

Case 1995-4

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Clinical History

A right handed white man presented at 43 years of age with impaired balance and difficulty using the left arm. There was no family history of neurologic disease. He had a five year history of hoarseness, mild dysarthria and a tendency to choke on liquids. Constipation and urinary incontinence developed in the two years before he presented and he had had a several year history of increasing depression. For the year prior to his presentation he noted problems with gait, particularly with turning and for the previous six months he experienced trouble using his left arm when dressing and slowness in typing. Over the weeks preceding presentation he noticed a tendency to "gape", holding his mouth open inappropriately and he developed difficulty with his left leg when shifting gears and a tendency to limp when fatigued. He had lived and worked on a farm and had been in contact with a variety of pesticides. He had a history of manic-depressive illness.

On examination at 43 years of age he was well developed and well nourished. BP was normal both supine and sitting. Pulse was 72. General physical exam revealed an enlarged, smooth prostate and vitiligo involving both arms and trunk. Neurologic exam revealed normal orientation, memory, normal language skills, praxis, calculation ability and left-right discrimination. Ocular fundi and visual fields were normal. Cranial nerve examination revealed a slight tendency to stare and lag of the left mouth on smiling. He had normal muscle bulk with "lead pipe" rigidity in the extremities, greater on the left side but there was no true pill-rolling tremor. Voluntary movements were slow, much more so on the left with a mild left pronator drift. Deep tendon reflexes were 3+ throughout with bilateral extensor plantar responses. Cerebellar exam was normal apart from slowed rapid alternating movements in the left arm. He had difficulty initiating gait and had en bloc turning, decreased armswing, and flexed posture. Sensory exam was normal. Cranial MRI was normal and he was treated with Sinemet. Four years later his principal complaint was frequent falls, especially when making turns. He also had episodes of "freezing up" and swings in his degree of bradykinesia. He walked with small steps. He was lost to followup until he died, aged 51 years.

GROSS AUTOPSY FINDINGS:

The brain weighed 1660g and had mild atrophy of the frontal and parietal lobes. The putamen showed bilateral moderate atrophy and the substantia nigra was very pale. The cerebellum was moderately atrophic and the basis pontis was slightly shrunken. Thalami, amygdalae and hippocampi were normal to gross inspection. The ventricles measured 54 ml.

MATERIAL SUBMITTED:

One luxol fast blue-H&E and one unstained section.

POINTS FOR DISCUSSION:

1. Diagnosis
2. Specificity of abnormal features