CASES DEMONSTRATED AT THE M.R.C.P. CLASS.

Case of Cerebral Arterio-Sclerosis with Bilateral Hemiplegia.

H.S., aged 48 years, was first seen in January, 1934, complaining of difficulty in articulation and weakness of the legs.

History.—About 3 years ago while the patient was lying down he experienced an attack of "pins and needles" in the right leg which was immediately followed by muscular weakness. Shortly afterwards the right arm also felt weak. There was no vomiting or loss of consciousness. He returned to work in about three weeks, but the right leg has felt somewhat weak ever since.

In August, 1933, he noticed that the left leg appeared to drag and that the little and ring fingers of the left hand seemed numb; these symptoms have persisted. In November last he states that he appeared to be talking as if he had a plum in his mouth, and about six weeks ago he began to complain of further weakness and tightness in the lower portion of the left leg. He has also been told that he has become quick-tempered. There has been no headache or giddiness.

Previous History.—History of right hip disease at 7-10 years of age, presumably tuberculous in nature.

Physical Examination.


Speech slow and somewhat slurred. Slight right-sided weakness of lower half of face.

No sensory abnormality beyond diminished deep pressure sense.

Arms: Extension of right elbow somewhat weak and arms somewhat spastic. Supinator, biceps and triceps jerks brisk, right greater than left.

Legs: Slight wasting of right leg amounting to $\frac{1}{16}$" in thigh and $\frac{1}{4}$" in calf. Knee and ankle-jerks brisk and about equal. Right ankle clonus—not left. Plantars both extensor. The right hip is ankylosed.

Gait shuffling and inclined to be tottering.

Incoordination of both arms on finger-nose test. Romberg's sign negative. Sphincters not affected.

Abdominal reflexes absent.
Other Systems: Heart slightly enlarged, with accentuated second sound.
B.P. 216/148.
Blood Wassermann reaction, negative.
Cerebro-spinal fluid:
   2 lymphocytes per c.mm.
   Total protein: 0.04%.
   Globulin: faint detectable trace.
W.R. and Lange, negative.
Blood Urea: 39 mg. per cent.
Urine shows trace of albumin.
Urea in urine: 2,000 mg. per cent.
Urea concentration factor, 51.

Discussion.
The case is one of arteriosclerosis—particularly cerebral arterio-sclerosis—in which bilateral cerebral thrombosis has occurred. The branches of the middle cerebral arteries are chiefly affected, the left side of the brain being more involved than the right. On the latter side, the facial area has escaped and the leg is more affected than the arm. The bilateral lesion has led to a slight degree of "pseudo-bulbar paralysis" shown chiefly in the dysarthria. The tongue and pharynx would have been more profoundly affected had the thrombosis involving the right side of the brain been more severe. This thrombosis began to develop only in August last, while that on the left side occurred three years ago. There is no aphasia.

The diagnosis is fairly straightforward. The best guide to the state of the cerebral arteries is not necessarily the condition of the palpable arteries (radial, brachial, etc.) nor the blood pressure, but the state of the retinal arteries as seen on examination of the fundus oculi. With the history in this case, a provisional diagnosis could be made from the condition of the right fundus. Less than half the candidates noticed these changes. Most of them arrived at a correct diagnosis on the history and neurological signs. A few missed the signs of upper neurone involvement affecting the left leg and were content with having found those on the right side.

Some made a diagnosis of disseminated sclerosis. Against this, in addition to all the signs of arterial degeneration, is the complete absence of any evidence of cerebellar or of cerebellar tract involvement—nystagmus, intention tremor, Romberg's sign, etc.—as well as the absence in the history of any true remissions. The negative Lange reaction in the cerebrospinal fluid would be another point against disseminated sclerosis. Very few who examined the case mentioned the possibility of syphilitic thrombosis, although with an onset at the age of 45, syphilis should first have been excluded by the Wassermann reaction. Nobody noticed the ankylosed right hip.

Case shown by C. Worster-Drought, M.D.
Cerebro-Macular Degeneration—Juvenile Form approximating to the Batten-Mayou Type.

H.G., aged 18 years, complains of stiffness and weakness of both arms and legs, inability to walk and failing vision.

**History:** Gradual onset during last 4½ years, starting in the left arm, spreading to the right arm and then to both legs.


"Fundus: On the right side, primary optic atrophy; disc extremely white with macular degeneration. Extending from the macula towards the disc is a circular area of retinal atrophy with speckled areas of pigment. On the left side, definite optic atrophy involving the papillo-macular bundle. The macula itself shows only increase of pigment." (Mr. A. Sorsby)

Postural sense and localisation poor in fingers and toes: sensation otherwise normal.

Arms of both sides are somewhat wasted, very spastic in extended position and usually held in flexion at wrist with fingers extended, the forearm showing a tendency to hyperpronation. All fingers can be moved voluntarily but in an awkward manner. Arm jerks unobtainable—inhibited by muscular rigidity.

The legs show clasp-knife rigidity, especially the right. Patient can overcome it to a certain extent.


Abdominal reflexes not obtained as abdominal muscles are in spastic contraction. No spontaneous involuntary movements. No definite ataxia of arms or legs. Sphincters normal.

Blood W. R. negative.

Cerebro-spinal fluid: no abnormality, with negative Wassermann reaction.

The patient exhibits a certain amount of emotional instability, laughing and crying with very little provocation. Intelligence is only very slightly impaired.

**Discussion.**

The diagnosis in this case is admittedly difficult. The patient was brought forward mainly to ascertain if the physical signs would be correctly made out and how they would be interpreted. A large proportion of the candidates recognized
the optic atrophy. Nobody noted the changes in the region of the macula. This, however, was to be expected. Most had great difficulty in deciding whether the muscular rigidity present was pyramidal or extra-pyramidal. As a matter of fact the spasticity affecting the arms is mainly extra-pyramidal, as indicated by their position and the inhibited deep reflexes, while that affecting the legs is a mixture of both, although pyramidal spasticity probably predominates. This is suggested by the exaggerated knee and ankle jerks and the indefinite plantar reflexes. Were the rigidity exclusively extra-pyramidal the plantar reflex would have been definitely flexor. Earlier in the course of the disease extensor reflexes were noted. Otherwise, most of the physical signs were correctly recorded.

Among the conditions suggested in diagnosis and which have to be considered are the following:—

(1) **Disseminated sclerosis.** In addition to the rarity of this disease originating at the age of 14 years, there is little to suggest such a diagnosis apart from emotional disturbance and a slight degree of euphoria. The type of rigidity of the arms (extra-pyramidal) is in itself almost exclusive and further, there are no signs of cerebellar involvement nor have there been any remissions since the onset of the illness.

(2) **Marie type of hereditary ataxia.** It is true that optic atrophy may occur in this variant of heredito-familial ataxia but the case does not show any real ataxia. The awkwardness of movement in the arms and legs is solely due to the spasticity. Further, the knee and ankle jerks in the Marie type of ataxia are more likely to be absent than exaggerated.

(3) **Progressive lenticular degeneration (Wilson’s disease).** A more praiseworthy attempt at diagnosis. The emotional changes are suggestive as well as the type of spasticity. The main fact against this diagnosis is the absence of spontaneous involuntary movements as well as the changes in the fundus oculi and the lack of evidence of hepatic disorder.

(4) **Schilder’s periaxial encephalitis.** If this were the diagnosis one would have to imagine that only the parietal lobes were affected and that the disease had spared the temporal and occipital lobes and almost entirely the frontal lobes. This is almost inconceivable. In addition, Schilder’s disease is one that affects exclusively the white matter of the brain; therefore, any indications of extra-pyramidal changes would exclude this diagnosis. Optic atrophy has been recorded in some cases of Schilder’s disease.

(5) Detection of the changes in the region of the macula, however, would exclude all the above conditions. The case admittedly is by no means a typical example of the Batten-Mayou type of cerebro-macular degeneration in that, so far, intellectual deterioration has been comparatively slight while the motor parts of the brain are much more affected than usual.

*Case shown by C. Worster-Drought, M.D.*