UPPER-MOTOR-NEURONE PARAPLEGIA

THREE ILLUSTRATIVE CASE REPORTS

Three patients presenting with upper-motor-neurone paraplegia were encountered in the medical wards of Somerset Hospital during 1961. Each case had a different aetiology, and each illustrated some unusual aspects of this condition. All three were curable. The case reports are presented below as a trio by the three Resident Medical Officers who each looked after one of these patients.

1. PERNICIOUS ANAEMIA WITH 'ACUTE' COMBINED DEGENERATION OF THE CORD

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This case report is presented to indicate the rapidity with which subacute combined degeneration of the cord can develop and progress in pernicious anaemia and produce a total paraplegia.

CASE REPORT

A 62-year-old male was admitted to Somerset Hospital on 15 December 1961. The referral note by the general practitioner contained the significant sentence: 'I am unable to get an adequate history'.

The patient was incapable of giving a good or accurate history. He was fully conscious, but was a very slow thinker, and was apathetic, failed to concentrate during conversation, and was sometimes disorientated and confused. Some of his remarks were irrelevant and inconsequential.

His chief complaints were progressive weakness of the legs, which developed slowly 12 months before, accompanied by swelling of the feet; loss of weight; a 9-month history of an uncomfortable sensation in the epigastrium ('soos 'n wind') associated with eructation of air and of fluid which caused a burning sensation at the back of his throat; and some difficulty and hesitation in urination for 1 month.

The significant abnormalities were confined to the central nervous system, apart from pitting oedema of the feet and ankles and a persistent temperature of 100°F. Power was decreased in the legs and there was some muscle wasting. All tendon jerks were present. The plantar reflexes were equivocal and there was bilateral ankle-clonus. Sensation was difficult to assess owing to lack of sustained attention, but the heel-knee test indicated poor coordination.

Special investigations. Haemoglobin level 7·8 G. per 100 ml., PCV 28%, white-cell count 8,000 per c.mm. The red cells showed marked anisocytosis with approximately 30% macrocytes, and there was some poikilocytosis. The white cells were normal. Apart from carcinoma of the stomach and other gastro-intestinal disorders, pernicious anaemia appeared the likeliest diagnosis, and the following special investigations were performed: cerebrospinal fluid — normal; chest X-ray — normal; occult blood test (benzidine test) on stool — negative; barium meal — normal; bone-marrow examination — the marrow was cellular with considerable activity of both red and white cell series, the red cells showing megaloblastic changes; total serum bilirubin — 0·5 mg. per 100 ml.; reticulocytes — 1·6%; investigations performed by the gastro-intestinal service at Groote Schuur Hospital revealed achlorhydria and achylia; no neoplastic cells were seen on gastric cytology.

Subsequent Course

Owing to the intervention of the public holidays in December and January, these investigations took 2 weeks to complete and during the first week of January 1962 (while awaiting the performance of the Schilling test) the patient became more ill and confused and generally weaker.

By 5 January his legs had become extremely weak; this was accompanied by retention of urine which required a self-retaining catheter. The knee jerks could no longer be elicited, while the plantar reflexes remained equivocal.

Intramuscular vitamin-B_{12} therapy was instituted on 5 January 1962, with an initial dose of 100 micrograms daily until 16 January, when the dose was increased to 1,000 micrograms daily for 14 days. The dose was then reduced to 100 micrograms daily until 5 April 1962, since when he has been receiving 100 micrograms weekly. His course after the start of vitamin-B_{12} therapy was as follows:

10 January: He looked very ill with a temperature of 104°F. (owing to infection of an ulcer which had developed on his buttocks), and both his legs were totally paralysed. The infection responded to therapy with chloramphenicol and novobiocin. The reticulocyte count (after 5 injections of vitamin-B_{12}) had risen to 8·6%.

19 January: Reticulocyte count 3·9%.

22 January: Apyrexial.

22 February: Mentally much brighter. The sense of position in his legs was grossly defective although power was returning to them. The haemoglobin level had risen to 9 G. per 100 ml.

1 March: The self-retaining catheter was no longer required. The haemoglobin level was 11·5 G. per 100 ml.

March - June: Power steadily increased in his legs and he gradually started walking in a walking-machine; at the beginning of June he could walk slowly by himself, but with an ataxic gait. Progressive improvement was continuing. The power was almost normal, but the sense of position was still defective. The plantar reflexes remained extensor. Knee jerks were present and ankle jerks were increased. The haemoglobin level had risen to 12 G. per 100 ml. Mentally he became much brighter, and cooperative and talkative. It was very obvious, by comparison with his state on admission, that there had been considerable intellectual deterioration at the time of his admission, which had greatly recovered at this stage.

14 June: He was discharged, walking, and instructed to attend for weekly injections of vitamin B_{12} as an outpatient.

DISCUSSION

The diagnosis of pernicious anaemia with subacute combined degeneration of the cord was confirmed by the haematological findings, the megaloblastic bone marrow, the achlorhydria and achylia, the reticulocytosis after vitamin-B_{12} therapy, the response of the anaemia to this therapy, and the presence of abnormal neurological signs most suggestive of subacute combined degeneration of the cord.

Prominent gastro-intestinal symptoms and loss of weight are well-known features in pernicious anaemia. The patient also had considerable mental and intellectual changes, which have been described in pernicious anaemia by various observers,[1,2] and have been re-popularized in recent years under the name of 'megaloblastic madness'. These changes responded to vitamin-B_{12} therapy.

The most noteworthy feature of this case is the rapidity with which the neurological involvement deteriorated *in the third week* after admission, before vitamin-B_{12} therapy had been instituted, leading to a total paraplegia with retention of urine. It was alarming, since therapy had been withheld pending the completion of the special investigations. Price[3] mentioned that the onset of subacute combined degeneration of the cord, though usually insidious, may be very rapid, and that deterioration may also be rapid. One wonders whether the acute infection of the buttocks, which was occurring at approximately the same time, played a part in *the rapid transformation of his legs from a state of weakness to total paraplegia in the*

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space of one week. The patient walked out of hospital 6 months after admission, with neurological abnormalities still present, but becoming progressively less.

SUMMARY

1. A case of pernicious anaemia is presented.
2. To illustrate the rapidity with which the neurological involvement progressed to total paralysis of the legs it is presented as 'acute combined degeneration of the cord'.
3. The abnormal haematological, neurological, and mental features responded to vitamin-B_{12} therapy.

I wish to thank Dr. G. J. Joubert, Medical Superintendent of Somerset Hospital, for permission to publish, Drs. M. Horwitz and J. B. King for their guidance, and Miss Pat Riley for assistance in the haematological investigations.

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