SUMMARY

A case of macrocytic protein deficiency anaemia in a boy suffering from milk allergy (eczema and urticaria) is described. He was able to take boiled cow's milk. The literature is briefly reviewed.

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Two Cases of Acute Idiopathic Porphyrria

A number of cases of acute idiopathic porphyria have been reported in the literature in recent years, but we think the disease interesting and rare enough to merit the recording of two further cases.

CASE 1

The patient, a single woman aged 20—height 5 ft. 8 in. (173 cm.), weight 6 st. 6 lb. (41 kg.)—was first admitted on June 18, 1948, as an acute abdominal emergency. She gave a history of five days' colicky upper abdominal pain—the onset coinciding with a period—nausea, and obstinate constipation. She had vomited on three occasions. Her past history was negative. Her parents and two siblings were alive and well.

On examination she looked ill. Her temperature was 99.6° F. (37.6° C.), pulse 80, and respirations 20. Her tongue was furred; there was no scleral or cutaneous pigmentation. The abdomen was soft, not distended, and without guarding or local tenderness. Peristaltic sounds were normal. Rectal examination was negative. Heart: apex beat forcible; no enlargement; no murmurs; rhythm normal; rate 80. The blood pressure was 170/120. Chest and C.N.S.: N.A.D. A few hours later she vomited; her temperature was 99° F. (37.2° C.) and pulse 72. The pain continued unabated, and laparotomy was performed. No abnormality was found.

After operation the pain persisted. The urine contained a trace of albumin and "excess urobilin," the blood N.P.N. being 70 mg. per 100 ml. During the ensuing week the pain continued, and on the fifth post-operative day she had an epileptiform convulsion. The blood pressure now was 160/100. The white blood cells numbered 31,000 (polymorphs 95%, lymphocytes 4%, monocytes 1%). The urine still showed a trace of albumin with an occasional granular cast. The blood N.P.N. remained at 70 mm. per 100 ml. The chest x-ray picture and cerebrospinal fluid were normal, and the blood Wassermann reaction was negative. She remained slightly febrile, and 14 days after operation complained of weakness of the legs. Examination revealed weakness, with muscle tenderness of the proximal muscle groups of both arms and legs. The cranial nerves were normal, and there was no sensory loss. A repeat lumbar tap revealed normal fluid. She now began to improve, the blood pressure falling to 130/80, and she was discharged on September 22. No definite diagnosis was made.

She was next admitted on June 13, 1949, with identical symptoms. The combination of severe abdominal pain and marked hypertension, together with the history of a negative laparotomy, suggested porphyria, and examination of the urine led to quick confirmation. Up to the present she has had five attacks. Each began and evolved in the same manner, and neurological complications occurred in all but

one. The last acute episode occurred in January, 1951. She has remained well since, although the urine is often coloured red.

CASE 2

A single man aged 41 was admitted on March 12, 1951. The history was of attacks of lower abdominal pain for four months, becoming more severe in the last three days, and violent eight hours before admission. He was nauseated and constipated.

On examination he was obviously in severe pain. His temperature was 98.2° F. (36.8° C.), pulse 56, and blood pressure 140/90. Some guarding and tenderness in the epigastrium were the only abnormalities noted. After some hours' observation it was decided to explore the abdomen with the possibility of a posterior perforation in mind. Nothing abnormal was found at operation.

Soon after operation he passed port-wine-coloured urine, and spectroscopic examination revealed the presence of porphyrins. On re-examination at this time he had become uncooperative and resentful; the blood pressure was 185/115, and there was obvious recent wasting of the leg muscles, especially the quadriceps. During the next few days the pain persisted, the blood pressure rose to 205/120, and the patient became deluded and irrational at times. The symptoms gradually abated, and by the 10th day after operation he was symptom-free and co-operative, the blood pressure was 130/80, and the urine was normal in colour.

He was discharged from hospital on April 14, and by that time the wasting of the legs was rapidly disappearing. He has been seen as an out-patient since, and has had no further attacks of pain or staining of the urine.

COMMENTS

Both cases were admitted as acute abdominal catastrophes. The association of severe unremitting colicky abdominal pain with vomiting and obstinate constipation suggests serious abdominal mischief, and in some cases may not unreasonably lead to laparotomy. The abdominal symptoms may be clamant, but are seldom supported by definite physical signs of local or general abdominal disease.

In neither case was a firm pre-operative diagnosis made, and laparotomy was undertaken mainly in the spirit of "it is better to be safe than sorry." The disease, although admittedly rare, is probably commoner than has hitherto been realized, and may well account for many "negative" laparotomies. It is, of course, very pertinent in the differential diagnosis of polyneuritis, Landy's paralysis, and even of some psychoses. As Petrie remarks, "The symptoms of acute porphyria tend to be remarkably stereotyped." This is well exemplified in these cases, particularly in Case 1, in which the story is almost monotonous. Yet the diagnosis on the first three occasions of her admission varied from intestinal obstruction to acute obstructive appendicitis. There is, however, the discrepancy between the symptoms and paucity of signs, which may suggest something unusual. If hypertension and neurological signs are present as well the diagnosis should be easy, but hypertension may not be present, at any rate in the initial stages, and the neuropathy tends to develop a little later. If the disease is not borne in mind the diagnosis will be missed, and this may well lead to an unnecessary laparotomy in a seriously ill patient, liable to serious and possibly fatal neurological complications at any time.

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