SOME UNUSUAL CASES OF NEUROSYPHILIS*†

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Case Reports

Case 1, a coal-miner aged 50, came to hospital on November 1, 1956. He had been struck across the bridge of the nose by a wire rope 3 weeks earlier, but the injury was trivial and he had continued to work. After a few days he noted that the sight of his left eye was dim and misty. His right eye became affected 2 weeks before admission. He noted that if he looked straight ahead his vision was adequate, but he could not see clearly to either side. During this period of 8 weeks he had dull occipital headaches, and, on being questioned, he said that he had suffered from headaches over the vertex of the skull for a year. He had attributed them to drinking,

but they had not disappeared after giving up alcohol 3 months earlier. He had no other symptoms.

Apart from accidents at work, one of which had led to the amputation of his right foot, his health had been excellent and he had no knowledge of previous venereal infection.

Examination.—The patient was alert, cooperative, and intelligent, with no impairment of memory.

Central Nervous System:

Cranial Nerves:

I. No disturbance of smell.

II. Bitemporal hemianopia (Fig. 1). Ophthalmoscopic examinations showed pallor of both optic disks, especially the left, the appearance of which suggested early atrophy.

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FIG. 1.—Case 1. Visual acuity in the left eye $<6/60$ and J17; in the right eye 6/9 and J1.

III, IV, VI. No motor disturbance. The pupils were equal and round, and reacted to light but not to accommodation.
Other: No abnormality.

X ray of Skull:

No abnormality; pituitary fossa normal.

Reflexes:

Arm: active and equal,
Knee: present,
Ankle: left absent (right could not be tested),
Plantar: left flexor (right could not be tested).

Blood:

Wassermann reaction strongly positive (20 doses);
Kahn test strongly positive.

Cerebrospinal Fluid:

Cells 114 per ml. (90 per cent. lymphocytes);
Protein 107 mg./100 ml.;
Wassermann reaction strongly positive;
Gold Sol 4553343120.

Treatment.—Potassium iodide was given, followed by injections of penicillin up to 40 mega units in 3 months.

Progress.—The headaches were relieved, but the visual acuity deteriorated rapidly and within 2 months the left eye had perception of light only, and the disk was diffusely pale. The field of vision in the right eye diminished very little, and though the atrophy came very close to the centre it has not yet involved the macula and the visual acuity for distance is 6/6. The pupil of the right eye now reacts very slowly to light but this reflex is absent in the left eye. The cerebrospinal fluid has

improved rapidly, but though the cell count is normal the protein content is still abnormally high, the Wassermann reaction is positive, and the Gold Sol on April 18, 1957, was 1112100000.

Discussion.—At first this case simulated a pituitary tumour very closely, the pattern of headaches and bi-temporal hemianopia being typical, but there were no x-ray changes in the skull and there were characteristic changes of syphilis in the cerebrospinal fluid.

The nature of the syphilitic lesion was discussed and the possibility of a gumma at the posterior aspect of the optic chiasma was considered. Surgery was ruled out, and the effect of specific treatment for syphilis was watched. As vision was failing rapidly, we had to give iodides and penicillin in adequate doses. There was no Herxheimer reaction, and for a short time the fields of vision appeared to remain unchanged, or even to improve (Fig. 2). Within a month, however, evidence of deterioration of the left eye was clear, and rapid loss of the visual field occurred, going on to virtual blindness of that eye by mid-January, 1957, *i.e.* within 10 weeks of starting treatment (Figs 3 and 4, opposite). The disk was obviously atrophic. It was then clear that we were dealing with optic atrophy which had started as a posterior chiasmic lesion and was progressing to bilateral loss of field. This is seen in Fig. 5 (p. 152).

FIG. 2.—Case 1. Visual acuity in the left eye 6/36 and J16; in the right eye 6/6 and J4.

It is known that the region of the chiasm is a favourite area for basilar syphilitic meningitis, and occasionally a gumma is found there with little

meningeal involvement. Igersheimer (1928) has noted the tendency of the temporal field defect to progress so that the entire field of one or both eyes

FIG. 3.—Case 1. Visual acuity in the left eye 6/60 – 36 and J20; in the right eye 6/6 and J1.

FIG. 4.—Case 1. Visual acuity in the left eye 1/60 and >J20; in the right eye 6/6 and J1.

FIG. 5.—Case 1. Perception of light only in the left eye; visual acuity in the right eye 6/6.

may be lost. If a diagnosis of syphilitic arachnoiditis could have been established early, it might have been advisable to treat this man surgically, attempting to relieve the tension in the nerve sheaths.

Case 2, an engineer and bookmaker aged 55, was an intelligent skilled man who noted about October, 1956, that the sight of his left eye was defective. He had suffered from pain at the backs of the eyes for 2 or 3 years. He was a non-smoker and drank very little alcohol, and his previous health had been excellent. There was no history of venereal disease.

Examination.—The patient was alert, intelligent, and cooperative. He had no skin or mucous membrane signs of syphilis.

Central Nervous System: The pupils were small, round, equal in size, and reacted well to light and convergence.

Cranial Nerves: Ophthalmoscopic examination showed that both disks were distinctly pale, the left being paler than the right. A central scotoma was present in the left field of vision; the right field was normal (Fig. 6, opposite). The other cranial nerves were normal.

Reflexes:

Arm: active and equal,
Knee: unequal, the right being very sluggish or absent and the left active,
Ankle: active and equal,
Abdominal: present,

Plantar: flexor.

No impairment of sensation detected. Romberg's test normal.

Organic: normal, though perhaps there was some loss of libido as his most recent coitus was in 1944.

Cardiovascular System: Heart was slightly enlarged and alteration of tone of the heart sounds was noted in the aortic area. Electrocardiograph showed myocardial disease, almost amounting to bundle branch block. X ray of heart not abnormal.

Blood:

Wassermann reaction strongly positive;
Kahn test strongly positive;
Syphilis flocculation reaction strongly positive;
Kahn verification test strongly positive.

Cerebrospinal Fluid:

Cells 270 per cm.;
Protein 280 mg./100 ml.;
Wassermann reaction strongly positive;
Gold Sol 4555554330.

Diagnosis.—Here we had a case of neurosyphilis and cardiac syphilis presenting as a central scotoma of one visual field, and cerebrospinal fluid findings indicating very active meningo-vascular syphilis or possibly general paralysis of the insane. The pallor of both disks made us think that the lesion was early optic atrophy of an unusual type, possibly associated with involvement of the blood supply to the left optic nerve.

Treatment.—Iodides by mouth and penicillin were given, the latter amounting to 20 mega units.

FIG. 6.—Case 2. Visual acuity in the left eye $< 6/60$; in the right eye $6/6$.

Progress.—After this treatment the cerebrospinal fluid was re-examined and reported as follows:

Cells 9 per cmm.;
Protein 120 mg./100 ml.;
Wassermann reaction strongly positive;
Gold Sol 5555443200.

Further Treatment.—Bismuth 2g. in 10 weeks was given, and the cerebrospinal fluid showed little change. The patient is now receiving additional penicillin.

Progress.—The visual fields have been charted regularly and there has been no improvement or impairment in a period of 6 months. His general health has improved greatly, he has greater energy, and the retrobulbar headaches have disappeared.

Discussion.—Central scotoma is a rare form of early optic atrophy. A lesion at the anterior part of the optic chiasma could have caused a central scotoma. Also a central syphilitic retinitis was possible, but was excluded in this case on account of the pallor of both disks. Endarteritis and thrombosis of the central retinal artery were possible but they have a different ophthalmoscopic appearance. Retrobulbar neuritis may be considered, and in this case was unilateral.

The literature generally places emphasis on the characteristic loss of peripheral visual fields of syphilitic optic atrophy; Uhtloft (1947) estimated that central scotoma occurred in 2 per cent. of cases,

and was accompanied by peripheral field defects. Sloan and Wood (1938) found central scotoma in 53 per cent. of their cases of syphilitic optic atrophy.

In this case the apparent arrest of visual failure and the improvement in the cerebrospinal fluid and the cardiac condition have encouraged us to hope that the disease will be arrested.

Case 3 (General Paralysis of the Insane), an unmarried bus driver aged 50, drove his bus along the wrong route one day, paying no attention to passengers or halts, and was abusive when protests were made. He stated that he knew what he was doing and he realized that his conduct was wrong. His previous health had been excellent, and he had no history of previous nervous or mental disease or venereal infection. He drank very little alcohol and had never been a heavy drinker.

Examination.—The patient was placid, helpful, and correctly orientated, and seemed to have a good memory and a lively interest in everything around him. He described the incidents leading to his medical examination with clearness and insight, but was unconcerned or mildly amused about them. Physical examination showed no abnormality.

Central Nervous System. The pupils were equal and reacted briskly to light and convergence, there were no tremors of tongue or face and no disturbance of vision or eye movement. Ophthalmoscopic examination showed no abnormality.

Reflexes:

Deep reflexes active and equal,
Plantar: flexor.
No disturbance of sensation found.

Cardiovascular System: Normal.

Blood: Strongly sero-positive to tests for syphilis.

Cerebrospinal Fluid:

Cells 36 per cmm.;
Protein 111 mg./100 ml.;
Wassermann reaction strongly positive;
Gold Sol 5555543110.

Treatment.—Iodides were given for a few days and then penicillin in doses of 1 mega unit daily for 20 days.

Progress.—No Herxheimer reaction was observed, and throughout this period of a month he was a pleasant cooperative patient, reading, playing cards, and joining happily in the activities of a general ward.

Soon after this he mentioned that he was hearing music in his left ear; he found this rather pleasant, but it was clearly an hallucination. In other respects his conduct remained unchanged for 5 days, then suddenly he became completely uncooperative, violent, and disorientated. Within 24 hours he presented the picture of acute mania. He was certified insane and transferred to a mental hospital.

Discussion.—In this case, the unusual features are the patient's degree of insight into his mental condition and the preservation of his intellectual faculties. The onset of unilateral auditory hallucinations was the prelude to acute mania. The question of a delayed Herxheimer reaction was considered but it was thought to be improbable as the time interval was 4 weeks.

Case 4 (Tabes Dorsalis), a farm worker aged 55, was first seen in July, 1956, when he presented with the characteristic clinical findings of tabes dorsalis. His medical record from other hospitals stated that in 1951 a diagnosis of syphilitic myocarditis and gumma of the leg had been made and he had been given a course of injections. In 1955 he was under treatment for ulceration of the legs and asthma. In March, 1956, he was treated for osteo-arthritis and was given 20 mega units of penicillin and physiotherapy. In May, 1956, he was diagnosed elsewhere as suffering from tabetic arthropathy (Charcot's disease) of the left hip.

Examination.—He was admitted to hospital in July, 1956.

Central Nervous System: He had small fixed pupils that did not react to light or convergence.

Reflexes:

Arm: active;
Knee: absent;
Ankle: absent;
Abdominal: noted with difficulty;
Plantar: flexor.

Romberg's test gave slight swaying. There was loss of vibration sense and of deep pain sensation in both legs.

Joints: X rays of both hips showed mixed, destructive, and proliferative arthritis suggesting a neurotrophic arthritis. There were minor osteo-arthritic changes in both knees and ankles.

Chest: This showed signs of slight emphysema and fibrosis at the apex of the left lung.

Urine: Loaded with pus and contained *Esch. coli*, enterococci, and *B. proteus*.

Blood: Definitely sero-positive to all tests for syphilis.

Progress.—He improved to a moderate degree while in hospital but decided to return home after 3 weeks' treatment.

He was re-admitted to hospital on October 23, 1956, when his condition had deteriorated very markedly. He had an irregular, low fever, large bedsores in the sacral area, and had lost weight. The movements of both hips were very markedly reduced, and he had considerable swelling of both knees and both ankles. A cough or other respiratory symptoms were not in evidence, but a few rhonchi and crepitations were heard at the base of both lungs. His urine was heavily loaded with pus and the same organisms (resistant to sulphonamides and antibiotics) were again isolated. A lumbar puncture was not done on account of the large deep ulcer over the sacrum.

An x-ray examination showed a great increase in the disorganization of both hips, with proliferative and destructive arthritis suggesting a neurotrophic lesion. There were similar, but less severe, changes in both knees and both ankles.

After a period of improvement the patient's general condition deteriorated rapidly and he died on November 9, 1956, i.e. 16 days after admission.

Autopsy.—It was found that he suffered from miliary tuberculosis. The hips, knees, and ankles were very disorganized, with necrosis of bone and capsular tissue, but without the microscopic changes of tuberculous arthritis. The original tuberculous lesion at the left apex had become active, with a small area of tuberculous broncho-pneumonia around the fibrosed focus. The brain and spinal cord showed no obvious wasting of the frontal hemispheres or posterior columns of the cord, and no change in the meninges.

Sections of the cerebral hemispheres showed no evidence of either meningovascular or parenchymatous syphilis. There was some slight loss of nerve cells from the frontal cortex but no proliferation of "rod cells". A section of the mid-brain showed no abnormality. Sections from various levels of the spinal cord showed a very slight thinning of myelin in the posterior columns, most obvious in the columns of Goll; this demyelination was, however, slight.

Discussion.—This was a case of tabes dorsalis with arthropathies which progressed rapidly in the last

3 months of life. Urinary infection and a deep ulcer over the sacrum complicated the picture. Radiological examination indicated a neurotrophic lesion of the joints. Three months before death an inactive fibrosed tuberculous lesion at the apex of one lung was observed. His final illness was miliary tuberculosis, the diagnosis being obscured by the co-existing tabes with urinary tract infection and

"bedsore". As tuberculosis, like syphilis, is becoming a rare disease, it is apt to be overlooked.

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