SECTION XII—"The Clinic"

Typical and Atypical Terminal Ileitis

By

A. GALAMBOS, M.D.*
and

W. MITTELMANN, M.D.
NEW YORK CITY, NEW YORK

"Terminal ileitis" or "regional ileitis" designates a condition described in 1932 by Crohn, Ginzburg and Oppenheimer as a "non-specific granulomatous inflammation of the terminal ileum."

The disease is a well defined ulcerative enteritis, in the greatest majority of the cases restricted to an average extension of about ten inches of the terminal portion of the ileum.

This process is followed by a disproportionally marked chronic inflammation or connective tissue reaction in the covering submucosa, muscularis and serosa, producing a striking thickening of the wall of the gut, with a subsequent narrowing of the lumen, which results in stenotic or even in obstructive symptoms. The involvement of the serosa leads at times to ascites, more often however to an adhesion formation between the ileal coils, or between the small and large intestines as well as between the intestines and abdominal wall. The very marked tendency to a characteristic fistula formation, producing communicating fistulous tracts among the organs involved, viz.: the ileum, colon, abdominal wall, retroperitoneal tissue, should be emphasized.

The granulomatous infection secondarily involves the mesenteric glands. The thickened, strikingly infiltrated mesentery acts like a retroperitoneal mass, fixing and inhibiting the free peristaltic movement of the thickened ileum, huddling it into a mass, producing the manifestations of an apparent adhesion, when, often there is no peritoneal adhesion actually present. It is instrumental in producing outside of adhesions, secondary break-downs of the tissues, suppuration, retro-and intra-peritoneal abscesses and fistula formation.

The underlying pathology may lead to a palpatory finding of an indefinite resistance, or of a large inflammatory tumor in the area involved.

The macroscopical appearance and the microscopical histological findings in the resected specimen are uniform and pathognomonic, often only per exclusionem.

The X-ray examination reveals more details and characteristic hints than does the physical examination. In well defined cases, the diseased area is marked by a large filling defect, with a typical "string-sign," representing the direct visualization of the stenosed lumen of the terminal ileal coils. Fistulous tracts may similarly give a resemblance to the string-like appearance on the films.

The main points in the history of the case are: the age of the patient, the protracted course of the disease, diarrhea, undefined pain, occasional fever, slight leukocytosis, secondary anemia, tendency toward fistula formation, failure of previous appendectomies to improve the condition.

It is interesting to note that practically all the patients who came under observation were young adults. The question arises as to the fate of these patients as they arrive at old age. If we disregard the quite improbable outcome of a complete reabsorption, except probably in its earliest stage, one is forced to the conclusion, that all these patients die before reaching old age.

Early recognition of the condition is urged, because early diagnosis followed by adequate surgery (resection), leads, as a rule, to a cure, while a failure to recognize the disease results in chronic marasmus, leading eventually to a fatal outcome.

Incorrect diagnosis—as the history of most of the cases proves—often leads to unnecessary appendectomies, failing to afford any relief to the patient's condition, in fact, often preparing the way to a secondary fistula formation.

One of the causes responsible for the failure to recognize the ailment, in our opinion is due to the fact, that the X-ray examination of the ileocecal region is performed routinely, six hours after the barium intake, . . . at a time when the terminal ileal coils in that particular area are as a rule empty already. The X-ray examination will reach no conclusion, and "negative findings" will be reported.

Instead of the routine X-ray examination with a fixed standard time of six or twenty-four hours, it would be more correct to have an X-ray study instituted using varying time intervals, and varying technic, dictated by the individual needs of the case. As an instance in both of my cases the routine six hour examination produced diagnostically irrelevant pictures. In the first case, the films taken two and three hours after the barium intake, in the second case the thirty-six hour pictures proved to be most decisive and diagnostically most important. As a rule, the two, three or four hour exposures will prove to be of the greatest value.
The disease does not seem to be extremely rare. Crohn reported in his first series in 1932 about 14 cases; two years later the number of cases referred to was 42. Kantor mentions six cases in his paper. We ourselves diagnosed terminal ileitis in the last ten months, three times. Two of this series already underwent an operation and the operative and pathological findings proved the correctness of the diagnosis.

In all of our cases diagnosis has been made pre-operatively, in fact, in the first case strong suspicion has been expressed for this diagnosis, even before the X-ray examination has been performed.

CASE REPORTS

Case 1, refers to a 42 year old white female, first seen at my office the 13th of March, 1934. She had diarrhea, of 15 years' duration. Her disease had a slow onset, and a slow progress. She had 3-4-5 mushy bowel-movements daily, without a noticeable admixture of pus, mucus or blood. She had no tenesmus, but she noticed an occasional periumbilical griping pain during defecation.

There was a more marked loss of weight during the last two years. She was beset by chills and fever (to 102 F.), of a few days' duration, once in every few months. Her general feeling during the intervals was quite satisfactory. Her doctor diagnosed these febrile attacks as "grippe." Her loss of weight during the last 6 months was between 20 and 25 lbs.

On the 29th of July, 1933, patient was suddenly taken severely ill, with chills, fever and periumbilical pain and was rushed to a Brooklyn hospital.

Fig. 1. Case 1. Terminal Ileitis. Typical case. Two hour film. Note the "string-sign" (arrow) and the "filling defect" in the ileocecal area. (Other parts of the G.I.T. are visualised by previous Ba. intake).

Fig. 2. Case 1. On both the upper and lower films, the "string-sign" and the "filling defect" manifest marked similarity to the picture of the previous figure. "String-sign" (arrow) proved at operation to be the direct visualization of a narrowed ileal lumen and not an ileocecal fistula. All other pictures are nearly identical.

The disease was suddenly taken severely ill, with chills, fever and periumbilical pain and was rushed to a Brooklyn hospital, where appendectomy has been performed. Patient remarks, that according to her surgeon's description the appendix removed has been found normal, but he noticed during operation certain unusual puzzling anomalies over the small intestines. The latter looked "swollen," like "sausages," or, resembling typhoid intestines. Between the appendix and sigmoid, retroperitoneal abscesses were found. They were opened and drained. Patient left the hospital after three weeks, and felt all right for the next three months, when the surgeon had to open the abdominal wall and drained externally an intestinal fistula. Patient felt generally comfortable as long as the fistula drained freely, but, as soon as it became blocked up, symptoms appeared. The surgeon had to intervene four more times to restitute and assure free drainage.

Her history was otherwise irrelevant.

On physical examination the abdomen showed spontaneous tenderness, which increased either upon touch or on examination even through the vagina or rectum. In the right lower quadrant there was a large indefinite mass palpable. The extreme sensitiveness of the abdomen decreased the following day, after the fistulous tract—incidentally just blocked—opened spontaneously, and drained pus again, freely. Presence of post-operative scar and fistulous tracts completes the findings over the abdominal wall.

Urine contained traces ofalbumen, with a negative findings in the centrifuged sediment. Feces gave slightly positive reaction for occult bleeding (Weber test). Hemoglobin was 58%. White blood count was 15,000, with 98% polymuclear, 7% large mononuclear cell, and 5% lymphocytes. The temperature on 'the first day of examination, when the fistulous tract failed to drain, was 102° F. The following day, after it broke open, temperature dropped to normal, pain subsided, white blood count fell to 14,000, with 72% polymuclear, 8% large mononuclear cells, and with 20% lymphocytes. The discharge consisted of a copious, odorless, thin and purulent material.

On account of this unusual history and physical findings, strong suspicion has been expressed as to the possibility of a terminal ileitis, and the subsequent X-ray examination served to substantiate this a priori diagnosis.

The X-ray examination of the gastrointestinal tract revealed the stomach, duodenum, jejunum and colon to be normal. All the pathology present was confined to the ileum.

The right lower quadrant of the abdomen was grossly characterized by a large filling defect of the ileal coils. The "string-sign" (Crane-Kantor) was demonstrable on all the films. It was interesting to note that the very last portion of the ileum in a length of 1-1½ inches was visualized as fairly normal. The proximal loops were not visibly distended. The 6 and 24 hour pictures were "negative," i.e. no visualization of the terminal ileal coils, nor of the proximal loops was demonstrated, evidencing the absence of an ileal obstruction. Films taken 2 and 3 hours after the barium meal proved to be the most convincing, visualizing the narrowed intestinal lumen ("string-sign"), with an otherwise falling filling of the other ileal loops (Stierlin's sign).

On re-operation, a sixty (69) inch portion of the terminal ileum, the cecum and two-thirds of the ascending colon have been removed, and an anastomosis between the ileum and the middle of the transverse colon instituted. The removed small intestine's wall was very thick, and its lumen greatly narrowed. No abscess was found at that time. The external fistula led into the ileum, four