A CASE OF ACUTE INTERMITTENT PORPHYRIA*

ROGER CHAPADOS, M.D., AND BERNARD PAIEMENT, M.D.†

THE HAZARDS associated with anaesthesia in porphyria have been fully discussed in recent publications.^{1,2,3} A 36-year-old painter was admitted to our hospital for treatment of abdominal pain and investigation of a weight loss of 30 pounds in one and a half years.

HISTORY

The patient's past history disclosed hospitalization at 16 years of age for pulmonary tuberculosis; he had left the hospital without having completed the treatment prescribed. He seems to have been in good health until one and a half years prior to this admission.

During the past one and a half years he had been treated twice for abdominal pains of three to five days' duration with nausea, vomiting, constipation, marked asthenia, and headaches. These crises appeared when he was working (painting). Between attacks he felt fine and had a good appetite. The first attack was treated at home. For the second, he was hospitalized elsewhere for 17 days, and was discharged without an exact diagnosis. When he was admitted to our hospital, he had had the aforementioned symptoms for five days.

The physical examination showed a very thin man, dehydrated and of greyish colour. There were micro-adenopathies at the axillary, cervical, and inguinal regions. The abdomen was soft and no palpable masses could be found. The remainder of the physical examination was negative.

Laboratory examination showed Hb. 14.7 gm., Hcrit. 45.3 per cent; white blood cells: 14,300; N. 62 per cent; L. 28 per cent; M. 7 per cent; E. 3 per cent. Chest X-ray showed sequelae of old tuberculosis. Barium meal and barium enema were normal.

Intravenous pyelography showed a normal left urinary tree, but on the right side there was a small dilatation of renal calyces, and the suggested shadow of a tumour in the right para-vertebral region "which might represent a lymphatic tissue tumour."

A diagnosis of chronic lead poisoning was postulated because of the symptoms (pain, constipation, headache, nausea, and vomiting) and the patient's occupation; but all the examinations to confirm this diagnosis were negative except for an elevation of the uroporphyrin in the urine.

In order to outline the para-vertebral shadow seen on the intravenous pyelography, a cavography was done, which showed a mass, probably of lymph node origin, compressing the inferior vena cava on the right side. A lymphoma combined with chronic lead poisoning was suggested. An inguinal lymph node biopsy showed nonspecific adenitis.

*Presented at the Abbott Meeting, McGill University, March, 1965.

†Department of Anaesthesia, Maisonneuve Hospital, Montreal, Quebec.

OPERATION

Three weeks after admission, the patient underwent a laparotomy to eliminate a lymphoma. (See Fig. 1.) Premedication was demerol 75 mg., atropine 0.4 mg. On arrival in the operating room, the patient was very depressed. Blood pressure was 110/70, pulse rate was 80 per minute.

Induction was with thiopentone 125 mg., succinylcholine 40 mg. plus decamethonium 3 mg. The patient was ventilated with oxygen and intubated with a portex no. 9, cuffed tube, whereupon the blood pressure dropped to 70/50. Pulse was still 80.

The blood pressure did not rise after vasoxyl 4 mg. was administered twice intravenously, but only after solucortef 200 mg. and dextran 6 per cent 500 c.c. intravenously. The anaesthesia was continued with nitrous oxide, oxygen, and cyclopropane. The blood pressure stayed at 100/70 with a pulse of 80.

The laparotomy was negative. Lymph node biopsies were done, and the pathological report showed non-specific lymphadenitis. The postoperative course in the recovery room was uneventful.

POSTOPERATIVE COURSE

The patient did relatively well during the first week, except for a feeling of weakness. Treatment with B.A.L. was started for lead poisoning. On the sixth postoperative day he developed a paralysis of both arms, and markedly diminished tendon reflexes. Three days later he suddenly went into cardiovascular collapse (B.P. 0), that only responded to noradrenaline and corticosteroids (B.P. to 120).

Electrolytes that day were normal: Na⁺ 137.7, K⁺ 4.8, Cl⁺ 94.7. The white blood count was 41,600, the temperature 100°, and the pulse 110. Temporary diagnosis was acute adrenal insufficiency, secondary to tuberculosis.

The paralysis spread, and on the eleventh postoperative day the patient was quadriplegic with paresis of the intercostal muscles but no sensory involvement. An examination for urinary porphobilinogen was now positive (Watson test +). Diagnosis was now *acute intermittent porphyria*.

DISCUSSION

This patient, in our opinion, presents two points of interest:

1. A problem of diagnosis. The first diagnosis which came to our mind after the operation was one of acute adrenal insufficiency, and it was not until the neurological signs appeared that the diagnosis of acute porphyria was considered. Dr. Robert Kark, in Medical Clinics of North America of January 1955, reports manifestations of pseudo-Addisonism in case of intermittent acute porphyria,⁴ but we could not find any mention of symptoms of acute cortical insufficiency in these cases.

2. The evolution of this case. There was a spectacular regression of the neurological symptoms after treatment with corticosteroids (prednisone 5 mg. q.i.d. per os.) and chelating agents (B.A.L.) for five days. The patient was able to go home

HÔPITAL MAISONNEUVE

[illegible]

FIGURE 1

for two days, four weeks after operation. Two days after his return to the hospital he presented a massive haematemesis and fell into shock. Three hours later he was dead, because of the impossibility of obtaining blood of his group from a depleted Red Cross Blood Bank, to treat the shock. At autopsy a giant gastric ulcer was found, probably a complication of the corticotherapy.

RÉSUMÉ

Ce travail présente un cas clinique de porphyrie aigue intermittente—qui s'est manifesté neuf jours après une anesthésie générale, par l'installation d'une quadriplégie. Les signes neurologiques ont évolué favorablement (corticothérapie et B.A.L.). Malheureusement ce malade est décédé d'une complication de la corticothérapie (hémorragie digestive massive—provenant d'un ulcère gastrique géant).

REFERENCES

1. DUNDEE, J.; MCCLEARY, N.; & McLoughlin, G. The Hazard of Thiopental Anesthesia in Porphyria. *Anesth. & Analg.* 41: 567 (1962).
2. NORRIS, W. & McNAB, G. W. Anaesthesia in Porphyria. *Brit. J. Anaesth.* 32: 505 (1960).
3. LEPINSKIE, F. F. Porphyria as a Problem in Anaesthesia. *Canad. Anaesth. Soc. J.* 10: 286 (1963).
4. KARK, ROBERT M. Clinical Aspects of the Major Porphyrinopathies. *Med. Clin. North America.* (1955), pp. 11-31.