LYMPHOSARCOMA OF THE GASTRO-INTESTINAL TRACT

WITH A REPORT OF THIRTEEN CASES *

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Lymphosarcoma of the gastro-intestinal tract cannot be considered as rare in occurrence, still it is so uncommon that isolated case reports are constantly appearing in the literature. There is wide variation in opinion regarding this condition, especially concerning the classification of various forms and its relation to other types of lymphatic tumors. Minot and Isaacs [1] have suggested the use of the terms lymphoblastoma or malignant lymphoma to include all types of malignant lymphatic tumors, and others have adopted the same nomenclature. Under this name they include (1) lymphatic leukæmia, (2) pseudo-or aleukæmic lymphatic leukæmia, (3) Hodgkin's disease, and (4) lymphosarcoma. There are considerable grounds for this classification as Ewing,[2] in considering the etiology, says: "It seems possible that in tuberculosis may eventually be found a connecting link between lymphæmia, pseudoleukæmia, some cases of Hodgkin's disease and some forms of lymphosarcoma." However, due to this variation in terminology, and also to the fact that many cases are reported either with no or only partial microscopic diagnosis, a complete tabulation with resultant conclusions is impossible. Many fibromyosarcomas are erroneously classed with the lymphatic tumors. We have two such cases, a brief résumé of which is given later.

Ewing [2] describes lymphosarcoma as a true malignant neoplasm arising in lymphatic tissue from proliferation of typical lymphocytes, and occurring as either a localized or diffuse process. In the stomach sarcomas constitute about 1 per cent. of all gastric tumors, most of which are either spindle-celled myosarcomas or lymphosarcomas. In the intestines sarcomas are relatively much more common. According to Liu [3] lymphoblastomas are the commonest tumors found in the small intestine, occurring three times as often as carcinomas. They may occur in any part of the intestine, but are most often found in the terminal portion of the ileum. They may be single or multiple. While rare in the appendix, Friend,[4] in 1926, found nineteen cases reported and added one of his own.

Sarcomas of the gastro-intestinal tract may occur at any age, but are found most frequently between the ages of twenty-five and forty-five.

Rankin [5] gives the sex occurrence as equal, but my own studies of the reported cases lead me to agree with Liu [3] that males are afflicted twice as commonly as females.

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Practically nothing is known regarding the etiology of these tumors. The theory of growth from embryonal cell groups has many supporters. Others feel that added to this there must be some exciting cause, such as tubercular infection. Certainly tuberculosis seems closely associated with sarcomas in many cases. Peterson [6] in a compilation of eighty-five cases found three where the tumors occurred six to ten weeks after a severe contusion of the region.

The symptoms presented by sarcoma of the stomach are those presented by any other tumor and vary with the size and location. As a rule gastric dilatation is not so marked as in carcinoma, and the tumor is apt to be larger and therefore more easily palpable.

In sarcoma of the intestines the one practically constant symptom is pain, generally of a colicky nature, occurring as a rule throughout the middle of the abdomen and not affected by eating. There may be slight loss of weight and slight fever. Vomiting is not a prominent symptom as a rule. There may be a history of increasing constipation. While narrowing of the intestinal lumen is a common finding, Libman [7] in 1900 could find no case of complete stenosis reported and I have found none since. Obstruction may be caused by intussusception which occurs not infrequently. Next to pain the most common finding is the presence of a palpable tumor increasing fairly rapidly in size. In some cases this is the only complaint the patient has.

The diagnosis is very difficult. Presence of a palpable tumor taken in conjunction with röntgenological findings of a partial stenosis, and occurring in a young patient, may at times make it possible, but not as a general rule. A good proportion of these patients come to the surgeon with symptoms of an acute abdomen and the correct diagnosis is only made at operation.

The duration of the disease is variable. Here it is that a uniform nomenclature and more accurate microscopic diagnosis in reported cases would help to clarify our ideas. Fisher [8] makes the statement that most patients suffering from intestinal sarcoma die within nine months. This is a shorter period than given by most writers, the average being about eighteen months.

The prognosis is also variable and depends on the type of sarcoma and also the kind and time of treatment. Farr [9] says the outlook in sarcoma of the stomach is no worse than in carcinoma, while Graves [10] speaking of sarcoma of the intestines says: "Death is the usual outcome." Some few cures have been reported. Kapel [11] reported one of fifteen and another of nine years' duration. One of our cases is alive and well eight years after operation.

There is some difference of opinion regarding the proper treatment of this condition. We must remember that no one man or clinic group has had a sufficiently long series of cases upon which to base their opinions—so that both the quoted opinions of others and our own conclusions are subject to change by future experience.

Considering the treatment the cases naturally fall into two groups. First is the very small group where the diagnosis can be made before operation and where the tumor is apparently resectable. Should these cases be submitted to the relatively dangerous operation of resection (Kapel [11] in his

report of resection in sixty cases of sarcoma of the stomach gives an operative mortality of 18 per cent.), or should they be given deep X-ray therapy, which apparently is so efficacious, temporarily at least, in causing similar tumors in other locations to disappear? Crumston [12] in his article on sarcoma of the stomach gives the treatment as excision. Minot and Isaacs [1] in reporting a case of lymphosarcoma of the small intestine which lived seven years without recurrence and then died two years later, say that no case of that type has lived so long except following surgical removal. They favor surgery and radiation, though they go on to say: "Radiation does not influence importantly the duration, it alleviates symptoms, decreases size of lesions and promotes patient's efficiency." Holmes, Dunn and Camp [15] in their article on lymphoblastoma of the stomach say: "The localized type of this disease should respond well to irradiation therapy or to a combination of irradiation and surgery." Rankin,[5] while saying that treatment is unsatisfactory, thinks surgery seems to offer the best chance and adds that radium may give good results. Desjardins and Ford [14] say that radiation may or may not cause definite prolongation of life, but it may keep the disease under complete or partial control for varying periods. Soiland [15] is rather pessimistic regarding X-ray treatment. He says: "It appears that we may successfully destroy the altered lymph nodes in every other part of the anatomy, even including those of the mediastinum, but we find our Nemesis in the abdominal cavity."

In the larger group of cases where diagnosis is only made at operation, there seems to be practically a unanimity of opinion that resection should be performed if possible, later writers favoring post-operative X-ray treatment.

In our experience resection, when possible, is still the method of treatment and we feel rather skeptical regarding the value of radiation. Even though incomplete, resection will often greatly prolong the life of the patient. In one of our cases (Case VII) the operator had to cut through tumor tissue in the mesentery of the intestine and yet this patient was well for nearly three and one-half years and had no X-ray treatment. In Case XIII where radiation alone seems to have caused a cure, the diagnosis was never proven and a later review of the patient would seem to show that the condition was tuberculosis of the peritoneum rather than sarcoma.

Resection should be performed whenever possible and may be followed by radiation if deemed advisable. If, however, radiation causes unfavorable symptoms such as vomiting or gastro-intestinal upsets we consider radiation of such doubtful benefit that we do not hesitate to discontinue it.

The use of arsenic should be mentioned. The present status of this drug is well summed up by Desjardins [14]: "Arsenic was used and when large doses were given over a prolonged period did result in transient improvement. However, results were not altogether satisfactory and these drugs are now seldom employed."

A brief summary of our cases follows: (The first six cases were explored only, no attempt at complete resection being made.)

CASE I.—Male, thirty years of age. Pain in lower right quadrant, one hour after eating, for three months. Vomited frequently. Operation showed numerous masses in the small intestine with involvement of retroperitoneal nodes. Specimen showed lymphosarcoma. Died five months after operation.

CASE II.—Female, seven years of age. Symptoms of acute abdomen, tuberculous peritonitis being the first choice. Operation showed intussusception of small intestine caused by prolapse of a tumor the size of a walnut. Enteroanastomosis without resection of tumor. Twenty-six days later a foot of the intestine with the growth resected. Several similar masses were found throughout the intestine. Pathological report—lymphosarcoma. Died six weeks after second operation.

CASE III.—Male, thirty-two years of age. Pain of seven months' duration, extending from symphysis to umbilicus. Operation revealed a tumor of ileum thirty centimetres from iliocæcal valve, with similar tumors in other portions of the ileum. Enlargement of lymph nodes. Pathological report—lymphosarcoma. Died two and one-half years after operation.

CASE IV.—Male, thirty-seven years of age. Discomfort and cramps across upper abdomen for one month. Swelling of feet and distention of abdomen. Operation showed huge tumor mass in retroperitoneal tissues, with metastatic nodes in meso of small bowel and one large tumor in the ileum, probably metastatic. Pathological report—lymphosarcoma. Died one month after operation.

CASE V.—Male, thirty-three years of age. Pain in epigastrium, right upper quadrant and occasionally right lower quadrant for four months. Vomiting at irregular intervals for six weeks. Slight loss of weight. Operation—diffuse infiltration of the small intestine encroaching on the lumen and extending into the meso at several points. One mass the size of a fist. Unquestionably lymphosarcoma. Pathological report—material lost in process of embedding. Died four months after discharge.

CASE VI.—Male, sixty years of age. Pain in lower abdomen especially on left side for three months. Lost a little weight. Operation showed a large mass involving large intestine and ileum adherent in pelvis; non-resectable. Pathological report—lymphosarcoma. Died fifteenth post-operative day.

CASE VII.—Female, thirty-four years of age. Pain around umbilicus for eight months. Loss of weight and strength. Palpable tumor in left side of abdomen. Operation—mass ten centimetres by eight centimetres involving whole circumference of gut about five centimetres from duodenojejunal junction. One large node in mesentery of gut. Resection with end-to-end anastomosis. Pathological report—lymphosarcoma. Died three years seven months after operation.

CASE VIII.—Male, thirty-five years of age. Epigastric pain for six months. Thirty pounds loss of weight. Increasing constipation. Palpable tumor. Operation—three separate tumors of small intestine contained in the first two feet of jejunum. Resection with end-to-end anastomosis of jejunum and duodenum. Pathological report—lymphosarcoma. Unable to trace patient.

CASE IX.—Male, thirty-five years of age. Symptoms of partial intestinal obstruction for five weeks. X-rays showed obstruction in splenic colon. Operation showed constricting mass at splenic flexure. First stage Mikuliecz and cæcostomy done. Pathological report—lymphosarcoma. Died suddenly day after operation.

CASE X.—Female, eighty-four years of age. Tumor size of orange, constant in size, for five weeks, to right of umbilicus. Increasing constipation. Operation showed intussusception at iliocæcal valve caused by prolapse of tumor of ileum about four centimetres in diameter. First stage Mikuliecz operation done. Pathological report—lymphosarcoma. Died on fourth post-operative day.

CASE XI.—Male, twenty-four years of age. Recurring pain in right lower quadrant for eighteen months. Came to hospital with symptoms of acute appendicitis. Operation—diffuse tumor involving entire circumference of terminal four inches of ileum. Tri-

angular segment of enlarged nodes running beyond duodenum. Resection of terminal ileum and ascending colon together with nodes, with side-to-side anastomosis. Pathological report—lymphosarcoma. Uneventful operative recovery. Given deep X-ray therapy. Well one year post-operatively.

CASE XII.—Male, thirty-four years of age. Pain in epigastrium of six months' duration, coming on one day to one hour after eating. Lost twenty pounds weight. Operation—mass size of palm of hand in posterior surface of stomach with enlarged nodes. Distal one-third of stomach resected. Pathological report—lymphosarcoma. In excellent health nine years later.

CASE XIII.—Male, thirty-seven years of age. Severe pains about umbilicus for one month. Lost a little weight. Palpable mass in right side, deeply adherent. X-ray showed general limitation of lumen of small intestine. No operation as condition was deemed inoperable. Diagnosis thought to be sarcoma, though a note was made that there is a possible tuberculous lesion at the apices, so the diagnosis may be tuberculosis of peritoneum. No pathological report. Tumor mass gradually disappeared under X-ray therapy. Three and one-half years later was in excellent health.

Two cases of myosarcoma have been operated.

CASE I.—Male, twenty-eight years of age. Sudden sharp pain in epigastrium to left mid-line. Thought to be perforated gastric ulcer. Operation showed huge tumor on anterior wall of stomach which perforated at operation. Resection done. Pathological report—myosarcoma. Given X-ray therapy, but died eighteen months later.

CASE II.—Female, twenty-eight years of age. Always constipated. Nausea and cramps in abdomen for two weeks. Operation—intussusception due to prolapse of tumor of cæcum. Intussusception reduced and tumor which did not appear grossly malignant, with surrounding cæcal wall, resected. Pathological report—myosarcoma of local malignancy type. Excellent health ten and one-half years later.

RECAPITULATION

There were thirteen cases of lymphosarcoma, all but two proven by microscopical examination, one not operated and one where the material was lost. Ten were males, three were females. Ages range from seven to eighty-four; average thirty-five. Ten cases involved small intestine, two large intestine, and one stomach.

Twelve cases were followed, one being lost track of. Seven died within one year; one died at the end of two and one-half years; one died after three and one-half years; one not operated, so diagnosis not proven, is well three and one-half years later; one resected, is well one year later; one, stomach resected, is well nine years later.

SUMMARY

1. A uniform nomenclature and more careful microscopical diagnosis are needed to enlarge our knowledge of this condition.

2. Treatment is unsatisfactory. Resection when possible followed by X-ray therapy, seems to offer greatest chances for a cure.

I desire to express my appreciation to Dr. Charles L. Gibson for permission to publish these cases, all of which were operated on in his service at the New York Hospital.

Since this paper was written we have had another case of lymphosarcoma of the ileum which I feel deserves inclusion in that we were able to make a diagnosis from the history before operation.

A man of thirty-five years, who was operated upon five months previously for chronic appendicitis now complained of pain to the right of the umbilicus, coming on about two hours after meals with the sensation of the formation of a mass which could be palpated at times. After a few minutes this pain would disappear and there would be a feeling of the letting up of a constriction and the passage through of liquid could be either felt or heard or both.

From this rather typical history the diagnosis of lymphosarcoma was made. X-rays gave the impressions of adhesions in the region of the cæcum. Operation revealed extensive lymphosarcoma of the ileum with involvement of the lymph nodes of the mesentery. Twenty cubic centimetres of ileum were removed with end-to-end anastomosis.

Pathological report was lymphosarcoma. The patient is still in the hospital, although he has made an uneventful recovery and is to receive X-ray treatment.

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