FURTHER ON THE PROBLEM OF REGIONAL ENTERITIS

P. Altankov, P. Chervenkov, L. Gospodinov

The range of information on regional enteritis, respectively ileitis, insofar etiopathogenesis, diagnostics and management are concerned is continuously extended and the interest in this malady is no more casuistical, but rather clinically conditioned. The widespreading is characteriszed by a certain degree of endemy, more frequent in some geographical regions. There are countries in which it represents a major part of the acute surgical abdomen (USA, England, Belgium, France), the highest number of cases being reported in the United States (Crohn and Van Patter alone published a series comprising more than 600 patients), in USSR — about 50 according to Lenda and in Bulgaria — 14. It is universally accepted that mostly affected are males, with a predilection for younger age, but nevertheless, cases in advanced age have been reported as well as small children (3-year-old) and twins (mono- and dizygotic).

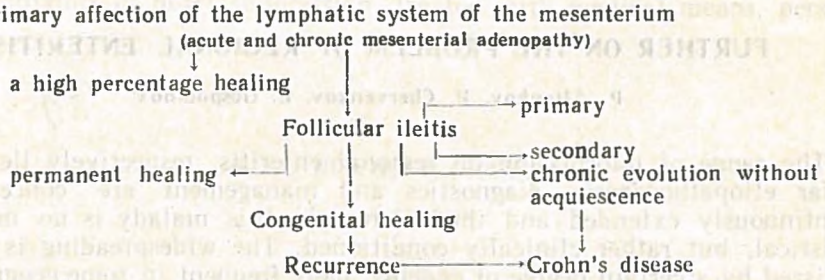
As a distinct nosological unit, the condition was described in 1932 by Crohn, Ginsburg and Oppenheimer, calling it "terminal ileitis" because they believed that it occurred merely at the distal end of the ileum (Crohn's iron law). Later on, however, similar morbid alterations were described in different sites of the intestine: jejunum (Harris, Bell, Brunn), duodenum (MacCarthy), colon (Morson, Waren, Duchess, Lenda), rectum (Popescu-Urlueni, Saptefrati). The latter evolution accounts for a revision of the original notion for the disease and its denomination — terminal ileitis (coined also Crohn's disease) was converted in regional enteritis, regardless of the fact that, according to Pugh, the terminal portion of the ileum appears involved merely in 74% of the cases. Segmentary affections of the small and large intestine are encountered as well as of the small and large intestines simultaneously with the ileo-cecal angle, as in one case of our personal series.

Of the numerous theories for the etiopathogenesis of regional enteritis: nervereflectory (Sozon-Yaroshevich), traumatic (Askanasi Stokes), chronic invagination (Mueller), irritative substances absorption (Waren, Sommers) and others, lately emphasis is being laid on the viral theory (postulated by Verges), accepting a lymphadenitis as the onset of disease, presumably caused by adenovirus.

Of the great number of observations made on regional enteritis, the inference seems warranted that the primary pathologic process develops within the regional lymph nodes and vessels of the mesenterium of the involved intestinal segment, leading to edema, tighthening and fibrous degeneration within the intestinal wall. Against the background of the changes just described, the circulation is impaired and trophic changes

occur accompanied by ulceration of the mucosa, hence opening the road for secondary infection.

Ever since 3—4 years Bousson, Bard and Jourde, accepting the evolution of the process outlined in this paper, draw attention to the follicular ileitis stage as initial in affections of the intestinal segment and evolving in various directions, submitting furthermore the following scheme:



It is obvious from the scheme that the development of follicular enteritis (in fact, these being the acute forms of Crohn's disease) within the wall of the involved intestinal segment should be considered as the onset of regional enteritis, whereas the acute or chronic mesenterial lymphadenitis is the primary etiological moment. In all likelihood, terminal ileitis is the most frequent because the terminal portion of the small intestine has the most strongly developed lymph apparatus, intestinal content toxicity therein is of a higher degree, the content stasis is prolonged and its motor activity — more intense.

The same authors describe findings in which, intra-operationem, mesenterial lymphadenitis is established and 2—4 months thereafter — follicular ileitis (duely diagnosed) and still later — chronich form of regional enteritis.

As early as 1939 Hadfield, and later Bousson, Chome, Mialaret and others (18, 8) related the acute forms of regional enteritis with Crohn's disease, assuming them as initial forms of the latter. The scheme suggested by Bousson casts greater clarity on the issue.

The problem of the role played by the appendix in the development of regional enteritis is rather obscure. The presence of a regional mesoadenitis concomitant the chronic appendicitis has been repeatedly proved. The changes in the appendix have been likewise proved during its removal for terminal enteritis, as in one of the patients of the present series. Bearing in mind its anatomical structure and draining lymph system as well as the favourable effect the appendectomy exerts on terminal enterites, the assumption seems warranted that the appendix has a definite bearing on the development of terminal enteritis.

Proceeding from the evolution of the anatomo-pathological process and clinical manifestations, the authors feel that it is reasonable to consider the morbid condition in question under two forms (phases): acute and chronic.

During the acute phase the presence is stressed of a submucous lymphoid hyperplasia in combination with trabecular sclerosis of the connective tissue and stasis within the mucosa and other layers and ganglionic hyperplasia as well. This form might be complicated by intestinal perforation or obstruction (5, 10, 17).

The chronic phase is characterized by a further evolution of this aspecific, inflammatory-degenerative process. The mucosa is enccmpassed by a diffuse edema and inflammatory granuloma, caused by plasmocytes, eosinophilic cells, never penetrating the submucous layer. Within the latter, massive sclerosis is detected with polymorphic cells of lymphocytic and reticular origin. The picture in this phase resembles that of cured ileocecal tuberculosis, although caseum is not present (Verges).

Nowadays, the hardly known syndrome of chronic intestinal obstruction, brought about by kinking of the intestinal segment under discussion and called kinking of Lane's band (Lane — 1911), is likewise assumed as a final outcome in terminal ileitis. It is furthermore stated that after subsidence of the inflammatory process in terminal ileitis, dense fibrous adhesions are formed, fixing the kinked and eventually narrowed intestinal segment against the abdominal wall. Similar adhesions with partial arrest of the passage were found during the operation of patient № 3 of the present series.

The diagnosis of both forms of regional enteritis is difficult. The symptomatology of the acute forms is identical to the picture of acute surgical abdomen and is usually proved on the operating table. The chronic form could be detected on the basis of some of its typical X-ray appearances: the sign of the chorda, of the basket, of pavement-like alternating hypotonic areas — all of them well known in roentgenology.

Treatment of regional enteritis is not definitely established and there are objections and criticisms directed to practically all methods applied in practice. Some authors claim that recurrences amount to 50% (Crohn), 65% (Pollock) and even 92% (Rohts), regardless of the method of management resorted to. In acute forms, with previously made diagnosis, a great many authorities recommend the adoption of a conservative policy: antibiotics, sulfanilamides, corticosteroids (17, 18).

The behaviour in acute forms of terminal ileitis, established during laparotomy, usually consists in appendectomy and infiltration of the involved mesenterial segment according to Forge and 20—40 cc 1% novocain solution. We practiced adding to the solution 400—600 000 U penicillin and 0.5 gr streptomycin. Some authors (Crohn, Pettinari and others) raise an objection against the appendectomy, expressing a groundless fear of fistulization etc. Falis, Helms, Grishkevich and the authors of the present article believe that appendectomy should not be discarded provided the regional enteritis is not distant from the ileocecal angle (2, 12). In destructive forms of the acute regional enteritis—perforation, phlegmon, necrosis—resection of the involved area until healthy tissue is reached is mandatory.

In chronic forms, originally, conservative treatment should be embarked on, as already stated, including a strict diet; such a policy proved effective in some cases. In most instances, anyway, operation should be undertaken, carrying out a bypass anastomosis or radical resection. The derivation procedure is simpler, but with it recurrences are rather frequent — 29% according to Colp, as in the last case of the series, subjected to ileo-transverso-latero-lateral anastomosis.

Resection of the affected region should be the method of choice in all instances of chronic regional enteritis — removal of 60 cm healthy intestine on each side, together with the corresponding portion of the mesen-

terium (Verges). In case economical resection is performed — up to 15 cm in healthy tissue (Deloyers H. Brener). In early resections, the incidence of recurrences, according to Pollock, amount to 12 per cent.

We carried out a follow-up study on five patients and we feel it is worthwhile presenting the case reports as they are of clinical interest, especially some selected cases.

Case Report I — K. I. K., male, aged 25 (history of illness 1562/1964). Complains of pains in the abdomen of one week duration, with loose bowel movements. Since two days the pains become stronger, he begins to throw up and the temperature rises up to 39°. On admission — palpatory painfulness in the ileocecal region with positive Blumberg symptom, without pronounced muscular tenderness (tension). Leukocytes 9800, hemoglobin 100%; in the urine — 8 —10 leukocytes and 2—3 erythrocytes. Emergency operation performed with preliminary diagnosis — acute appendicitis. The appendix is found slightly edematous and accordingly removed. The distal portion of the ileum, at a point distant 12—13 cm, is found edematized. The regional lymph nodes of the mesenterium are increased up to the size of a pea grain. The mesenterium is infiltrated with 200 cc solution of penicillin-novocain. Uneventful postoperative period and the patient is discharged on the 6th day — healthy. The histological investigation of the appendix reveals: chronic exacerbation of appendicitis with profuse lymphocytic infiltration of mucosa and fibrous degeneration of submucosa.

On the check-up examination one and a half years later, the patient has no complaints; laboratory investigations: within normal limits, X-ray: the intestine exhibits hastened up passage of contrast matter — after 4 hours it is found in the colon with no traces at all in the small intestine.

Case Report II — B. A. B., male, 9-year-old (history of illness № 10625/1959). Complains of vague pains in the abdomen with nausea dating 5—6 days ago. Since two days they become more intense and localized to the rightside, irradiating in the thigh. Temperature — 37,5°. On admission — palpatory pains in the ileocecal area with manifested Blumberg's, Rovsing's and Obraztzov's signs. Pulse rate — 110, leukocytes — 10.000. Emergency operation carried out with preliminary diagnosis — acute appendicitis. Profuse yellowish liquid discharged from the open abdomen. The appendix is found filled up of content, with irrigated vessels and edematized apical half. The distal portion of the small intestine, immediately beneath the ileocecal valve, is found with edematous wall over a length measuring about 10 cm, reddened, with irrigated vessels. The mesenterium of this intestinal segment is likewise edematous, with enlarged lymph nodes. Terminal ileitis is accepted, probably in connection with the acute appendicitis. Routine appendectomy is carried out and the mesenterium is infiltrated with 20 cc novocain-penicillin solution; 300 000 U penicillin and 0,5 gr streptomycin administered in addition into the peritoneal cavity.

Pastoperative period uneventful; the child receives 5 gr streptomycin and 3 000 000 U penicillin; discharged in good general condition. No complaints are recorded at the regularly carried out yearly prophylactic follow-up examinations.

Case Report III — B. T. M., a 30-year-old Lebanese is concerned with history of illness No. 14981/1959. Admitted at the internal ward a week ago with preliminary diagnosis — gastric ulcer. On the afternoon of the day he was referred to the surgical clinica, he sustains severe pains in the abdomen, with onset during the previous night. A wait-and-see policy adopted for several hours, originally for suspected perforating ulcer and thereupon — for acute appendicitis. It is decided to carry out appendectomy under conditions of pronounced Blumberg's, Rovsing's and Obraztzov's signs, pulse rate 110 per min, 16 500 leukocytes and temperature — 37,2°.

During the operation, the appendix is found moderately irrigated, 7—8 cm long, with pronounced hyperemia of the vessels, warranting completion of the appendectomy. The distal portion of the small intestine, at a point distant 10—12 cm, is found with strongly hyperemized vessels, edematous and reddened; it is adherent to the mesenterium and to the posterior abdominal wall, accounting for a relative impatency. Terminal ileitis is accepted, the adhesions — relieved and the mesenterium of the intestinal segment — infiltrated with 60 cc novocain-penicillin solution; in addition, 1 gr streptomycin and 600 000 U penicillin introduced into the peritoneal cavity. The postoperative period is supervised jointly with a therapeutist due to concomitant bronchopneumonia. The patient receives large quantities antibiotics, cardiotonics and vitamins. Discharged without abdominal complaints and referred to a rehabilitation unit. The investigation of the appendix reveals chronic exacerbated appendicitis.

Case Report IV — H. A. H., male, aged 32 years (history of illness № 3665/1964). Two months ago, following ingestion of a peppery food, he sustains severe abdominal pains; after several days they become moderate. Two-three hours after eating a meal, he receives nausea with frequent vomiting which accounted for relief of symptoms. During the past two months, recrudescence of pains occurs, becoming permanent, with sweating at night, frequent succussion sounds; unshaped stools are passed 2—3 times daily. He looses weight (5 kg). Past history — in childhood he complained of abdominal pains very often; 8 years ago he fell ill with grippe accompanied by pains in the abdomen. On admission — tongue furred, palpatory painfulness in the ileocecal region; by palpation of the abdomen the presence of a dense mass is discovered, vaguely outlined, without distension of the abdominal wall. Leukocytes — 9400, Hb 90%, urine — within normal limits.

With diagnosis chronic exacerbated appendicitis, the patient is subjected to operation on the day of admission. The appendix is with no visible changes, the cecum — a compact, tumorous-like mass, the size of a child's fist, with thickened wall and pasty consistency, purple red. The terminal portion of the ileum, at a point 14—15 cm from the cecum, is likewise edematized, with purple-red colour, compact and with pasty consistency. The proximal portion of the intestine is moderately dilated and the peristalsis therefrom is not conferred to the terminal ileum. The corresponding mesenterial segment is edematous and the lymph nodes within the latter — enlarged, ranging from a pea grain to a cherry. Appendectomy is carried out and a lymph node obtained for biopsy. Bypass anastomosis is embarked on in view to the presence of chronic regional ileocecal enteritis

and progressively narrowing lumen, leading to intestinal obstruction (the onset of the latter coincides with the clinical exacerbation described). A latero-lateral ileo-transversostomy is carried out for the anastomosis, distant 50 cm from the ileocecal valve. The postoperative period is uneventful and the patient is discharged in good general condition, liable to further out-patient follow-up examinations.

The histologic investigation of the lymph node reveals diffuse reticular hyperplasia with pronounced fibrous alteration of the reticulum, hyperplasia of the lymph follicles with profuse outgrowth of their germinative centers; the nodular capsule is thickened and fibrous. It exhibits total obliteration of the lumen with copious growth of aspecific, granulating tissue, the mucosa is indiscernible, the submucosa and muscularis are fibrously changed (M. Gardevski).

On the follow-up examination after 3 months, the patient complains of malaise, poor apetite, sporadic pains in the abdomen and movement of bowels 2—3 times daily (stools unshaped). Reduced sexual potency. Laboratory investigations: within normal limits. On the check-up X-ray investigation, the large intestine, the cecum excluded, is filled up by means of enema with passage of the contrast matter through the anastomosis created. On tracing up the passage of the contrast matter received per os, a permanent stasis (retention) is established in the terminal ileum. The lumen of the latter in the vicinity of the ileocecal valve is normal whereas, proximally — it is dilated. A slight defect in filling up is noted in the cecum. With a view to the persistence of the process, radical surgery was proposed to the patient, but he refused.

Ileocecal localization of regional enteritis is very rare. According to Demole (11) up to 1959, nine cases in all have been published.

Case Report V — J. D. J., female, aged 55 (history of illness № 2316/1966). The illness dates back 4 months ago when she feels acute low abdominal pains, with frequent appeals for defecation and liquid stools. Thereafter, the pains become intermittent and she is subjected to treatment for colitis. Owing to failure of the latter, she is referred to the hospital with suspected abdominal tumour. On admission, the general condition is good and by palpation of the ileocecal area, the presence of a hard and fixed mass is discovered, with longitudinal form, along the length of the ascending colon, palpatorily painful. Laboratory investigations of blood and urine reveal insignificant deviation, whereas the X-ray examination demonstrates tumour of the cecum, very likely of malignant nature.

With a preoperative diagnosis — carcinoma of the cecum — she is referred for surgery. During the operation, carried out under intubation (endotracheal) anesthesia and rightside laparotomy, the cecum is exposed and found encompassed by a tumorous mass, constricting (stenosing) the lumen, with greatly enlarged lymph nodes in the mesenterium. Aiming its removal (of the neoplasm), the cecum and colon ascendens up to the middle of the transverse colon are liberated; the terminal portion of the small intestine and the middle portion of the transverse colon are resected performing termino-terminal anastomosis between the resected ileum and the resected colon. Uneventful postoperative period and the patient is discharged healthy in two weeks. She has no complaints at all on the follow-up examination. The histologic investigation of the preparation reveals no

evidence for malignancy. Within the thickened wall a connective tissue inflammatory infiltrate is detected displaying mainly perivascular disposition, consisting of eosinophilic leukocytes, lymphocytes and plasmatic cells. In certain areas, the infiltrates are grouped in the form of lymphatic follicles with differentiation of terminal centers therein. Numerous dilated lymph vessels, filled up with homogenous pale pink tissue are encountered among the connective tissues. Strongly pronounced follicular hyperplasia with growth of germinative centers is also observed.

The histologic changes correspond to the picture of chronic, fibrous, productive proliferative, inflammatory processes (M. Gardevski).

Inferences

1. Regional enteritis as a distinct clinical unit already assumes an important role in acute abdomen conditions.
2. Of the numerous etiopathogenetic theories proposed, the transformation of mesenterial lymphadenitis in acute (follicular enteritis of Bousson) and chronic forms of the Crohn's disease is worth of special mentioning.
3. The symptomatic complex of intestinal incompetence — Lane's kinking — should be assumed occasionally as a sequel of terminal ileitis.
4. It could be assumed that the appendix has a definite bearing on the development of terminal ileitis.
5. The case material reported illustrates the diagnostical difficulties in acute (first two cases) and chronic forms in which diagnosis „chronic form" has not been established (observations III and IV).
6. The only reasonable treatment of chronic forms is early radical resection; appendectomy should be also included in the complex surgical management of all forms.

REFERENCES

1. Аминев. — *Вестн. хир.*, 1961, 2.
2. Бурба, В. Е. — *Вестн. хир.*, 1957, 2.
3. Гришкевич, В. М. *Вест. хир.*, 1962, 9.
4. Дереджан, А. — *Хирургия*, 1955, 2.
5. Калев, Н. — *Хирургия*, 1962, 9.
6. Ленда, Б. А. — *Вестн. хир.*, 1958, 5.
7. Петков, П. — *Хирургия*, 1960, 11.

К ВОПРОСУ О РЕГИОНАРНЫХ ЕНТЕРИТАХ

П. Алтынков, П. Червенков, Л. Господинов

РЕЗЮМЕ

Указывается на клиническое значение регионарных ентеритов, в особенности для спешной хирургии. Обращается внимание на теорию о переходе мезентериальной лимфопатии в острые и хронические формы заболевания, как самую правдоподобную их этиопатогенеза. Приведенные лучаи (4) являются подкреплением указанной теории.