Parkinsonism secondary to neurosyphilis

A case report

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Summary

A case of parkinsonism associated with neurosyphilis, which improved markedly after appropriate treatment, is described. Although neurosyphilis is a rare cause of parkinsonism, it should be considered when parkinsonism appears in the 30-50-year age group, particularly if other anomalous neurological findings are present. Adequate therapy may improve the patient's condition and prevent further progression.

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Although the association between neurosyphilis and parkinsonism is rare, it is accepted that *Treponema pallidum* can cause the features of this condition.[1] The signs of involvement of the basal ganglia may appear soon after the initial infection or may be delayed for some years.[1] Usually the classic neurological abnormalities of neurosyphilis such as pupillary signs, optic atrophy, pyramidal signs, sensory loss and abnormal cerebrospinal fluid (CSF) findings are found together with the parkinsonism.

Case report

A 48-year-old Black man was admitted to hospital complaining of chest pains and a productive cough of 1 week's duration. On examination he was demented, malnourished and dehydrated, pyrexial (38,9°C) and pale. His fingers were clubbed but there was no jaundice or cyanosis. Bronchial breathing was heard over his left dorsal lung base. No other abnormality was detected and a tentative diagnosis of lobar pneumonia was made and confirmed by chest radiographs.

A neurological examination performed during his stay in hospital revealed that he was poorly orientated for time, space and person and that he had parkinsonism. There was no history of parkinsonism in the family and the patient as well as his relatives denied drug intake (e.g. phenothiazines or reserpine) and exposure to industrial toxins.

Careful assessment of the patient revealed a bilateral resting tremor of 4 - 7 Hz in the upper limbs, cogwheel rigidity in the wrists and neck muscles, a positive glabellar tap, poor eye blinking and limited facial expression. His gait was slow, shuffling and hesitant. He had bilateral primary optic atrophy and his pupils were small (3 mm), irregular and failed to respond to light, but accommodation was intact (Argyll Robertson pupils). There was a mild bilateral coarse horizontal nystagmus and weakness of upward gaze, sensory hearing loss on the right (tuning fork examination), and wasting of the tongue causing dysarthric and slow speech. Examination of the limbs disclosed slight generalized weakness and bilateral brisk tendon jerks with slight predominance in the lower limbs. The left plantar response was extensor and the right flexor. Hoffmann's sign was negative. Frontal lobe release signs were present (pout, palmomental and head retraction). Co-ordination was impaired and there was some degree of dysdiadochokinesia and intention tremor in the upper limbs. Perception of pin-prick was normal in the arms and legs, but position sense and vibration sense were impaired in all limbs, predominantly on the right. A marked degree of rombergism was also present. His sphincters as well as testicular sensation were normal.

Laboratory analysis revealed a white blood count of $14,2 \times 10^9$/l, a haemoglobin value of 9,7 g/dl, an erythrocyte sedimentation rate of 40 mm/1st h (Westergren), and normal serum electrolyte and glucose values, liver and renal function, vitamin B_{12} and folate levels. The Wassermann reaction in serum was repeatedly positive. The fluorescent treponemal antibody absorption (FTA-ABS) test was strongly positive in the blood and CSF. On lumbar puncture the CSF was under normal pressure (160 mm H_2O) and clear with 105 cells (80% lymphocytes), protein 1,1 g/l (normal 0,1 - 0,4 g/l), glucose 2,6 mmol/l (normal 2,5 - 5,5 mmol/l), and chloride 118 mmol/l. Bacteria were not observed. Computed tomography showed no mass, but features of communicating hydrocephalus associated with moderate cortical and cerebellar atrophy were present.

The patient was given a course of procaine penicillin G (600 000 U intramuscularly daily for 21 days) which followed a 3-day administration of prednisone 60 mg daily to avoid a possible Jarisch-Herxheimer reaction. When seen 3 months later, he was much more alert and less handicapped by his parkinsonism. He was able to walk without much support, although his gait remained slow with poor arm swing. The initiation and accomplishment of movements improved significantly and there was no retro-, pro-, or anteropulsion. The rigidity in his limbs decreased and there was only slight residual trembling in his hands. His speech was clearer and the facies less expressionless. Repeat lumbar puncture revealed clear fluid with 31 cells (mostly lymphocytes), protein 0,82 g/l and normal glucose content. The FTA-ABS test was positive.

Discussion

This patient had typical parkinsonian signs, Argyll Robertson pupils, a positive Babinski reflex on the left, cerebellar signs, impaired intellectual capacity and abnormal CSF findings.

Although there is little doubt that acquired syphilis can cause a parkinsonian syndrome, the association of these two conditions in the same patient could have been coincidental.[2] However, the fact that a significant improvement in the patient's condition and the CSF findings followed treatment with penicillin strongly suggests that the syphilitic infection of the nervous system was in some way connected with the development of the parkinsonian syndrome as well as the associated neurological signs. It is possible that cure of the pneumonia and the patient's rehydration contributed to the improvement of his parkinsonism.

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Parkinsonism is a rare sequel of neurosyphilis. There are only 59 reported cases of syphilis as a cause of Parkinson's disease. The first one by Hutchinson[3] appeared in 1898. The syphilitic manifestations were thought to be hereditary and the patient subsequently developed paralysis agitans in his forties. This is interesting historically because there was no evidence that parkinsonism had any relation to the syphilis. In 1953 Neill[4] described a case of a man aged 43 who developed parkinsonism slowly over 2 years, with tremor, rigidity and other typical signs of the disease. He was treated vigorously with penicillin, and a significant improvement of his parkinsonism was noted. In 1954 Lit[5] described 2 patients both of whom improved on eradication of the syphilitic process, one much more than the other. In 1968 Siegfried[6] reported a case in which parkinsonism was associated with neurosyphilis, but no mention of response to therapy was made.

The pathological findings associated with meningovascular syphilis suggests that the inflammatory process may occasionally spread to involve basal ganglia and other mesencephalic structures. Wilson and Cobb[7] introduced the term 'mesencephalic syphilis' to describe the pathological association between neurosyphilis and parkinsonism. Mella and Katz (quoted by Neill[4]) recorded typical mesodermal syphilitic involvement of the basal ganglia in a 46-year-old man with a diagnosis of paralysis agitans. Autopsy revealed perivascular infiltration in the globus pallidus, chronic leptomeningitis, atrophy of the frontal lobes and granulations in the fourth ventricle, all pathognomonic of neurosyphilis. Stevenson and Allen (also quoted by Neill[4]) recorded a similar case of parkinsonism which showed typical changes of dementia paralytica in the cerebral cortex together with widespread lesions in the corpus striatum and its connections at autopsy.

Since neurosyphilis definitely affects the midbrain area and small vascular lesions throughout the mesencephalon have been described, it would seem logical to conclude that there is an aggravation of an underlying parkinsonian process in the majority of the so-called parkinsonian syphilitic patients, and in a few rare cases the luetic lesion is responsible for the entire parkinsonian picture, since it may disappear when proper treatment is instituted. In the present case it seems that the parkinsonian picture was probably aggravated by an underlying neurosyphilis since an improvement (but not complete cure) of the patient's condition occurred after appropriate therapy had been instituted.

This case report is noteworthy in that neurosyphilis and Parkinson's disease are both uncommon conditions in the Black population. Neurosyphilis should therefore be considered in the differential diagnosis of parkinsonism, specifically when other anomalous associated neurological symptoms are present. In this event adequate therapy may improve the patient's condition and help in averting further progression of the parkinsonism.

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