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CROHN'S DISEASE

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CASES of Crohn's disease appear to be increasing in frequency. Of the true pathology little as yet seems to be known and the causation still is covered by the unsatisfactory word "idiopathic." The presenting symptoms vary considerably and it seems difficult to foretell which cases will resolve and which will develop the serious complications of obstruction, perforation and fistula formation. Thus to describe any case as having classical symptoms is hardly accurate and yet the following case seems to fulfil so closely the postulates of Crohn that it seems worth recording.

Case History.—Sapper S. N., for the first time $9\frac{1}{2}$ years ago, started to complain of abdominal symptoms. He was at that time 18 years of age and working as a cabinet maker. Suddenly, without cause, he developed diarrhoea with abdominal pain. This pain which was situated in the lower abdomen was very severe and was unaccompanied by vomiting. The pain itself was very severe. It doubled him up and he was unable to straighten himself. At the same time his motions varied from six to twelve per day. This attack lasted one week. Six to nine months later he had a further similar attack which persisted on and off for nine months. The frequency of stools varied very considerably but the intensity of the pain was what disturbed him most. His stools were consistently negative to all dysenteric organisms. He lost several stones in weight. His normal weight had been $10\frac{1}{2}$ stones. His doctor's prescriptions of bismuth, etc., did not help his condition but spontaneous recovery took place and he had another period of twelve months when he was quite well again except for very minor attacks lasting only one or two days.

On the appearance of his third attack he was sent to his civilian hospital as a suspected colitis and was an in-patient for three months. He had the full gamut of investigation—faeces, sigmoidoscopy, bismuth enema, through-going barium meal, all with negative findings. He was passing some bright red blood but this was attributed to hæmorrhoids. He left hospital, he says, no better, very weak and he had lost any weight he had regained. At times his motions reached twenty per day, which left little time for cabinet making! This state of affairs continued until 1938, when again for twelve months he had a period of recovery and so was enrolled in the Army as A 1 in 1939.

He was in a Military Hospital in 1940 and again in 1941. He was admitted for fifteen weeks in 1942; again X-rayed, sigmoidoscoped, and the pathologist again, supplied with numerous stools, all of which were negative. He left hospital in September, 1942, and was re-categorized B 1, and was actually put on a draft for overseas. At the last moment, as he was still having several motions per day, he was taken off the draft. He was posted to a series of units at home still with pain and diarrhoea persisting and in September, 1943, he was re-categorized C 2—permanent.

He now passed into the hands of the psychiatrist who said his "nerves were in a shocking state." This is not surprising if having twenty motions daily. From there he went from one job to another—clerk, storeman, etc., and finally was again admitted to a general hospital in May, 1944.

Physical examination revealed a miserable little man, age 28 years, but looking 20 years older and weighing 6 stones. He was having six to twelve motions daily, very severe pain and an irregular daily pyrexia rising to 100 to 102 with pulse 100 to 120. He was racially unfitted to bear pain and after 9 years it had started to show its effects and with each attack

of pain he cried out and demanded morphia or other analgesic. His abdomen was retracted and firm and, although no mass was palpable, tenderness was marked in both iliac fossæ and in the suprapubic region.

Repeated examination of his stools was negative and therefore one ruled out a chronic lesion of his colon. His blood was examined for possible agglutinins with negative results. This was carried out on account of his continued pyrexia. He was having urinary frequency and pain at the end of micturition and yet the daily twenty-four-hour quantity was normal and the urine was free from abnormal contents—thus suggesting a normal urinary tract with an extra vesical inflammatory lesion irritating the distending bladder and causing severe dragging pain as the bladder emptied. This fact was confirmed on rectal

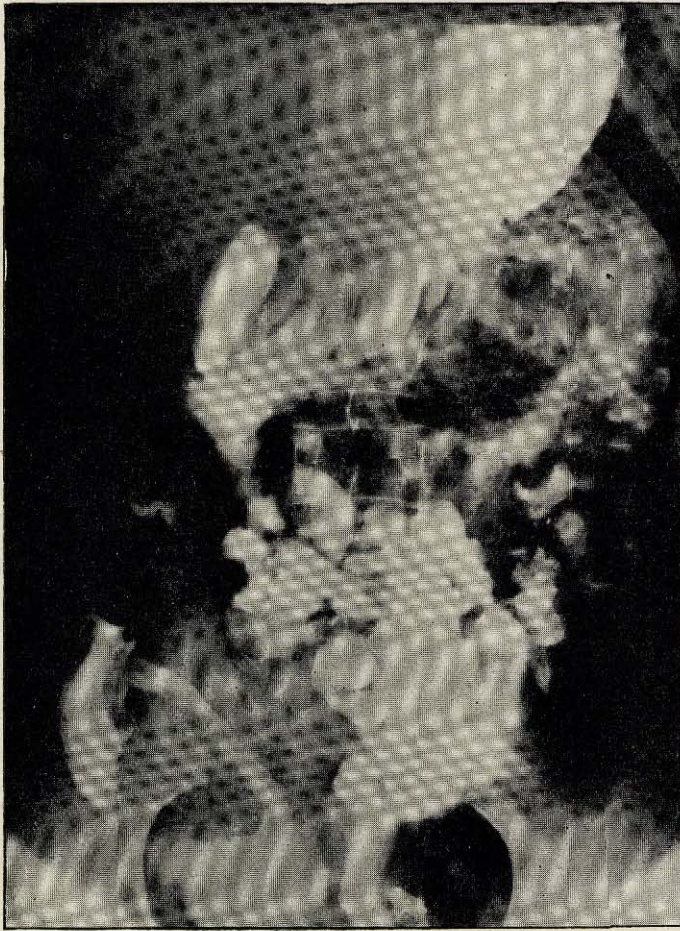


FIG. 1.—Cæcum starting to fill. Narrowing of terminal ileum. Some obstruction proximal to this in the pelvis.

examination by the presence of a very tender mass in the recto-vesical pouch. Abdominal tenderness was present across the lower abdomen and entrance to the false pelvis but neither ascending nor descending colon was tender or thickened. It was considered that the tenderness was due to a loop of terminal ileum inflamed and thickened that had fallen into the pelvis and was adherent to the posterior wall of the bladder and that a fistula with abscess had formed or was forming.

Sigmoidoscopy was not carried out at this stage but, some months before, it had shown a localized area of œdema of the mucosa at the recto-sigmoid junction—suggesting again the

presence of localized extramural inflammation. The fever throughout was irregular and not characteristic, rising to 101 to 102 with his pulse ranging from 100 to 120. Blood-count showed a secondary anæmia with hæmoglobin 77 per cent and white cells varying from 11,400 to 18,900. The through-going bismuth meal clinched the diagnosis. It showed the "string sign" of Crohn, marking the site of the terminal ileum and, equally important, that the last loop of the small intestine was fixed and immovable in the pelvis—tilting up the patient made no change in its position (fig. 1).

Treatment.—Operation was decided upon. The operation of choice was to be a right colectomy but it was envisaged that the loop of ileum might now be so fixed in the pelvis that removal would be impossible without damage to bladder or rectum. If this were so, what should one do? Short circuiting ileum to transverse colon and leaving this long loop either excluded or not excluded from the faecal stream frequently does not give satisfactory drainage (a case so treated a few weeks previously and seen by one of us strongly confirmed this as the right colon became distended and had to have a cæcostomy performed leaving the field for the final resection a very unpleasant one).

It was decided, if the ileum could not be freed, to leave it *in situ* with both ends open and exteriorized, and at the same time to proceed to complete the right colectomy. Later after weeks of irrigation with saline, etc., and after the inflammation had subsided, one might attempt to remove this U-shaped loop from the pelvis.

In actual fact at operation the loop did separate. An abscess was found in the pelvis and a small sinus communicated with the ileum. This operation was not easy but a right colectomy was carried out. A drain was placed in the pelvis and post-operative convalescence was uneventful and straightforward. Much of the success of the operation was due to the satisfactory pre-operative, operative and post-operative resuscitation with blood, plasma, glucose-saline, etc., which the patient received from the transfusion officer, Captain (Miss) B. Winterton, R.A.M.C.

The specimen corroborated the known facts of Crohn's disease. (1) The thickened mesentery, one inch—with abscesses in it; (2) the disease stopping abruptly at the ileo-cæcal valve; (3) œdema and swelling more marked than fibrosis; (4) shortening and "concertinaing" of the affected gut; (5) adhesions and fistula.

The specimen measured about two feet when removed but when the mesentery was freed it measured about five feet. In fact, what is the length of the small intestine? The anatomy books state twenty-two feet and yet at times it is possible for a ten to twelve-foot tube to appear at the anus and still project at the mouth. The freeing of the mesentery gives an artificial idea of its length.

Pathological Investigation.—(1) Stools Culture (20 times): *B. coli.* and *Streptococcus faecalis*—tubercle bacillus not seen. Macroscopically: Loose, pale, foul smelling. No blood. No mucus. Microscopically: Pus cells, few monocytes and epithelial cells, no cysts or trophozoites of *Entamoeba histolytica*. No helminthic ova.

(2) X-ray: (i) Pelvis, (ii) Chest, Nil.

(3) Urine: Nil.

(4) Blood-Count:	W.B.C.	11,400	18,900
Neutrophils		74.5	71.0
Eosinophils		0.0	1.0
Basophils		0.5 later	0.0
Lymphocytes		19.5	18.0
Mononuclears		5.5	10.0

(5) Sigmoidoscopy: Œdema at recto-sigmoid junction.

(6) Blood Group: A.

Report by Radiologist (Captain H. Haggar, R.A.M.C.).—Barium Meal Examination: Stomach and duodenum were radiologically normal, but a radiogram taken two and a half hours after ingestion of the meal showed: (1) An irregular narrowing of the lumen of the distal foot of the ileum; (2) absence of normal segmentation of the involved ileum; (3) a coil of involved ileum lying in the pelvis (fig. 1). There was no dilatation of the small gut proximal to the diseased area.

Screen examination at this stage with the patient in various positions (prone, supine, head low, feet low) showed the loop of ileum in the pelvis to be firmly fixed. A further

radiogram taken after six hours showed a similar state of affairs, but in addition a poor filling of the cæcum (fig. 1). At twelve hours the small gut was empty and in twenty-four hours no trace was visible in the entire alimentary canal.

After operation the specimen was filled with barium solution and air, and a soft radiogram taken (fig. 2). The narrowed lumen and thickened wall is seen in detail. There is an enlarged lymph gland lying in the mesentery of the ileo-cæcal angle. It contains two small areas of calcification.

Pathologist's Report upon the Specimen (Major Winston Evans, R.A.M.C.).—(a) Macroscopic: The affected portion of the small intestine was about 15 inches long and extended to the ileo-cæcal valve. The cæcum was normal and the portion of small gut beyond the lesion was unaffected. The attached mesentery was much thickened and contained two large glands containing calcified matter. The peritoneal surface was engorged and showed

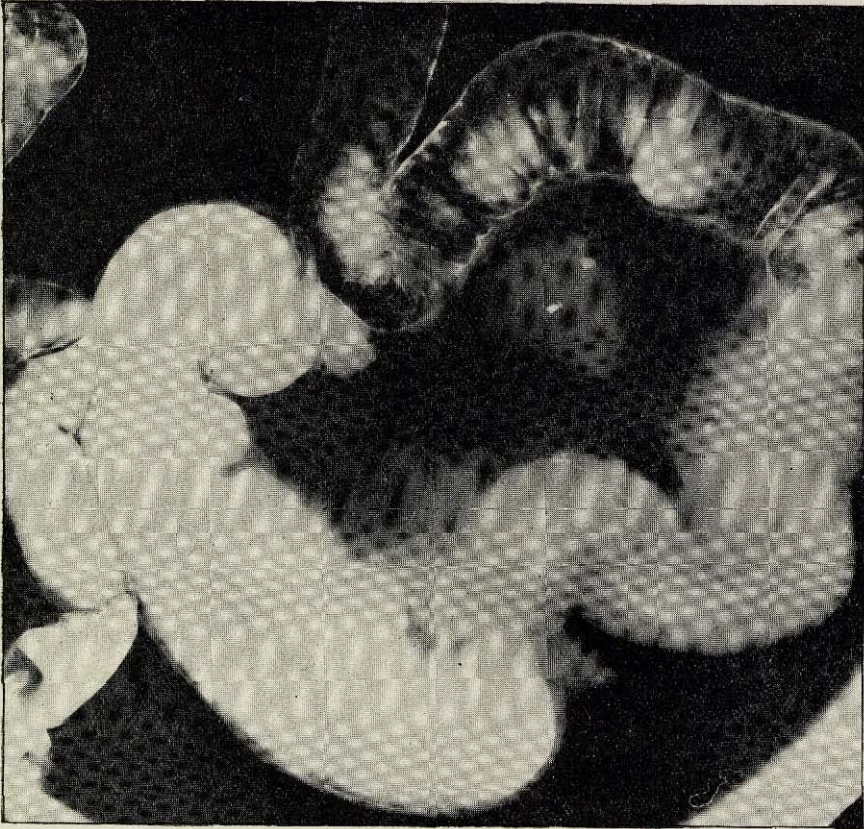


FIG. 2.—To illustrate the regional and terminal distribution of the disease. Proximal gut normal in texture and size. Appendix, although not seen in this, was normal.

a perforation where it had been adherent to the bladder. Section of the bowel wall showed it to be turgid, much thickened and hypertrophied with a tortuous and irregular narrow lumen. The mucosa and submucosa was thickened and oedematous with loss of intestinal folds and in parts ulcerated. In other parts the mucosa had been completely destroyed and replaced by a layer of atrophic epithelium. (b) Microscopic: Histological examination shows a chronic inflammatory process involving all the layers of the gut with a marked infiltration with plasma cell and fibroblastic reaction. The mucosa in different sections shows varying stages of destruction and is completely absent in some areas, a covering of epithelium remaining. The submucous lymph follicles are enlarged and some show an endothelial cell

proliferation. Giant cells systems are very scanty. There are no areas of caseation or focal necrosis.

Lymph glands show evidence of old tuberculosis with much fibrosis and a replacement of the remaining germinal centres by giant cell systems and focal areas of caseation.

SUMMARY.

(I) Details of a long-standing case of Crohn's disease treated by resection of terminal ileum and right colon are given.

(II) Diarrhoea, abdominal pains, and loss of weight were the presenting symptoms.

(III) The symptoms had persisted intermittently over a period of 9½ years.

(IV) The diagnosis was made on clinical examination, confirmed by the negative laboratory findings and clinched by the typical X-ray picture.

(V) Radiography of the resected specimen gives a good idea of the area involved, how contraction occurs in its length and how tight a stricture of the small intestine must be before obstructive symptoms appear.

The treatment of the above soldier was carried out in a General Hospital commanded by Colonel R. G. Shaw, M.C., to whom our thanks are due for permission to forward the case for publication.

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