H. O. WONG AND K. W. CHOW: URETERIC IRREGULARITY IN RENAL-ARTERY STENOSIS

FIG. 1.—Case 1. The collection of blood-vessels from the right common iliac artery extend upwards into the hilum, filling the post-stenotic portion of the renal artery. The lower-pole branches of the right renal artery are now better opacified and look irregular. *Inset*: Retrograde aortogram demonstrating a small right renal artery which shows a localized constriction with post-stenotic dilatation of the vessel. Picture taken $\frac{1}{2}$ second before main picture.

FIG. 2.—Case 1. Excretory urogram taken 15 minutes later shows irregular indentations or scalloping of the right ureter in its upper and middle thirds. The indentations correspond exactly to the site of the tortuous vessels. Note the better concentration of the contrast medium on the right side.

FIG. 3.—Case 2. A delayed film following percutaneous aorto-renal arteriogram, showing periureteric collateral vessels on the right side (arrowed). Note the difference in size of the kidneys.

W. E. GIBB *ET AL.*: SURVIVAL IN WHIPPLE'S DISEASE

FIGS. 1 and 2.—Small-bowel biopsy, February 1963. (P.A.S. stain. Low and high power pictures.)

Survival in Whipple's Disease

[WITH SPECIAL PLATE]

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Whipple's disease, "intestinal lipodystrophy," is now being recognized more frequently and over 100 cases have been recorded since the original description (Whipple, 1907). In the past it had been concluded that the disease progressed steadily to death within a few months or years from the onset, but more recently records have been published of patients living for longer periods (Anton, 1961) and of others responding to treatment with steroids or with adrenocorticotrophic hormone (A.C.T.H.) (Cheers *et al.*, 1961). We wish to emphasize both these points in reporting the case of a patient who has survived for about 20 years from the onset of his symptoms and whose health has materially improved during the past three years, after treatment with A.C.T.H.

CASE REPORT

A leather-goods worker aged 33 was first seen in 1949, at which time he had had a mild polyarthritis for about five years. This chiefly affected the left wrist-joint and the metacarpophalangeal joints of the left hand. He was otherwise well, but in spite of analgesics he continued to have pain. His E.S.R. was normal.

In 1950 he started to have abdominal pain and to lose weight, and his E.S.R. rose to 26 mm. per hour. He was first admitted to hospital in August 1952 with loss of weight, diarrhoea, and a purpuric rash, when it was established that he had steatorrhoea, rarefaction of bones, iron-deficiency anaemia, and a purpura responding to ascorbic acid. Neither a gluten-free diet nor a short course of deoxycortone acetate (D.C.A.) had any effect on his condition, but after a course of cortisone by mouth lasting six weeks he gained 1 st. (6.4 kg.) in weight.

During succeeding years he was admitted several times for chest infections, and on one occasion, in 1957, he had a marked sulphaemoglobinaemia due to the large doses of codeine compound tablets he had been taking for the "arthritis."

In 1960 he had considerable pain in his limbs and was taking at least 30 codeine compound tablets daily, which did not give relief; he was also passing as many as 12 offensive fatty stools daily, and his weight had fallen again, to under 9 st. (50.8 kg.).

Examination showed him to be a wasted anaemic man with a smooth tongue and a generalized brownish colour but with no buccal pigmentation. There was a persistent bluish tinge to the mucous membranes. The blood-pressure was 110/70 and there was bilateral pitting oedema of his ankles. The abdomen was protuberant but no masses were felt and there was no ascites. Proctoscopy showed nothing abnormal.

Initial Investigations.—Hb 7.4 g./100 ml. (50%). E.S.R. 34 mm./hour. W.B.C. 7,000/c.mm. (normal differential). Serum calcium 8.6 mg./100 ml. Serum phosphate 2.4 mg./100 ml. Serum alkaline phosphatase 18.8 K.A. units. Serum electrolytes: sodium 130, potassium 4.1, and chloride 100 mEq/litre. Alkali reserve 24.5 mEq/l. Blood urea 40 mg./100 ml. Magnesium 2.9 mg./100 ml. Electrophoresis of the serum proteins showed a raised α_2 -globulin and diminished γ -globulin. Barium meal and follow-through showed normal progress of the opaque medium through the small bowel, but there was much snowflake flocculation of barium in the jejunum and dilatation of the ileum with exaggerated folds. Barium enema revealed nothing abnormal.

In 1952 a six-day fat balance showed absorption to be 70%; and investigations in March 1952 showed that up to 97.4% of the faeces consisted of fat, the stool melting on warming and solidifying when cool.

Peroral jejunal biopsy showed infiltration of the submucosa with macrophages containing periodic-acid-Schiff positive staining

material. He had been thought to be suffering from Whipple's disease for many years, and this confirmed the diagnosis. With intensive parenteral and oral treatment with iron, calcium, and vitamin B₁₂ the haemoglobin rose to 12.1 g./100 ml. (82%) and an attempt was made to counter the severe loss of weight and the diarrhoea. He was started on oral prednisolone 20 mg. daily, but this caused intractable dyspepsia, and in spite of a reduction in dosage had to be abandoned. During the course of prednisolone his stools became normal in appearance and frequency, but he felt no better in general health and did not gain any weight. A.C.T.H. was tried, and this, in a total of 90 units a week in divided dosage, reduced the bowel habit to one motion a day, and occasionally two or three motions. His weight had increased to 8 st. 9 lb. (54.9 kg.).

Additional investigations on the absorption of haematinics showed a rise in serum-iron level from 48 to 91 μ g./100 ml. after an oral dose of 600 mg. of ferrous fumarate. A Schilling test with ⁵⁸Co-labelled vitamin B₁₂ revealed normal absorption, a 24-hour excretion of 17.6% being obtained.

Serological tests with regard to the arthritis showed a mildly positive latex-fixation reaction, but differential agglutination titres and antinuclear factors were consistently negative.

The patient was again admitted on account of a sudden fall in haemoglobin with the passage of bright red blood per rectum. Sigmoidoscopy suggested an obstruction at about 15 cm. from the anal margin. Mr. J. O. Robinson performed a laparotomy and excluded a neoplastic cause for the bleeding.

At laparotomy the whole of the small gut and mesentery were affected, appearing white and oedematous. When the jejunum was opened for a further biopsy the mucous membrane was seen to have a frosted appearance. A large mesenteric gland taken at the time contained macrophages.

In February 1963 a further small-bowel biopsy specimen again showed clearly the typical histological picture (Special Plate, Figs. 1 and 2) and that this had not changed since 1961. Nevertheless his general health has improved and his weight has increased to 9½ st. (60.3 kg.). A maintenance dose of 80–90 units of A.C.T.H. weekly is being continued.

COMMENT

We record this case, of a patient who first developed Whipple's disease 20 years ago, to illustrate the markedly chronic course which the disease may pursue. Another feature is the striking improvement in his general health during the past three and a half years while having A.C.T.H. This degree of improvement could not be obtained with D.C.A., and although he benefited from a short course of cortisone the effect of subsequent oral prednisolone could not be assessed because of gastric intolerance. Lastly, although there was considerable improvement in his general health, this was not reflected in the serial small-intestine biopsies.

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