Idiopathic muscular strictures of the sigmoid colon

C. CASSANO AND A. TORSOLI

From the Istituto di Clinica Medica Generale (II) dell' Università, Rome (Italy)

In a paper dealing with diverticular disease, Morson (1963) stated that four out of 155 specimens of sigmoid colon removed at St. Mark's Hospital, London, for 'diverticulitis' showed an increase of the muscular thickness of the wall without any detectable diverticula. There were no signs of inflammation in the wall, and the muscular abnormality could be compared to that seen in patients with diverticulosis. From this observation, he suggested that the muscular thickening could precede the appearance of diverticula or predispose to the occurrence of diverticulosis. This hypothesis seems to have been confirmed by Arfwidsson (1964) who found initial intramural diverticula in cases of spastic colon with remarkable thickening of the musculature. The muscular involvement of the colon in diverticulitis has also been pointed out by Celio (1952) who compared the myopathic component of the disease to the hypertrophic stenosis of the pylorus.

Idiopathic muscular strictures of the large bowel in human beings have not been described until now. In the horse, segmental muscular stenosis has been mentioned in the oesophagus and small intestine (Helmke, 1939). We have observed two cases of muscular stricture of the sigmoid not related to diverticular or other diseases. It seems helpful to give a brief description of our experience.

CASE REPORTS

CASE 1 This is a necropsy study of a 21-year-old woman who died of renal failure. She had never complained of digestive trouble; her bowels had always been regular.

After the necropsy the large bowel was fixed in dissection with 10% saline formaldehyde at a pressure of 70 cm of water. After fixation, radiographs were taken with air contrast, and transverse sections of the large bowel, embedded in paraffin and sectioned at 5 microns, were stained with haematoxilyn and eosin, Mallory, and Pas.

Morbid anatomy All the large intestine from the ileo-caecal valve to the anal canal was studied. The outside of the fixed specimen showed a stricture in the sigmoid which was harder and more fibrous than the rest of the large bowel. This narrowing was also evident in the air-contrast film (Fig. 1a).

On opening the specimen, the caecum, right transverse, left colon, and the rectum were normal. The sigmoid showed reduction of the lumen in its entire length except at one point where a circumscribed dilatation of the wall was seen. The mucosa had redundant folds with a longitudinal pattern (Fig. 1b); the walls showed a considerable increase in the thickness of the muscular layer except at the dilatation where it was thinner than the surrounding parts (Figs. 1a, 1c).

Microscopically (Figs. 1c, 2a, 3a) the mucosa and the submucosa were normal and showed some signs of

FIG. 1a. Case 1. X-ray air-contrast picture of the isolated colon, showing the normal aspect of the ascending tract and the stricture of the sigmoid with remarkable thickening of the wall: there is a break between the stenotic sigmoid and the normal rectum and at about the middle of the sigmoid a circumscribed dilatation of the wall.

FIG. 1b. Macroscopic appearance of the specimen between the normal descending colon and the narrowed sigmoid: in the narrowed sigmoid, thickening of the wall and many mucosal folds with a longitudinal pattern.

FIG. 1c. Transverse sections of the sigmoid colon (4×) at the level of the dilatation of the wall. The thinning of the longitudinal musculature and the almost complete disappearance of the circular musculature are noted. Also the mucosal folds are missing at this level.

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vascular congestion and a few focal collections of neutrophilic granulocytes. Thickening of both circular and longitudinal musculature could be easily recognized and was more evident in the inner layer. The musculature appeared to be normal in structure, but in some sections oedema was evident. The ganglions of Auerbach's plexus seemed to be cytologically normal and more numerous in the narrowed portion of the bowel than in those adjacent. The serous membrane showed a normal aspect.

At the level of the dilatation of the wall (Fig. 1c), the mucosa was flattened, the mucosal folds had disappeared and the muscular layer was thin. The dilatation involved about one third of the entire circumference and its radius was wider than that of the common diverticula. The picture resembled in some way the sacculations that at times can be seen in the sclerodermic colon.

CASE 2 A 53-year-old woman had suffered from constipation from the age of 13. At first the bowels were open at intervals of a few days, then at longer periods, till she had to use laxatives and enemas. Nevertheless she had stopped using laxatives during the last few years because they were no longer effective. Lately the constipation had become worse and she began to suffer from a tight, painful sensation in the left hypochondrium. This seemed apparently related to the intestinal stasis. She complained also of headache, abdominal distension, and listlessness; all this had worsened in the last two years.

An X-ray examination of the large intestine taken about 10 years ago revealed that the calibre of the sigmoid was slightly reduced. However, there were no detectable changes in the wall.

Physical examination showed a fit woman with a normal abdomen. The colon, increased in consistency and slightly painful, was felt in the left iliac fossa. The laboratory tests did not show any abnormal pattern. Proctosigmoidoscopy was also normal.

The gastrointestinal transit time, studied by means of radiopaque markers (Halls, 1965; Hinton, 1967), revealed that after 10 days all the markers were still retained in the large intestine; the greater number seemed to be situated in the left colon above the sigmoid (Fig. 4a). X-ray examination of the large intestine by double-contrast enema (Figs. 4b, 5a, b) showed a segmental stricture in the descending and sigmoid colon and the rectosigmoid junction. The change of luminal calibre was gradual, cranially, but clear cut at the distal end. The outline of the wall appeared to be normal everywhere and also in the stenotic tract.

A provisional diagnosis of muscular stricture of the sigmoid colon was made. It was believed that this stricture could be the cause of the constipation and surgical treatment was suggested.

At laparotomy the sigmoid appeared normal, with a slightly reduced calibre. A left hemicolectomy was performed, and the surgical specimens were fixed according to the procedure described in the previous case. After fixing the sigmoid was found to be stenotic; it felt harder than the descending colon.

Upon opening the specimen a stricture of the sigmoid was evident and the radiological changes of intestinal calibre, gradual in the upper end and clear cut in the lower one, were confirmed. A noticeable corrugation of the mucosa was seen, the folds having sometimes no definite orientation, sometimes a transverse direction (Fig. 6). Thickening of the muscular layer was seen
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(Fig. 3c). Microscopically (Figs. 7 and 8) mild congestion of the mucosa and submucosa was seen. The muscularis mucosae seemed slightly thicker, with an irregular pattern of its muscular cells. The circular muscle was remarkably thickened, while the taeniae appeared to be quite normal. No diverticula were found. The ganglia of the myenteric plexus appeared to be numerous and more voluminous in the stenotic portion, but were structurally normal. No changes in the serosa were noted. No part of the wall components showed signs of inflammation or degeneration.

After the operation the patient’s symptoms disappeared and her bowel movements returned to normal.

DISCUSSION

Our cases show a hitherto unknown muscular involvement of the sigmoid colon. The most important finding is the considerable increase in thickness of the musculature with a consequent remarkable reduction in calibre of the damaged segment. In
FIG. 4a. Case 2. X-ray film 10 days after the administration of radiopaque markers which are still retained in the large intestine; most of them are situated above the sigmoid.

FIG. 4b. X-ray film during the first phase of a double-contrast enema; the stricture of the sigmoid is clearly visible.

FIGS. 5a - 5b. Case 2. X-ray films after gas insufflation (double-contrast enema). The stricture begins from the descending colon to the sigmoid and ends abruptly at the wall of the rectum. The outlines and the inner surface of the sigmoid are regular.
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FIG. 6a. Case 2. Macroscopic appearance of the isolated distal colon. The corrugation of the mucosa and the development of the mucosal folds are noted. The increase in thickness of the wall is more evident at the higher magnification.

FIG. 7a. Case 2. Transverse sections of the sigmoid colon. Fig. 7a shows the normal aspect (×50) of the mucosa, the slight increase in thickness of the submucosa and the thickening of the circular musculature.

FIG. 7b. An increase in number of Auerbach's ganglions is also evident (×50).
the first case, the thickening involved both muscular layers and it appeared to be asymptomatic. Some focal inflammatory changes of the mucosa and submucosa and furthermore a circumscribed dilatation of the wall were noted. In the second case, characterized by severe constipation, the muscular abnormality involved only the circular layer. This was not accompanied by inflammatory or degenerative change. After surgery (left hemicolecction) the bowel habit returned to normal.

A similarity exists between the muscular changes in these two cases and the changes of the muscle in diverticular disease. Nevertheless, it is impossible to say whether or not there is a relationship between the two clinicopathological pictures. In diverticular disease the increase in thickness of the muscles seems related to the shortening of the intestine (Morson, 1963), while, in at least one of these cases, there was no indication that the sigmoid was shortened. The almost normal aspect of the sigmoid upon removal and the reproduction of the stricture after distension and fixing are proof that the muscular thickening can have an important functional component. A pharmacological effect of saline formaldehyde could possibly have contributed to the recurrence of the stricture (Grupp and Goerttler, 1957).

It should be stressed that in both cases the stricture was localized in the sigmoid colon, that is, the colonic segment which normally has the thickest muscular wall and in which mechanical activity is most intense (Cassano, Torsoli et al., 1966).

These strictures could be defined as 'idiopathic' in order to point out the unknown aetiolog and their presumable autonomous meaning; but further studies are necessary to elucidate the background of the pathological picture. It is probable that they are not often exactly recognized, the diagnosis in most cases being that of simple chronic constipation, diverticulitis, sigmoiditis, or cancer.

SUMMARY

Two cases of muscular stricture of the sigmoid colon, apparently idiopathic, have been described. In the first case the increased thickness of the muscle was asymptomatic and involved both the circular and longitudinal layer. It was accompanied by some scattered inflammatory changes. In the second case, characterized by severe constipation, the muscular thickening concerned only the circular layer and there were no signs of inflammation. After a left hemicolecctomy the constipation disappeared.

Some similarities between this pathological picture and that of diverticular disease are mentioned. The need for further studies to elucidate the meaning of these muscular thickenings has been stressed.

REFERENCES


ADDENDUM

Since this article was written, we have seen two new cases of sigmoid muscular stricture. In both the
pathological findings were strictly comparable with those described above (severe thickening of both muscular layers without evidence of inflammation and the ganglia of Auerbach's plexus appearing to be normal). The first case concerned a 53-year-old man with longstanding constipation; in the second patient, a 35-year-old man who was operated upon for a subacute intestinal obstruction, a small diverticulum was found in the thickened muscular wall.

FIGS. 1-3. Pathological appearances in a man aged 53 with longstanding constipation.