the caudate nucleus with emotional and cognitive functions. The basal ganglia and higher centres produce transmitter substances which have been identified as glutamic acid, dopamine, acetylcholine, noradrenaline and gamma aminobutyric acid. Physiological investigations have shown that the primary role of the basal ganglia is to scale the amplitude and velocity of movements rather than to initiate movement. The basal ganglia structures thus have a critical role in preparation for and execution of movement. Basal ganglia dysfunction causes impaired voluntary movement and abnormal involuntary movements.

3. Parkinsonian disorders are classified as primary or secondary. Primary Parkinsonism is a diagnosis made by excluding the causes of secondary Parkinsonism. The clinician is alerted to the possibility of secondary Parkinsonism by atypical Parkinsonian clinical features and by the signs of coexistent diseases.

CLINICAL MANIFESTATIONS

4. Classic primary Parkinsonism is characterised by the four cardinal signs of tremor at rest, rigidity, bradykinesia and postural instability. There may also be associated mood and intellect changes, oculomotor control problems and changes in the autonomic and sensory systems.

5. Of all the possible signs and symptoms the most common are rest tremor and bradykinesia. Bradykinesia may give rise to a generalised slowing down of movements, lack of facial expression and decreased frequency of blinking, dysarthria, monotonous speech, drooling, small handwriting, difficulty in rising from a chair or turning in bed, a shuffling gait of little steps, decreased automatic movements such as arm swinging, start hesitation and freezing.

6. The motor signs are due to levodopa depletion, the depression and "freezing" to low noradrenaline levels and the dementia which affects about 1/3 of Parkinsonism patients to low acetyl choline levels.

AETIOLOGY

Primary Parkinsonism

7. The cause of this is unknown. It is a common condition which affects one person in a thousand. It is of uniform distribution throughout the world and is slightly more common in men. It is rare in those under 50. In about 5-10% of cases there is a family history but identical twin studies have failed to reveal concordance and it is generally agreed that it is not a hereditary condition.

8. There is no evidence of a viral or other infective origin for primary Parkinsonism. No environmental causal factor has been identified.

9. Patients with primary Parkinsonism smoke less than the general population. The significance of this is unknown.

Secondary Parkinsonism

10. This occurs in relation to infection (particularly encephalitis), therapeutic drugs, toxins, severe trauma and, rarely, in relation to vascular disease.

11. The diagnosis of post-encephalitic Parkinsonism is suggested by a history of encephalitis lethargica, features of Parkinsonism plus the additional features of post-encephalitis. These include spasms of eye deviation and oculogyric crises, pupillary disorders and dystonias. The onset of post encephalitic Parkinsonism is usually within ten years of the primary infection. In general the course of this form is benign.

12. Neuroleptic drugs such as the phenothiazines, reserpine, tetrabenazine, methyl dopa, and lithium may also lead to secondary Parkinsonism. In 95% of such cases withdrawal of the offending drug leads to remission of the Parkinsonian features within weeks or months.

13. Secondary Parkinsonism may occur as a result of heavy metal ingestion, including manganese, cobalt and mercury.

14. The condition may occur in relation to diffuse cerebrovascular disease resulting from atherosclerosis.

15. Secondary Parkinsonism may also follow generalised brain anoxia as a result of cardiac arrest or carbon monoxide poisoning. It may also complicate severe diffuse brain trauma. This is seen most characteristically in the punch-drunk syndrome. A head injury which might lead to Parkinsonism would be profound and the time interval between trauma and appearance of basal ganglia features is short.

CONCLUSION

16. Parkinsonism is due to a disorder of the extrapyramidal system or basal ganglia. It may be a primary condition, the aetiology of which is unknown but for which no causal environmental factor has been identified. It may be secondary to other diseases, toxins or severe head trauma.

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