SARCOMA OF THE SMALL INTESTINE.*

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THE clinical and pathological aspects of sarcoma of the small intestine have been thoroughly reviewed in the comprehensive papers of Baltzer, Rheinwald, Lecene and Libman. While little can be added to their conclusions, the writer desires to report two new cases and to summarize the results of operation in the large number of cases which are now on record.

A statistical review of sarcoma of the intestine proves the rarity of the affection, as Baltzer in 1894 was able to collect 14 cases, Libman 59 cases in 1900, and Lecene 89 cases in 1904. The autopsy records in various large hospitals also confirm the view that sarcoma is infrequent in the intestinal tract, especially when compared with carcinoma. Nothnagel found 243 instances of carcinoma of the intestine in 2124 autopsies on cancer cases, while of 243 sarcomata but three were in the bowel. Smoler in 13,036 autopsies found 13 cases of primary sarcoma of the small intestine. Sarcomata of the large intestine, excluding the rectum, are much less common. Of Krueger's cases, 16 occurred in the small intestine, 6 in the large intestine and 16 in the rectum. Jopson and White, in 1901, found 22 cases of the large intestine, whereas Libman's paper appearing a year earlier contained 59 cases of sarcoma of the small intestine.

Sarcoma of the small intestine does not appear to affect any particular age, although Baltzer found that the majority of his cases occurred in the fourth decade. The 75 cases in which the age is mentioned may be divided as follows: 1–10, nine; 10–20, ten; 20–30, seventeen; 30–40, eighteen; 40–50, fourteen; 50–60, five; 60–70, two.

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The rather large number of cases occurring at an early age is a fact of much interest. The tumor in Stern's case was present at birth and caused intestinal obstruction from which the child died. In addition to this instance, sarcoma of the intestine has been observed in children of five and six years of age for which successful operations have been performed (Power, Barling, Zwalenburg).

Any portion of the small intestine may be the seat of a primary sarcoma. The following is an analysis of 53 cases in which the part involved is mentioned. As many of the case reports merely state that resection of the small intestine was performed, they could not be included. Duodenum and jejunum, 3; jejunum, 12; jejunum and ileum, 2; ileum, 32; entire intestinal tract, 4.

All writers on the subject mention the predisposition of the male sex in intestinal sarcoma. Adding the cases which I have collected to Lecene's we find of 101 instances, 67 occurred in males and 34 in females, or practically twice as many in the male sex.

As lymphosarcoma constitutes one of the chief types of intestinal sarcoma and as such growths tend to spread early to the neighboring lymphatic nodes, the mesentery of that portion of a bowel in which the sarcoma arises is involved frequently. In 45 autopsies 34 (75 per cent.) instances of mesenteric involvement are recorded by Lecene, a fact demonstrating the importance of thorough removal of the mesentery of the affected bowel. On the other hand metastasis to the superficial lymph-nodes or those in the retroperitoneum or mediastinum is rare.

Involvement of practically all the abdominal viscera has been noted in advanced cases, although the liver and kidney are especially liable to metastatic deposits. Direct extension to the peritoneum of adjacent viscera is quite common, and at the time of operation several loops of gut may require resection. Involvement of the bladder is met with frequently because the tumor in many cases occupies a pelvic position.

The histological variety of sarcoma is of great interest in

connection with the question of metastasis. The majority of recurrences or metastases have arisen in lymphosarcoma or in the round-cell variety. The spindle-cell sarcoma, on the other hand, has a pronounced tendency to remain localized. This fact is explained partly by reason of the stenotic action such tumors exert on the intestine, in consequence of which the indications for early operation arise before marked extension can occur.

The association of single traumatic insults has long been held important in the development of sarcomata in general. The numerous instances recorded by Coley, Lowenstein and others support this view. It is not surprising, therefore, that such a factor is mentioned in some of the reported cases and is of particular interest, as the disease occurs much oftener in the working class. Zwalenburg records an abdominal injury in a boy aged five; six weeks later a tumor one inch in diameter was noted at the site of injury. Nothnagel observed a case of lymphosarcoma developing on the base of an old tuberculous ulcer. The association of tuberculosis and lymphosarcoma elsewhere has been observed and is regarded as an accidental association. Three cases of sarcoma have been reported to have occurred in the ileum years after severe attacks of typhoid fever. Firth noted an instance developing five months after an operation for strangulated hernia. Syphilis has also been present in several cases.

From these factors of more or less etiological importance, we are unable to draw any conclusions which might throw light upon the cause of intestinal sarcoma.

Kasemeyer has investigated very thoroughly the subject of intussusception caused by tumors, and has collected 284 cases, of which 85, or 30 per cent., were caused by malignant formations. Of these 85 cases, 57 were carcinoma and 26 were sarcoma. The symptoms of intussusception as seen in children, the severe abdominal pain, vomiting, bloody and mucous stools, are seldom present in intussusception secondary to tumor formation. In such cases a chronic course is pursued and the symptoms extend over months even with an

intussusception present, as is demonstrated by the dense adhesions about the bowel or by extension of the invaginated tumor to the intestinal wall with which it comes in contact. The infrequency of complete obstruction following tumor intussusception is explained by the fact that the infiltrated intestinal wall undergoes dilatation.

Tenesmus may be the chief symptom complained of, but is as inconstant as is meteorismus and abdominal tenderness. The presence of a sausage shaped tumor, the situation of which varies, along with other symptoms of chronic intestinal obstruction, has been regarded as distinctive of tumor invagination by several observers, and the diagnosis correctly made (Ewald, Kasemeyer).

Many varieties of sarcomata have been observed in the intestine; the 99 cases in which the type is mentioned are divided as follows: Lymphosarcoma, 34; round-cell sarcoma, 43; spindle-cell sarcoma, 13; fibrosarcoma, 3; mixed-cell sarcoma, 1; myxosarcoma, 2; myosarcoma, 2; melanotic sarcoma, 1.

The lympho- and round-cell sarcomata greatly predominate. Many cases diagnosed as round-cell sarcoma probably belong to the lymphosarcoma group, but the histologic descriptions are too incomplete and indefinite in many cases to make the classification correct.

The tumors in the majority of cases originate in the submucous tissues (lymphosarcoma) or in the connective tissue of the muscularis or perivascular region, and in some instances reach a considerable size without producing any ulceration of the mucous membrane. They may extend parallel to the long axis of the bowel, producing a gradual infiltration of all the tissues but not causing stenosis. The bowel above the area of infiltration frequently undergoes dilatation and resembles an aneurism; the lumen of the intestine, in such cases, is filled with necrotic tumor tissue, pus and fecal material. Dilatation of the intestine is seen in the round-cell and lymphosarcomata, whereas stenosis and obstruction result from the fibrosarcomata. In exceptional cases the tumor extends

through all the coats of the gut, gradually involving neighboring coils and forming a large adherent mass. The tumor may be single or multiple; in the latter event the growths appear as plaques or small nodules under the mucosa. The single tumors, especially if pedunculated, are singularly prone to produce intussusception, although this complication has developed in the infiltrative types of tumor.

Marked variations exist in the size of the tumors, although as a rule the growth has reached considerable proportions before the diagnosis has been made or the operation performed. The shape is spindle, the contour irregular and the consistency firm in most cases.

Although partial occlusion of the bowel is present in about one-half of the cases complete stenosis practically never develops from the mere presence of the sarcoma. Even in large tumors encroaching upon the intestinal lumen, a narrow passageway can be demonstrated, thus explaining the chronic intermittent symptoms of intestinal obstruction. When complete occlusion occurs and is followed by the symptom of ileus, the condition is caused by adhesions or by an intussusception.

Sarcoma of the small intestine manifests itself in the beginning by symptoms of an indefinite nature. In the majority of patients generalized abdominal pain is first noted; this is followed by loss of appetite, nausea, vomiting, the bowels are irregular, diarrhœa alternates with constipation, and distention of the abdomen soon follows. The patients are very thin, pale and weak, when first seen. Moderate elevation of temperature and slight leucocytosis may be present. Unless the acute obstruction is due to kinking of the intestine or to an intussusception, complete constipation is unusual, although repeated attacks of obstinate constipation may be complained of. Baltzer and Nothnagel both asserted that apart from complications, sarcoma of the intestine does not produce symptoms of stenosis. This view has been disproved by subsequent articles, in which it has been shown that at least 55 per cent. of the cases do have symptoms indicative of some degree of in-

testinal obstruction, but the course is not similar to the stenosis caused by cancer of the bowel. When carcinoma produces an obstructive lesion, the course is generally a protracted one and the patient's loss of strength and weight is slow and gradual. Sarcoma, on the other hand, causes rapid loss of weight, the disease rarely lasting over a year and the average duration, according to Rheinwald, being four to five months.

A careful study of the histories of many cases shows that attacks of constipation and diarrhœa are common, although these symptoms are wanting in a small proportion of the cases. It is also worthy of note that in many instances vague intestinal disturbances are the earliest symptoms noted, and that operation performed a few weeks or months later will often reveal a larger or even inoperable sarcoma.

Blood in the stools has been present in a small proportion of the cases, and is sometimes one of the earliest symptoms mentioned.

In a few instances the patients have noted the presence of a tumor. This on examination varies considerably in size, the surface is smooth and nodular, and unless seen quite late, the growth is freely movable. Its consistency is as a rule dense and hard. In late cases metastatic nodules are palpable and the primary growth demonstrated with difficulty.

As the result of pressure of the tumor on the intestine, distention may result, and pressure on the vessels may produce ascites, or œdema of the legs, distention of the veins of the abdominal or thoracic walls, jaundice, dysuria or diminution in the amount of urine (Libman). Examination of the blood shows merely a secondary anæmia.

Libman has classified the varieties of the disease as follows: (1) Latent cases, the disease being first discovered at autopsy. (2) Cases with the clinical picture described by Baltzer, either the general symptoms, the distention of the abdomen, or the tumor being first noted. (3) Cases in which the first symptoms are due to an intussusception or other variety of intestinal obstruction or to perforation. (4) Cases resembling tuberculous peritonitis. (5) Cases in which jaun-

dice is the first symptom. (6) Cases resembling ovarian cysts. (7) Cases bearing a close resemblance to appendicitis, an observation noted first by Libman and described in several reports since that time.

An early diagnosis in these cases seems impossible because the symptoms are so mild and transitory in the beginning. When, however, a tumor is discovered, freely movable, producing pressure symptoms of a mild type, with the absence of severe obstruction symptoms, sarcoma of the small intestine should be suspected.

The treatment of intestinal sarcoma is of course surgical, although in inoperable lymphosarcomata benefit has been followed by the administration of arsenic. Libman recommended its use even in cases in which successful resection of the intestine has been performed.

For a long time sarcoma of the intestine was regarded as almost invariably fatal. This view is not sustained by an analysis of the cases reported in the past decade, in a large number of which many years have elapsed without recurrence since the time of operation. The vague nature of the symptoms delays operation, although a palpable tumor is almost invariably present at the time of operation and a history of a chronic intestinal disturbance can be obtained in the majority of cases.

The number of resections of the small intestine for sarcoma is 75; of these 15 are collected by Zwalenburg, 37 by Moynihan, 6 by Lecene, 17 by Speese. There were 55 recoveries (74 per cent.), and 19 deaths following operation. Nine instances of recurrence are noted, the periods varying from three months, 5 months (2), 12 months (2), 15 months. The cases in which recurrence arose in 7 instances were diagnosed as lymphosarcoma or round-cell sarcoma, thus emphasizing the malignant nature of this variety; one case of myxosarcoma recurred.

When the infiltration of the bowel is too extensive for removal or metastasis has occurred, the abdomen should be closed without further exploration. If stenosis is present some sur-

geons advise an artificial anus to relieve the immediate and urgent symptoms.

The large number of intussusceptions noted in the series is a matter of considerable interest and importance. In 14 of the 74 resections, this complication was encountered. Ten of these 14 cases recovered, 1 died immediately after operation, and 3 from recurrence. The type of tumor has no influence upon the development of an intussusception, for the complication has occurred in the round-cell, the lymphosarcoma and other forms. A pedunculated tumor may predispose to invagination, but it also follows cases in which the intestinal wall is extensively infiltrated by the tumor.

The amount of small intestine resected in the majority of cases is from 10 to 40 cm. Barclay removed 190 cm., and Storp 510 cm. of the bowel. In the former case the patient suffered from frequent and liquid stools, and in the latter no metabolic or other disturbances were noted.

The effect of the removal of large amounts of small intestine has been investigated experimentally by Flint, whose conclusions are of great importance in view of the radical measures which may have to be undertaken in some of the cases. It was found that in dogs as much as 50 per cent. of the total intestine may be removed without fatal results, and the animals may gradually return to a condition of practically normal weight and metabolism when maintained on a favorable diet under good conditions. Resections of 75 per cent. or more of small intestine may be survived, but such animals do not show a return to normal weight with the establishment of a good compensatory process.

Animals at first suffer from a severe diarrhœa, ravenous thirst and appetite, and loss of weight, from which they gradually recover until conditions may return to those of a normal animal. They remain extremely sensitive to unfavorable conditions of diet and living.

The compensatory process consists in a hypertrophy and hyperplasia of the remaining portion of the small intestine. There is no regeneration of villi or crypts.

Human cases behave in general like animals and show similar metabolic disturbances. There are over 58 cases in the literature in which over 200 cm. of small gut have been resected. The mortality is 16 per cent., which is lower than it should be, as only the successful cases have probably been reported. Metabolic disturbances in human beings bear no definite relationship to the amount of small intestine resected.

Resection of over 400 cm. of intestine has been followed by recovery, while death from inanition has resulted from resection of 284, 289, 300, 380 cm. respectively.

Profound digestive disturbances have resulted from removal of 192 and 204 cm. of ileum.

Progress in human cases should be guarded. Apparently successful resection may, for lack of suitable compensation, succumb ultimately to a slow process of inanition. Experiments and series of human cases emphasize the fact that neither the stomach nor the colon is able to compensate for the loss of large portions of small gut.

The writer desires to express his thanks for permission to report the following cases, operated upon by Dr. John B. Deaver at the University Hospital.

Male, aged fifty, has been suffering with hemorrhoids for several years and for the past several weeks has complained of constipation, distention of the abdomen, severe cramps and vomiting. The constipation was relieved by enemas and laxatives, the resulting movements were as black as ink, although free blood was not noticed. He has had successive attacks of pain, tenderness and obstinate constipation. The mass was not discovered until the time of examination, seven weeks after his symptoms began. The examination disclosed a round mass in the right lower quadrant of the abdomen. The tumor is tender, regular in outline, and is movable.

Blood examination, red blood cells 4,980,000, polynuclears 70, white blood cells 9,800, lymphocytes 20, hæmoglobin 100, monoleucocytes 0, transitionals 1, eosin 0.

Examination of the fæces for occult blood was negative.

Operation.—A large mass about the size of an orange was

found in the ileum about 3 feet from the ileocæcal junction. The ileum was resected with its corresponding portion of mesentery, and end-to-end anastomosis was performed. Five days after the operation the patient developed a fecal fistula; this was followed by peritonitis, from which he succumbed eleven days after the operation.

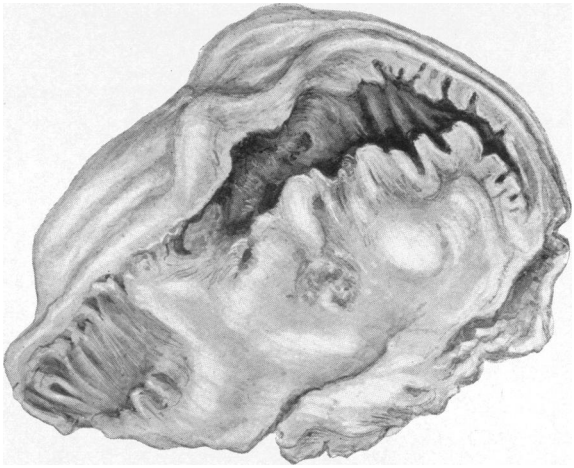
Pathological Examination.—The specimen consists of 57 cm. of ileum. The intestine at one area contains a globular mass 8 cm. in diameter, the wall of the intestine is enormously thickened, measuring 3 cm. The section through this thickened portion shows that the intestinal mucosa is greatly ulcerated and that the lumen of the bowel is represented merely by an irregular area of ulceration through the centre of the tumor mass. At one point the lumen is almost completely occluded by the tumor tissue (Fig. 1). The tumor mass, as represented by the greatly infiltrated wall of the intestine, is composed of firm whitish tissue which is completely surrounded by the serous coat of the intestine. In the mesentery several enlarged nodes having the same characteristics as the primary tumor are found.

Microscopic examination shows a very cellular formation composed of small, round, deeply staining cells, having a fairly uniform appearance. The stroma is composed of thin fibrils which ramify between the tumor cells, which extend to the mucosa and infiltrate and destroy the intestinal glands. The structure of the muscular coats of the intestine is completely obliterated by the cellular infiltration. The tumor contains very minute areas of necrosis and is fairly well supplied with new blood-vessels. The lymph-nodes removed from the mesentery show a similar involvement.

Diagnosis.—Lymphosarcoma.

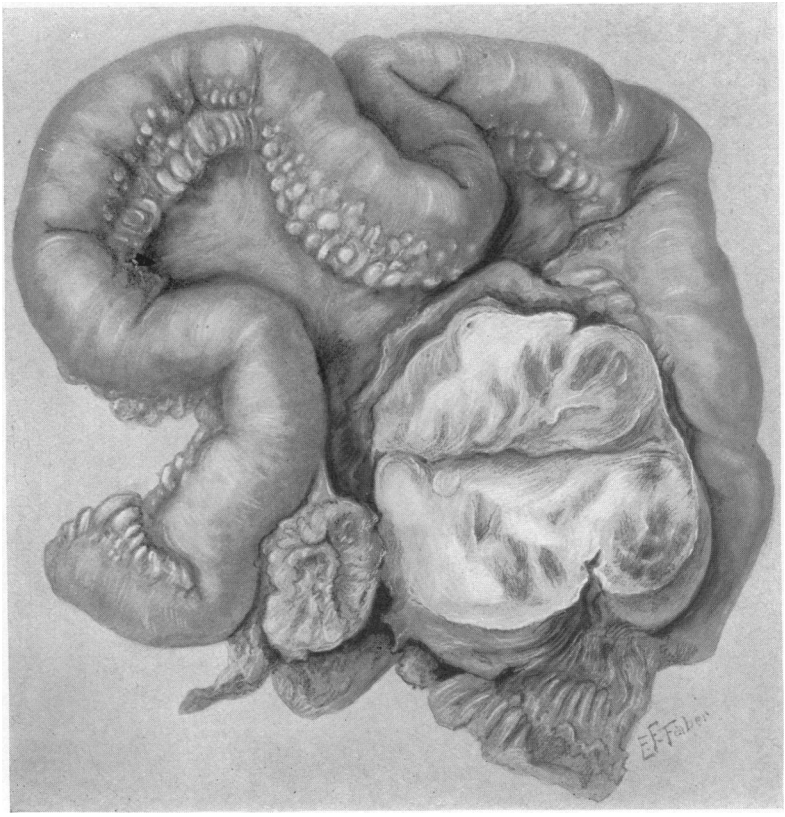
Female, aged fifty-seven, was admitted to the University Hospital complaining of pain in the abdomen. Her past medical history is unimportant. One sister died of cancer of the stomach. Her present illness began one month before her admittance, when she was suddenly seized with agonizing pain in the abdomen. The pain was localized to the region of the umbilicus; the attacks were accompanied by vomiting. The attack lasted twenty-nine hours. The patient recovered and was well for a period of three weeks, when the pain again returned. The pain has been persistent, is constantly localized to the region of the umbilicus; the bowels are regular; there has been some distention of the abdomen. On examination a mass the size of a grape fruit is palpable in the lower and middle portion of the abdomen. The upper limit of the tumor is about one inch below the umbilicus.

FIG. 1.



Lymphosarcoma of intestine showing partial occlusion of the lumen.

FIG. 2.



Myxosarcoma of mesentery.

The mass is smooth, round and slightly movable. Red blood cells 3,710,000, polynuclears 70, white blood cells 20,000, lymphocytes 23, hæmoglobin 60, monoleucocytes, 3, transitionals 4, eosin 0.

Operation.—On opening the abdomen a mass was found in the mesentery, in the midline; the surrounding coils of intestine were attached to it by adhesions. The coil of ileum which surrounded the tumor and the mesentery were excised and a lateral anastomosis formed. A supravaginal hysterectomy was performed for a large subserous fibroid tumor. Recovery; no evidence of recurrence three months after operation.

Pathological Examination.—The specimen consists of a tumor which is surrounded by a loop of small intestine, which measures 80 cm. in length. The tumor, which measures 8 cm. in diameter, is situated near the base of the mesentery and is attached to the intestine for a distance of a few centimetres only. The wall of the intestine appears normal and is not compressed by the tumor mass. On cross section the tumor is soft in consistency, the cut surface for the most part is white and contains numerous reddish areas and small points of necrosis.

On microscopic examination the growth for the most part is composed of tissue containing large stellate cells. The connective tissue in these areas is of very loose texture, and contains within its meshes a homogeneous substance taking a faint blue stain. Large numbers of blood-vessels with thin walls are present. A considerable amount of free blood is found in the fibrous tissue. In addition to the stellate cells mentioned, there are many areas in which large numbers of cells are closely packed together, the cells being spindle in type, some are large, some small and many being arranged around the blood-vessels. Minute areas of necrosis are encountered, and in these situations leucocytes are found between the tumor cells. Many nonstriated muscle fibres are seen in the more superficial portions of the tumor.

Diagnosis.—Myxosarcoma of mesentery.

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