and forearms. The wasting and weakness extend very gradually to the upper segments of the limbs, and the muscles of the trunk and face are rarely affected even after thirty or forty years. Bulbar symptoms are unknown. It is important to notice that the reaction of degeneration can be found in some of the muscles in practically every case, while there may or may not be cramps, pains, coldness and lividity of the skin, fibrillary tremors, or slight sensory changes.

My patient, F. J., aged 33, married, with one child, is a strong and otherwise healthy man, in whom the condition first appeared when he was 19 years old. He was then an active footballer and member of a rowing club. He even now plays golf and finds no fatigue from a five-mile walk, though the disease has progressed steadily. The feet were first affected and then the hands and forearms followed. There is no history of any previous illness, and he has rarely required medical treatment. The great age which many of his relatives attained, even when affected like himself, is striking. His heart, kidneys, and lungs are normal, the expansion of his chest is 3 inches. The muscles of the trunk, face, eyes, throat, and tongue are unaffected. He can whistle and pout the lips. The action of the bladder and bowels is normal. There is extreme wasting of the muscles of the feet and lower part of the legs, and intrinsic muscles of the hands, and the forearms, and slightly of the thighs. There is no movement at all in the toes or at the ankles; neither abduction nor adduction or flexion of the feet. The great toe and the others are flexed and quite motionless; the arch of the foot is preserved, and his gait in walking is surprisingly good, though slightly high-stepping. He flexes and extends the leg on the thigh and the thigh on the hip freely, but in getting up from the ground he likes a little assistance, such as putting his hand on his thigh, but he can bend down to touch the ground and rise up again easily.

Though the small muscles of the hands are so wasted, the grip is strong, and he has no difficulty in buttoning his clothes or in writing. Supination and pronation have escaped, but extension at the wrist is weak, and the hands when at rest take the claw form. There is little or no fibrillation or muscular irritability on percussion. There is no ataxy. He stands easily with his eyes shut, and the pupils react to light. The knee-jerks are present, but weak; the Achilles tendon-jerk is present, but there is no plantar reflex or clonus. He has no pain or cramps, and the sensation to light touch or pin-prick appears normal over hands, feet, and trunk. There is no reaction to either current in feet, calves, or hands, but some of the extensors at the wrists show reaction of degeneration—that is, no faradic reaction but good galvanic, with the positive pole equal to or greater than the negative.

In this patient, then, we get wasting of the distal segments of the limbs with loss of power, beginning in early adult life and steadily progressing. The reflexes are diminished and reaction of degeneration is present. Finally, it appears from his statements that eleven males and ten females of his family are similarly affected. In this family the first case recorded is that of the patient's great-grandfather, and out of fifty-four of his descendants twenty-one are already affected and others are not yet old enough. The disease seems to pass rather more readily by the male line than the female, and it has little effect on the duration of life. In a group of five brothers and sisters affected, two are alive aged 77 and 79, and three died at ages of 62, 82, and 86.

One would like to know whether the affection did actually begin with the great-grandfather. It cannot have existed many generations back at the present rate of increase, for the district would be full of cases; whereas the disease is always a rare one.

The type is clearly distinct from the myopathies. Thus—

(1) Pseudo-hypertrophic paralysis has neither the distal distribution of wasting nor the reaction of degeneration, and it generally shows some muscles enlarged.

(2) The rare cases of distal myopathy such as the one recorded by Gowers¹ appear to have the facial muscles involved and no reaction of degeneration.

(3) The Werdnig-Hoffmann type is really myelopathic, but the wasting begins in the proximal segments during the second and third years of life, and death usually follows in three or four years. Reaction of degeneration is found in these cases, and several may occur in one family.

(4) In amyotonia the trouble begins at birth or soon after. There is no definite wasting, but paresis of the distal segments which may improve if the patient survives. I think there is no affection of other members of the family here, but reaction of degeneration may be present.

My group of symptoms, then, differs widely from all these. The pathology of this peroneal type does not seem to have been made much clearer of late. The sclerosis in the columns of Goll and Burdach, and the lesions in the

anterior horn cells and in the peripheral nerves, appear to vary in intensity in different cases, but we are completely ignorant of the cause which produces them. Whether it be "a developmental weakness, an inherited susceptibility to some toxin," or a true infection, spirochaetal or otherwise, remains to be proved.

REFERENCE.

¹ *British Medical Journal*, 1902, ii, p. 89.

PARALYSIS OF ALL FOUR LIMBS CURED BY REMOVAL OF A SPINAL TUMOUR.

BY

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A WOMAN, aged 51, was admitted to hospital in October with complete paralysis of the right arm and leg and partial paralysis of the left arm and leg. She also complained of intense headache and gnawing and shooting pains in both arms; these were so severe that hypodermic morphine was needed to give her any sleep.

The history given was that fourteen months previously she had first noticed numbness in the right hand, but this did not bother her until in February she tripped over something and had a bad fall, which shook her considerably. After that she began to lose the use of her right arm. By May the arm was so weak that she had to give up her work, and she was beginning to drag the right foot. There was also then some aching pain in the shoulders and right arm. Early in July the right hand had become useless, and the trouble in the right leg had advanced so much that she could not walk, also the left leg was affected. By the end of August the right arm and leg were paralysed, and she could only move the left leg slightly, and there was weakness in the left shoulder.

On admission the right arm and leg were motionless; she could draw up the left leg slightly as she lay in bed. She could not abduct the left shoulder, but could adduct and rotate it, and flex the forearm. The grasp of the hand was very weak, and she could not feed herself with it. Both the trapezii were wasted, but not the arm muscles. Reflexes were increased.

She complained of intense headache and agonizing shooting and aching pains in both shoulders, and of "pins and needles" all over the body. The pain had been getting worse and worse for the past six weeks. There was anaesthesia of the right hand and forearm, and diminished sensation in the same area on the left arm. Sensation over the legs and body was less than normal, and distinction between heat and cold was not accurate. There was no incontinence, but it was difficult to get the bowels to act.

The neck could be rotated and flexed, and an x-ray photograph showed no disease of the bones. On lumbar puncture the cerebro-spinal fluid was not under pressure. It contained globulin in large excess, colloidal gold 55544,33221, Fehling reduced normally, no increase of cells. The Wassermann reaction was negative. The cerebro-spinal fluid of the cisterno puncture contained only a very faint trace of globulin, colloidal gold 01110,00000, no cells. It would be interesting to know if this striking difference in the cerebro-spinal fluid below and above the tumour occurs in other cases.

The diagnosis made was a lesion of the spinal cord about the level of the second cervical vertebra, and, on account of the intense pain, probably a tumour in the meninges pressing on the back of the cord. Mr. Beresford was asked to do a cisterno puncture and insert some lipiodol. X-ray photographs after this showed some lipiodol just above the level of the first cervical, and opposite the level of the first and second cervical vertebrae.

After the operation the movements of the left arm soon began to improve, next those of the left leg. It was nearly a month before the right leg showed signs of recovery, and the right arm later still. Massage and

faradism were used to hasten the return of movement. After ten weeks the patient could walk with a little assistance, spasticity not having quite disappeared. Movements of the left arm and hand were perfect, and she could lift a glass to her mouth with the right hand.

The operation described by Mr. Beresford:

On November 8th, 1927, under intratracheal anaesthesia (Dr. Eccles), the laminae of the second, third, fourth, and fifth cervical vertebrae were removed. The dura was not pulsating and felt hard in the second, third, and fourth cervical region. On opening the dura a tumour was seen in the postero-lateral aspect pressing into the right side of the cord. The tumour, which was attached by a pedicle to the dura, was enucleated without difficulty. Its appearance was rather like a large raspberry, fleshy to feel, and not very vascular. It measured 2.75 by 1.5 by 1 centimetres.

The dura was closed with continuous catgut, and, after closing the wound, the head, neck, and shoulders were supported in a plaster cast. The patient left the theatre in good condition, and got immediate relief from her intense headache. Sections of the tumour show it to be an endothelioma. This tumour is said to be non-malignant, and therefore the prognosis in this case is good.

COMPLETE TRANSVERSE RUPTURE OF THE JEJUNUM WITHOUT EXTERNAL WOUND.

BY

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COMPLETE transverse rupture of the intestine is rare in the absence of external wounds. Moreover, recovery from such a condition is exceptional. The following details of a case of such rupture of the jejunum seem worthy of record, particularly in view of the satisfactory issue.

A man, aged 46, was brought to the Nelson Hospital on July 20th, 1927, suffering from severe abdominal injury. The history given was that while riding a motor bicycle he had lost control and crashed into a wall, his abdomen striking a projecting corner.

On arrival at hospital the patient was in great pain and suffering from severe shock. He was in a cold sweat, and complained of intense pain in the abdomen, which at times caused him to writhe in agony. The pulse was 64, and the temperature 97.5° F. Breathing was laboured with grunting expiration and almost entirely thoracic; vomiting did not occur. The pupils were equal and contracted, and reacted to light and accommodation. The whole of the anterior abdominal wall was intensely rigid and motionless, while the only external sign of injury was a slight discoloration or contusion just below the ribs in the left hypochondrium. On palpation the abdomen was extremely tender everywhere, and a point of maximum tenderness could be elicited in the region of the tip of the spleen. There was definite dullness in the flanks, especially on the left side, but the patient was so ill that shifting dullness was not investigated. Liver dullness was present. The bladder was not distended; a catheter was passed and normal urine withdrawn. A diagnosis of grave abdominal injury with rupture of a viscus, possibly the spleen, was made, and the patient was treated for shock, 1/2 c.cm. of pituitrin being given together with a rectal saline injection, morphine, and atropine.

Two hours later Dr. Martin Randall opened the abdomen; a large left paramedian incision was used, with the centre opposite the umbilicus. No gas escaped on opening the peritoneum, and no fluid or odour suggestive of rupture of a hollow viscus was perceptible. There was a large quantity of almost pure blood in the peritoneal cavity. Examination of the liver, spleen, stomach, and pancreas revealed no injury, but while the last-named was being investigated a loose torn end of bowel came into view. This proved to be the jejunum, about four inches beyond the duodeno-jejunal flexure. The other end was found without difficulty, the tear separating the two pieces of bowel extending a full inch into the mesentery. The edges of the severed bowel were somewhat irregular, but it was practically a clean transverse tear. The removal of about half an inch on each side sufficed to form a clean-cut edge for suture, and an end-to-end anastomosis was performed. The peritoneum having been sponged quickly, the incision was closed, but a drainage tube was placed in the left loin and another in the pelvis, since some fluid was still present in each of these situations. After the operation the temperature was 97.4° F., and the pulse 88. The patient was given 1/4 grain of morphine at midnight, and saline by the rectum and subcutaneously.

At 6 a.m. the next day he was given a further 1/6 grain of morphine, and throughout the day continuous rectal saline. The temperature rose to 99° F., and the pulse to 116. Throughout the day the patient was given an ounce of water at a time, and during the day 1 drachm of brandy. In the evening he was given

an enema, and he passed a considerable quantity of flatus, with a slight action of the bowels. At night he was given 1/6 grain of morphine and at 4 a.m. on the next day a further 1/12 grain. During the day he seemed to have improved, the temperature being 98.6° F., and the pulse 104; vomiting occurred four times, and in the evening the stomach was washed out with sodium bicarbonate, which arrested this. From this time onwards the patient continued to progress satisfactorily; distension was relieved by turpentine enemata. The temperature remained normal or slightly subnormal, and the pulse rate had fallen to 70 by July 24th. The patient suffered from some distressing hiccup, which however, responded to a simple mixture of bismuth, and he continued to progress until he was discharged from hospital on August 27th, 1927, in good health.

A series of cases recorded by Massie¹ at Guy's Hospital between the years 1899 and 1922 shows that rupture of the intestine occurred in 34 instances. Of these, 26 were situated in the small intestine, and the jejunum was more commonly injured than the ileum; of the 14 jejunal cases, 7 occurred within eighteen inches of the duodeno-jejunal flexure. It was also shown that the part most susceptible to injury is that part which crosses the vertebral column, and that the second and third loops of the gut are most frequently involved. Most of these lesions were partial, and nearly all cases where the duodenum was involved were associated with other severe injuries in the upper abdominal wall. This writer also points out that whereas cases of ruptured stomach and duodenum are accompanied by other injuries, generally intra-abdominal, cases of rupture of the small intestine are usually single, and are not associated with any other injury. This fact is borne out by the present case.

The main symptoms and signs are pain, shock, and rigidity. It has been said that impairment of liver dullness is to be expected, but in the series collected by Massie this occurred in only 3 out of 34 cases; in the present case there was no impairment. Of 31 cases reported of complete ruptured small intestine the mortality was found to be 78 per cent., and in 9 cases in which the rupture was complete the mortality was 87 per cent. The high mortality in cases of complete rupture is said to be due to the increased amount of shock as compared with cases of partial rupture.

Another series of cases, numbering 22, and occurring at the London Hospital between the years 1913 and 1922, is recorded by Huddy.² This writer emphasizes the importance of early operation, the significance of pain as an indication for operation, and the fallibility of relying on the pulse rate—that is, not operating because it is slow. Again, such signs as loss of liver dullness, surgical emphysema, and distension are not of great clinical value; the latter is generally a late result, and is often found with other injuries, such as damaged kidneys or fractured pelvis; emphasis is also laid on the fact that the mortality increases with the length of the history, and that operation should not be delayed if there is no improvement after three hours. In the present case diagnosis of grave internal injury was easy on account of the extreme rigidity, great shock, and the definite evidence of effusion into the peritoneum.

The decision to operate depends on the diagnosis of grave internal damage and not on a diagnosis of any exact form of injury, and the earliest moment at which the condition of the patient is such as to make it possible should be the time chosen for operation.

The points that seem to be of special interest in this case are the following. On opening the abdomen no gas was detected, nor was there any fluid or smell suggestive of a rupture of a hollow viscus. The only fluid that escaped was blood, possibly diluted by peritoneal serous exudate. Nothing resembling food could be found in the effusion, and the inevitable presumption is that the intestine was empty when injured; the shock was followed by a pyloric spasm, which persisted until the operation was performed, because the stomach was not empty, the patient having had tea a short time before the accident occurred. The success of the operation was probably largely due to its being undertaken early.

I am indebted to Dr. Randall for assistance in describing this case and for permission to publish it.

REFERENCES.

¹ Massie: *Lancet*, 1923, ii, 640. ² Huddy: *Clinical Journal*, February 6th, 1924, p. 66.