REPORT OF A CASE OF BILATERAL DEGENERATION OF THE GLOBUS PALLIDUS.

By R. Mowbray, M.D., M.R.C.P., D.C.H., Major R.A.M.C.,

AND

H. K. Fidler, B.Sc., M.D., Major R.A.M.C.

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It is felt that the following case and post-mortem report is of such clinical and pathological interest that it is worth while recording it in its present form. Under existing conditions of service we have been unable to go deeply into the literature concerning such conditions.

Case Report.

Lt. R. F—, R.N.V.R., aged 26, was fit and well until a motor accident on November 26, 1942, when he sustained the following injuries:

(1) Multiple abrasions and minor lacerations of the face.
(2) Small linear laceration of the right upper eyelid.
(3) Subconjunctival haemorrhage of the right eye with no evidence of other damage to the eye.
(4) Fracture of the nose with displacement.
(5) Loosened teeth with the right first incisor broken off and driven back into the gum.
(6) Fracture of the right forearm, with rotation of the fragments.

He was taken to a general hospital, and when examined there was not unconscious and gave no history of loss of consciousness. A general anaesthetic was given and the right forearm manipulated and reduced with a satisfactory result. On November 28 he was thought well enough to transfer a short distance by ambulance to the area orthopaedic centre situated at another general hospital. On arrival at this hospital his condition was quite satisfactory. On November 29 his scalp wound began to bleed and was opened up by Lt.-Col. Armstrong under pentothal anaesthesia. Blood clot was evacuated from the wound, but no definite bleeding vessel was found; therefore the wound was sutured and dressed. While in the theatre for this operation an examination of the nose was made by the E.N.T. surgeon, who reported as follows:

"There is a fracture of both nasal bones and the nose may be freely moved on the face. Position good and both airways are clear at the moment; there is therefore no treatment so far as the nose is concerned; splints are not required. Ears are normal."

The patient was returned to the ward, where he became cyanosed and his pulse rate increased to 140 per minute. He was given oxygen and carbon dioxide, and his jaw pulled well forward. He soon improved, but later in the day his temperature rose to 101·4° F. and he had a rigor.

Lt.-Cmdr. Ayoub examined his eyes on December 1 and reported that their condition was satisfactory. On December 2, under pentothal anaesthesia, Lt.-Col. Armstrong replastered the right forearm, after which the patient's condition appeared to be satisfactory and the wounds were healing in a normal manner until December 7, when it was noticed that he was very restless in bed and appeared to be unable to lie still.

When examined on the morning of December 9 he was found to have definite choreiform movements of both arms and legs, face and body. The tongue was typical of chorea, and a worm-like grip was present in the left hand. Speech was

affected in so much that it was explosive and not clear, his replies to questions were short, and because of his speech he was unable to carry out a sustained conversation, but mentally he appeared normal at this time. The movements were observed to stop during sleep. An examination of the nervous system showed nothing further abnormal.

The wounds on his lip and the scalp and eye wounds had become septic, and on examination of his throat there were found a few patches of yellow exudate on both tonsils.

A throat swab taken on December 9 showed *Staphylococcus aureus* and haemolytic streptococci, but no diphtheria bacilli or Vincent's organisms. The leucocyte count was 9,500 per c.mm., with a differential count of neutrophils 67 per cent., lymphocytes 29 per cent. and monocytes 4 per cent. The cerebrospinal fluid was also examined and found to be clear, with less than two cells per c.mm., protein 40 mgm. per cent., globulin no excess and sugar normal. Direct smear and culture were negative.

On December 11 his condition was much the same as before, but next day it was thought that he had improved a little and he could control his movements to a certain extent voluntarily. His urine examined on this day was normal.

However, when examined on December 13 his condition was definitely worse; he had generalized, purposeless, jerky, ill-sustained movements, which were most marked in the left arm, which was slightly weak as well. All deep reflexes were exaggerated, the abdominals were present and the plantars were flexor. There were no sensory changes, and no incontinence of urine or faeces. The only other abnormality found was diminished movement of the soft palate. Surg.-Lt.-Cmdr. Davies, R.N.V.R., was asked to see the patient, and he agreed with the diagnosis of chorea, but could not suggest any cause for it.

Next day the patient complained of difficulty with swallowing, and on examining the palate it was found that the movements were spasmodic. His choreiform movements seemed a little better.

On December 15 the ulceration on his lips was definitely worse; he began to complain of cramp-like pain in his legs, and there was slight weakness in his left peroneal muscles. The condition of his palate was the same, the lip ulceration showed a slight tendency towards healing, but the scalp wound was still septic and his left tonsil was covered with a dirty greenish slough, the rest of his throat being mildly inflamed. A specimen of urine tested was normal. A swab from the lip grew diphtheroids and scanty haemolytic *Staphylococcus aureus*, but the throat swab grew morphological K.L.B. The white cell count was 10,800 per c.mm., with 76 per cent. neutrophils, 20 per cent. lymphocytes and 4 per cent. monocytes. No abnormal cells were seen. A further throat swab taken on December 23 again grew morphological K.L.B.

By December 26 the condition of the throat was much improved and the slough had gone, and his scalp and lip wounds were improving. A smear from the scalp wound grew haemolytic *Staphylococcus aureus*, but no diphtheria bacilli. His chorea was, however, getting worse, and on the night of January 1 he was very restless and attempted to get out of bed, talking nonsense all the time; he was also incontinent of urine. On January 3 Lt.-Cmdr. Ayoub examined his eyes, and reported that the right pupil was larger than the left, and that there was slight fluffiness of the right disc margin. He thought the cause was a mid-brain lesion. Major Lloyd, the area psychiatrist, also examined the patient and agreed with the diagnosis of chorea, which was associated with marked hallucinations, but because of the patient's condition he was unable to make any detailed psychiatric examination.

On January 4 Brigadier James saw the patient, but was unable to shed any further light on the case. A white cell count done on this day showed 11,500 cells per c.mm., with 75 per cent. neutrophils, 3 per cent. basophils, 18 per cent. lymphocytes and 4 per cent. monocytes. All the cells appeared normal.

Lumbar puncture was performed on January 5, but the C.S.F. was mixed with blood due to trauma. A throat swab grew haemolytic *Staphylococcus aureus*, but no diphtheria organisms.

On January 6 it was reported that he had not slept at all well during the night, going to sleep for only short periods at a time, during which periods his choreiform movements stopped. He had been definitely more noisy and had talked a lot of

nonsense, sometimes rambling on about his accident, or pretending to be driving a motor car or piloting an aeroplane. He was showing definite signs of exhaustion, and his mental state was getting worse; it was getting very difficult to get him to answer simple questions, and feeding was becoming a problem. On examination there were still the same typical choreiform movements of the face, arms, legs and body, the tongue movements were typical, as also was his grip, but the power in his limbs was definitely decreasing. All deep reflexes were present, and no gross lesion of the cranial nerves could be found. The abdominal reflexes were absent. His legs were wasted, and his feet had taken up a position of extension and inversion; there was no true paresis or hypertonia of the ankles, but movements were weak. Plantars were flexor. He had now become incontinent of both urine and faeces.

On January 10 he seemed a little quieter and more sensible, and the chorea was not quite so violent. The heart sounds were tic-tac in character, with a short sharp aortic second sound. Next day he was worse again, and at times he was confused; he had no idea about the date or the time of day, and had only a vague idea of how long he had been in hospital, but later on he became quite rational. At times he would become rather violent and try to hit people, but this seemed to be due to an ungovernable emotion of dislike for certain people; later on he would remember all about it and be sorry for what he had done. His movements were worse, but there was no change in the physical signs in the nervous system.

Lt.-Cmdr. Ayoub again examined his eyes and reported that there was no choking of the discs, but that they were not as distinct as possible owing to the mucoid corneal covering. Another lumbar puncture was carried out, the fluid being below normal pressure and the Queckenstedt normal. There were 4 lymphocytes and 18 red blood cells per c.mm., protein 70 mgm., globulin slight excess and sugar normal. Culture and smear were negative. The increase in protein and globulin was probably due to the previous lumbar puncture, when pure blood had been obtained.

By January 14 his movements were worse; he did not recognize those who were looking after him, and he appeared unable to speak coherently. From this date onwards he became steadily worse, becoming completely disorientated and having great difficulty in swallowing, not apparently due, however, to any complication associated with his diphtheria infection; his choreiform movements remained much the same in degree.

On January 20 he developed a cough without any very definite gross physical signs in the lungs, and he died suddenly on January 22.

A lumbar puncture had been performed on January 16, when the cerebrospinal fluid was found to be under normal pressure and showed 8 lymphocytes per c.mm., with 40 mgm. of protein and no excess of globulin, the sugar being within normal limits. Blood urea done on the same day was 96 mgm. per 100 c.c., with a normal urine.

During the whole course of the illness the temperature did not rise above 99° F. except after the operation on November 29 and just before his death.

Radiographs of the skull and right forearm had been taken on the day following his accident and showed no fracture of the skull, the radius and ulna being fractured at the junction of the middle and upper thirds.

One attempt was made to find out something about this officer's family history, but apart from hearsay evidence that there was no family history of chorea or fits or mental defect we were able to get nothing definite. He himself was regarded as somewhat of a clever eccentric before his accident.

The treatment of this case was from the outset somewhat empirical, as a definite diagnosis beyond chorea could not be made. The surgical treatment consisted of the usual routine measures for such injuries, and apart from the occurrence of sepsis, which was dealt with, seemed satisfactory and his wounds were healed at the time of his death.

From the point of view of medical treatment the exhibition of aspirin or sodium salicylate had little effect on the condition. Luminal was of no use at all; large doses of medinal did seem to help a little, but paraldehyde was the best sedative tried. Morphia had little beneficial effect. He was given 80,000 units of anti-diphtheritic serum intramuscularly on December 23, and his throat seemed to improve in a normal manner following this injection. 23 gm. of sulphanilamide

was also given in the hope of helping the wound sepsis, but the whole course of the disease was one of progress downwards to death.

Autopsy (10 hours post mortem).

The body was wasted, but otherwise well developed. The fracture of the right arm had united and the injuries of the face had healed completely.

The nasopharynx contained a little muco-purulent exudate. The tonsils were large and cryptic, but the mucous membrane appeared normal. The right lung weighed 570 gm. and the left lung 455 gm. Both showed a moderate degree of congestion, oedema and hypostatic lobular pneumonic consolidation. The heart weighed 255 gm., and apart from a moderate degree of right-sided dilatation appeared normal. The liver weighed 2,005 gm. It was enlarged, dark reddish-brown in colour and the cut surfaces were bloody, but otherwise normal in appearance. The other abdominal viscera appeared normal.

There was no evidence of any injury to the skull. There was an apparent increase in cerebrospinal fluid, and the meningeal vessels were congested. The external surfaces of the brain appeared normal. The cut surfaces showed a moderate degree of generalized vascular congestion, and an otherwise normal appearance everywhere, except for the globus pallidus on each side, where very striking macroscopic lesions were found (Fig. 1). These structures were soft, buff-coloured, porous and collapsed. On each side the entire structure was involved and was clearly demarcated from the surrounding tissues, which appeared free from disease. The latter, which include the caudate nucleus, putamen, optic thalamus and internal and external capsules, were of normal colour and consistency. The white bands of the medullary laminae which divide the globus pallidus in two appeared broader than normal—an appearance which was probably only relative, due to collapse of both zones of the globus pallidus. The medulla oblongata and spinal cord appeared normal.

Microscopic Examination.

There was some fragmentation of muscle fibres in the heart. The kidneys showed vascular congestion, and rather marked cloudy swelling of cells lining the convoluted tubules. In the liver there was a moderate degree of generalized congestion in the sinusoids and a little fatty metamorphosis in the parenchymatous cells, particularly in the central zones of liver lobules.

Sections from the central nervous system included the globus pallidus, putamen, optic thalamus, internal capsule, caudate nucleus, substantia nigra, red nucleus and subthalamic regions, motor cortex, floor of the fourth ventricle and medulla oblongata. They were stained with Ehrlich's haematoxylin and eosin and Pal's modification of Weigert's method for myelin sheaths. Myelin degeneration was almost complete in the globus pallidus on each side. The normal structure was replaced by a loose reticulum, in which there were many scattered compound granular macrophage cells. These were somewhat more abundant around the blood vessels, where they formed rather ill-defined perivascular cuffs. The only other type of cell seen was a small cell with a dark staining nucleus and scanty basophilic cytoplasm. These were not numerous, and were scattered diffusely throughout the lesion. Only a few nerve cells were present. They were at the edge of the lesion, and showed varying degrees of degeneration from mere swelling of the cell to chromatolysis. No red blood cells or pigment was found to suggest that any previous haemorrhage had occurred. The edge of the lesion was well demarcated, and the surrounding brain tissue appeared unaffected except for a slight apparent increase in small round-cells in the zone of demarcation.

Weigert-Pal stained sections failed to reveal any lesion other than that described in the globus pallidus.

Service conditions made it impossible to carry out any further histological investigation of the tracts to and from the globus pallidus.

Comment.

This case is of unusual interest, because it showed a degenerative lesion which was apparently confined purely to the globus pallidus on each side and the symptoms

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were those of a progressive type of chorea. No athetoid movements were observed. No liver lesions suggestive of those found in Kinnier Wilson's disease were seen.

From the post-mortem findings there is nothing to suggest that fat embolism, haemorrhage or any other vascular disease had a place in the aetiology. Microscopy indicated that the lesion was one of pure degeneration.

It would appear to us that the most likely aetiological factors are either trauma at the time of his accident or the anoxaemia which followed his first pentothal anaesthetic. In either case the addition of some constitutional defect of the nervous tissue concerned may have been an additional factor.

Summary.

(1) A case is described in which bilateral degeneration of the globus pallidus is accompanied by the clinical picture of severe chorea.

(2) The possible aetiology is shortly commented on.

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