CROHN'S DISEASE OF THE STOMACH?


JANE FULLERTON, M.B., B.Ch., D.P.H., F.C.Path., Consultant Pathologist.

St. Olave's Hospital, London, S.E.16.

The condition now known as regional ileitis was established as a clinical, radiological and pathological entity by Crohn, Ginzburg and Oppenheimer in 1932. The histological nature of this disease was further underlined by Hadfield in 1939. Regional ileitis was thus established as a disease of unknown aetiology, characterised by a thickened segment of ileum, consisting histologically of non-caseating granulomatous foci, together with a granulomatous involvement of the regional lymph nodes. Skip areas in the bowel were described as a typical feature of the condition.

This type of granulomatous condition affecting the ileum together with the colon (Crohn, 1949), and cases involving the colon alone (Armitage and Wilson, 1950), were later described. Regional ileitis, together with the involvement of the upper gastrointestinal tract was also reported (Rose 1949, Martin and Carr 1953, Richman, Seifer, Winkelstein, Kirschner and Steinhardt 1955, Miller, Sandweiss and Schwachmann 1956). Finally, cases have been described where this granulomatous condition affects the stomach alone.

Because of its comparative rarity and interest, a further case of isolated granulomatous gastritis is reported.

Case Report

Mr. W.F., a 57 year old caretaker, first presented at this hospital in August 1953. He gave a 15-year history of recurrent episodes of pain in the epigastrium and right hypochondrium. The pain would start one hour after a meal, persist for about 10 minutes and was occasionally associated with vomiting. He was tender in the epigastrium and

REFERENCES


*Present address: The Lahey Clinic, Dept. of Surgery, 605 Commonwealth Avenue, Boston, U.S.A.
Investigations performed, which are summarized below:-


March 1962 Barium meal—a shallow gastric ulcer was seen on the upper third of the lesser curvature. A diverticulum of the second part of the duodenum was noted.

May 1962 Gastroscopy—the gastric rugæ were poorly developed.

August 1962 Gastroscopy—once again gastric atrophy was noted.

Re-Admission. He was re-admitted to hospital in October 1963. Epigastric pain and vomiting were still prominent symptoms, but over the preceding 6 months he had developed anorexia and had lost 3 stone in weight. Except for clubbing of the fingers, there were no abnormal physical signs. A barium meal done on this occasion showed a constant narrowing and rigidity of the distal third of the stomach. There was an almost complete loss of mucosal folds in the whole of the stomach (Fig. 1). These appearances were strongly suggestive of a carcinoma of the stomach, of the linitis plastica type. The duodenal diverticulum was again noted.

Operation. At operation the stomach was considerably thickened, particularly in its distal half, but it was quite supple and freely mobile. There was a healed gastric ulcer at the lower end of the lesser curve. The regional lymph nodes were not enlarged. There were numerous adhesions in the upper abdomen. There was no other abnormality. A Polya-type sub-total gastrectomy was performed. He had a persistent pyrexia post-operatively and discharged a large quantity of pus through the upper end of the incision 3 weeks after the operation. His temperature settled and his general condition rapidly improved after this, but he had obviously developed a large gastric fistula. This slowly healed over the next four weeks.

Pathology. Macroscopic examination of the specimen showed a gross thickening of the stomach. The gastric rugæ were pale and flattened and had a cobbledstone appearance seen in cases of regional ileitis. There was no mucosal ulceration.

Microscopical examination of sections taken from several parts of the specimen showed an atrophy of the gastric mucosa. The layers of the stomach were infiltrated by lymphocytes and there were numerous focal granulomata present in all the layers (Fig. 2). Giant cells were associated with each granuloma, but there was no evidence of necrosis or caseation (Fig. 3). Sections examined under polarized light showed typical Schaumann bodies containing crystals (Fig. 4). Sections examined for acid fast bacilli and fungi were negative.

Investigations. Because of these findings, further investigations were carried out:-

Serum Proteins—Total 6.5 g./100 ml.; electrophoresis—slightly decreased albumin and raised Alpha, globulin. Liver Function Tests—Bilirubin 0.3 mg./100 ml. Alkaline Phosphatase 10 units. Thymol Flocculation—negative. Blood film—no abnormality seen. Hæmoglobin 13.3 g./100 ml.
FIG. 3.—Three focal granulomata shown in the submucosa (x110). Giant cells are associated with each granuloma, and there is no evidence of necrosis or caseation.

FIG. 4.—High-power view of part of a granuloma under polarized light (x530). This shows a typical Schaumann body containing crystals.


Intradermal skin tests and subsequent skin biopsies to antigen prepared from tissue involved in Crohn's disease of lymph glands, ileum and colon were all negative.

Barium examination (done 4 months post-operatively)—there were no visible mucosal folds in the gastric remnant, but otherwise the esophagus, gastric remnant and small bowel were normal.

The patient was last seen 5 months post-operatively. He had not gained any weight and still complained of disabling post-prandial epigastric pain and vomiting.
TABLE 1

THE CLINICAL FEATURES OF REPORTED CASES OF ISOLATED GRANULOMATOUS GASTRITIS.

<table>
<thead>
<tr>
<th>No.</th>
<th>Author and Sex</th>
<th>Age and Sex</th>
<th>Clinical Features</th>
<th>Physical signs</th>
<th>Histamine test meal</th>
<th>Barium meal</th>
<th>Gastroscopy</th>
<th>Other tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Gore and M. Mc-</td>
<td>26 M</td>
<td>Pain, vomiting,</td>
<td>-</td>
<td>-</td>
<td>Cone shaped rigid deformity of antrum diagnosed as carcinoma</td>
<td>- W.R. negative, Mantoux negative, X-ray hands N.A.D.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Carthy, 1944</td>
<td></td>
<td>weight loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Appell and others, 1951</td>
<td>48 F</td>
<td>Vomiting, weight loss</td>
<td>-</td>
<td>Achlorhydria</td>
<td>Pyloric obstruction due to carcinoma</td>
<td>- W.R. negative.</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>McKusick, 1953</td>
<td>49 F</td>
<td>Pain, vomiting,</td>
<td>-</td>
<td>Achlorhydria</td>
<td>Contraction and rigidity of stomach—limitis plastica</td>
<td>- W.R. negative.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>weight loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Scott and others, 1953</td>
<td>25 M</td>
<td>Anorexia, pain, vomiting, haematemesis</td>
<td>-</td>
<td>-</td>
<td>Small stomach with atrophic rugae and poor peristalsis</td>
<td>Gastric atrophy</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Self, 1957</td>
<td>35 F</td>
<td>Anorexia, pain,</td>
<td>-</td>
<td>-</td>
<td>Rigid pyloric antrum—limitis plastica</td>
<td>- W.R. negative, Mantoux negative, X-ray hands N.A.D.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>vomiting, diar-</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>rhoea, weight loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Eckstein and Parker, 1958</td>
<td>38 M</td>
<td>Pain, weight loss</td>
<td>-</td>
<td>-</td>
<td>Rigid pyloric antrum—limitis plastica</td>
<td>- W.R. negative, Mantoux negative, X-ray hands N.A.D.</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Bruce and Daber, 1954</td>
<td>7 F</td>
<td>Pain, vomiting, weight loss</td>
<td>-</td>
<td>Achlorhydria</td>
<td>Small stomach with atrophic rugae and no peristalsis—limitis plastica</td>
<td>Gastric atrophy</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Bowen and Berry, 1961</td>
<td>59 F</td>
<td>Pain, haematemesis</td>
<td>-</td>
<td>-</td>
<td>Narrowed antrum—carcinoma</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Fahimi and others, 1963</td>
<td>51 M</td>
<td>Pain, vomiting, weight loss</td>
<td>-</td>
<td>Achlorhydria</td>
<td>Dilated stomach antral narrowing—carcinoma or healed ulcer</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Fahimi and others, 1963</td>
<td>56 F</td>
<td>Pain, vomiting, Epigastric weight loss, haematemesis</td>
<td>-</td>
<td>-</td>
<td>Shrunken stomach—carcinoma or healed ulcer</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>Fahimi and others, 1963</td>
<td>47 M</td>
<td>Acute pain</td>
<td>Some free acid</td>
<td>-</td>
<td>Antral narrowing—carcinoma</td>
<td>-</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>Kune and Fullerton, 1964</td>
<td>57 M</td>
<td>Anorexia, pain, vomiting, weight loss</td>
<td>-</td>
<td>-</td>
<td>Narrowing and rigidity of distal stomach with loss of mucosal folds—limitis plastica</td>
<td>Gastric atrophy</td>
<td></td>
</tr>
</tbody>
</table>

Discussion

The Clinical Features of Isolated Granulomatosus Gastritis

Eleven further cases of well documented isolated granulomatosus gastritis have been found in the British and American literature. The clinical features of these are summarized in Table 1. The sexes are evenly distributed. The ages range from 25 to 59. The majority of cases have presented with epigastric pain, vomiting and loss of weight. Haematemesis was the main presenting symptom in two cases. One case presented as severe epigastric pain of sudden onset. There were no characteristic physical signs. Histamine-fast achlorhydria was present in 4 of the 5 cases where this investigation was done. Gastroscopy was done in 3 cases, all showing gastric atrophy. Barium meal examination was done in 11 cases, and it either showed an appearance resembling limitis plastica or else appearances of an antral carcinoma. A pre-operative diagnosis was made in 11 cases and it is emphasized that in all of these the diagnosis was carcinoma of the stomach, often of the limitis plastica type.

The Management of Isolated Granulomatosus Gastritis

The diagnosis will only be made by the pathologist, unless the surgeon is aware of the existence of this uncommon condition. At operation the part of the stomach involved in the granulomatosus process will feel thickened and rubbery and there will usually be fleshy regional glands present. The condition may be suspected at operation when, despite extensive gastric involvement, the stomach is relatively mobile, not fixed to any of the adjacent structures, and the omentum, peritoneum and liver are free of metastases. The operation of choice is either partial or total gastrectomy according to the extent of involvement. This was done in 10 of the 11 reported cases and a follow up is reported in 5 cases, all of whom appear to be cured.
TABLE 2

THE CLASSIFICATION OF GRANULOMATOUS GASTRITIS.

1. AETIOLOGY KNOWN OR CLASSIFIABLE.
   (a) Regional ileitis with gastric involvement.
   (b) Sarcoidosis with gastric involvement.
   (c) Chronic specific granuloma of the stomach, e.g. tuberculosis.
   (d) Reaction to foreign material, e.g. chronic beryllium disease.
   (e) Eosinophilic granuloma of the stomach.

2. ISOLATED GRANULOMATOUS GASTRITIS OF UNKNOWN AETIOLOGY.
   (a) Isolated sarcoidosis. (Sarcoid Reaction).
   (b) Crohn’s disease of the stomach.
   (c) Reaction to unknown ingested substances.
   (d) ?

A gastro-jejunostomy and vagotomy was done in one case, but there is no follow up. In the present case only a partial gastrectomy was done, though the whole of the stomach was involved in the process. The patient continues to have symptoms, probably because we are dealing with an incompletely treated condition. Should he continue to have these disabling symptoms, he will almost certainly come to total gastrectomy.

The Classification of Granulomatous Gastritis

Cases of granulomatous gastritis readily fall into one of two groups (Table 2). In the first group are cases where the cause of the granulomatous condition is either known, e.g. chronic beryllium disease, or else the condition is classifiable as part of a more generalised disease, e.g. sarcoidosis. In the second group are cases of isolated granulomatous gastritis of unknown aetiology. Such a case is reported in this paper. Despite full investigations, it was not possible to classify this condition. The presence of Schaumann bodies with typical inclusion crystals did not help to solve the problem of classification, as the granulomata of sarcoid, non-caseating tuberculosis, intestinal Crohn’s disease and chronic beryllium disease are indistinguishable (Jones et al. 1960 and 1964). Indeed it is staggering to note that the response of the tissues to such widely different agents as a bacillus and a metal may be identical.

Should we label this case as “Gastric Sarcoidosis”, “Reaction to Unknown Ingested Materials” or “Crohn’s disease of the Stomach”, as has been done in some of the other cases? Is the stomach that the pathologist examines merely the end-result of tissue response to a number of different agents? Clearly then, assigning an aetiological label to this condition we have got no further in the understanding of the disease, but have only given way to intellectual complacency.

Summary

A case of isolated granulomatous gastritis of unknown aetiology is reported. The clinical features of the other reported cases are reviewed. The management and the problems in the classification of this condition are discussed.

We would like to thank Mr. R. A. V. Lewys Lloyd, Senior Surgeon, St. Olave’s Hospital, London, for permission to publish this case, and Dr. W. Jones, Williams, Institute of Pathology, The Royal Infirmary, Cardiff, who prepared the special histological sections, contributed the Crohn’s tissue antigens, and gave us helpful advice in the preparation of this paper.

REFERENCES


