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## LEIOMYOSARCOMA OF THE DUODENUM

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LEIOMYOSARCOMA OF THE DUODENUM was first reported by von Salis in 1920. Since that time 60 cases have been noted in the world literature. The last comprehensive study was presented by Weinstein and Roberts in 1953. Their collective review of 28 cases suggested a poor prognosis, particularly with respect to the operative mortality which was 50 per cent.

Since 1953 the rate of reporting leiomyosarcoma in this location has accelerated, and 29 additional cases have been documented. Information has accrued which suggests the need for reappraisal of the subject. It is clear, for example, that this lesion is not as rare as it was formerly thought to be. The risk of surgical therapy can no longer be viewed as excessive since there has been no operative mortality in the cases recorded since 1953. Data have accumulated on the prognosis after surgical therapy. Finally, studies correlating the pathologic anatomy and the roentgenographic appearance of the tumor have frequently made it possible to arrive at a correct diagnosis of this malignant lesion preoperatively.

In the present study, all previously reported leiomyosarcomas of the duodenum have been reviewed, bringing the total to 61 (Table I).

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Trends in the diagnosis, therapy, and prognosis have been analyzed in order to arrive at a clearer understanding of the surgical management of these tumors.

### INCIDENCE

Of the 46 cases in which sex was mentioned, 25 were female and 21 male. Age distribution ranged from 21 to 80 years. Peak incidence occurred in the 40 to 49 year age bracket (see Table I), about one decade earlier than for the more common malignant conditions of the gastrointestinal tract.

The relatively equal frequency of malignant as compared to benign smooth muscle tumors of the duodenum may be inferred from the statistics of Starr and Dockerty who reported 16 smooth muscle tumors, of which 9 were benign and 7 malignant. These figures are substantiated by the data of Griffin and Disch who were able to collect 41 cases of duodenal leiomyoma in 1957, at which time approximately 50 leiomyosarcomas had been recorded.

Leiomyosarcoma is relatively infrequent compared to adenocarcinoma of the duodenum. Brenner and Brown collected 474 carcinomas in 1955, at which time approximately 40 leiomyosarcomas had been reported. The ratio of myomatous to entodermal malignancy is thus about 1 to 12.

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TABLE I.—SUMMARY OF CASES

<i>Author and year reported</i>	<i>Location (part of duodenum)</i>	<i>Age, yrs. sex</i>	<i>Symptoms</i>	<i>Physical findings</i>	<i>Gastro-intestinal x-ray film</i>	<i>Gross pathology</i>	<i>Operation</i>	<i>Result</i>
von Salis (1920)	4	40 M	7 yrs., fever, constipation	Mass	Not stated	Size of baby's head; necrosis with enteric and parietal fistulas	4 laparotomies	Died 4 mos. postop.
Brdiczka (1931)	1	60 F	2 wks., pain and bleeding	Mass	Filling defect; displacement of stomach and duodenum	Size of child's head	Segmental resection	Died 1 wk. postop., pulm. embolus
Anderson and Doob (1933)	3	37 M	5 yrs., bleeding, wt. loss, eructation	Mass and anemia	Mass seen on KUB; G.I. series negative	15 x 18 x 12 cm., fistula from bowel lumen to necrotic center; hepatic metastases	None	Died
Silverstone (1934)	3	51 F	2 yrs., pain, vomiting, wt. loss	Mass	Negative	5 x 4 x 3 cm.	Local excision	Died 4th postop. day of thyrotoxicosis
McFarland (1935)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Stommel (1935)	2	39 M	5 wks., pain, jaundice, weakness	Mass and jaundice	Negative	Size of fist, adherent to pancreas and portal vein; hepatic metastases	Laparotomy	Died postop.
Seymour and Gould (1936)	1	54 M	3 mos., pain, jaundice, weakness, fever, vomiting, wt. loss	Mass, hepatomegaly, palpable gallbladder, jaundice	Filling defect	4 x 3 cm., necrotic center with fistula to retro-duodenal abscess	None	Died
Nickerson and Williams (1937)	2	65 M	2 yrs., pain, wt. loss, diarrhea	Not stated	Not stated	15 x 15 cm., necrotic center	None	Died
Nickerson and Williams (1937)	2	70 F	Several mos., pain and weakness	Not stated	Not stated	Ulcerated polypoid	None	Died
Mendillo and Kaufman (1938)	2	49 M	3 yrs., pain, bleeding, wt. loss	Anemia	Diverticulum in 2nd part of duodenum	10 x 7 x 4 cm., necrotic center with short fistula	Local excision	Well 8 mos.
Foshee and McBride (1939)	3	63 M	3 mos., weakness, wt. loss	Mass	Negative G.I. series, extrinsic pressure defect on colon	10 x 10 x 6 cm.	Local excision and duodenojejunos-tomy	Died 20th postop. day, anastomotic leak
Harrington and Ganshorn (1940)	3	38 F	8 mos., weakness	Anemia	Filling defect with ulceration	4 x 2.5 x 2.5 cm., with ulceration	Segmental resection	Well 4 mos.
Mayo (1940)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Itikawa (1940)	3	42 M	13 yrs., pain and bleeding	Mass and anemia	Filling defect with ulceration	Fist-sized, with satellite egg-sized nodules, ulcerated	Palliative gastroenterostomy	Not stated
Henning and Garland (1941)	2	65 F	3 mos., bleeding, nausea, dyspnea	Mass and anemia	Filling defect	8 x 3 x 5 cm., adherent to pancreas, necrotic center	Segmental resection, cholecystoenterostomy, gastroenterostomy	Died 2 dys. postop.

TABLE I.—SUMMARY OF CASES (Con't.)

Author and year reported	Location (part of duodenum)	Age, yrs. sex	Symptoms	Physical findings	Gastro-intestinal x-ray film	Gross pathology	Operation	Result
Shackelford <i>et al.</i> (1942)	2	61 F	3 mos., bleeding, pain, diarrhea	Anemia	Filling defect	Lemon-sized tumor	Pancreaticoduodenectomy	Died 9 mos. later of recurrence
McCullough (1944)	3	42 F	Several mos., pain, fever	Mass and anemia	Not done: KUB showed mass	8 cm. mass, central necrosis with fistula; abscesses and metastases in liver	None	Died
Williams (1944)	4	30 F	6 dys., pain	Anemia, abdominal distention and tenderness	Not done	4 x 3 cm. tumor, direct extension to liver and colon; free perforation	Laparotomy, peritoneal drainage	Died, 2 dys. postop. of peritonitis
Brunschwig and Tiholiz (1946)	3	Not stated	Not stated	Not stated	Ulcer with fistula to retroduodenal abscess	11 x 7 x 3.5 cm., invading head of pancreas	Segmental resection	Well 3 yrs.
Nitshe and Suckle (1947)	2	64 F	Few days, bleeding	Shock	Negative	1.5 cm., periampullary	None	Died, hemorrhage and peritonitis (perforation)
McLean (1948)	2	50 M	4 mos., jaundice vomiting and weight loss	Jaundice	Not done	Lemon-sized, invading portal vein with compression common duct	Gastroenterostomy, cholecystoenterostomy	Died 16th postop. day
Heyman and Clark (1950)	2	42 F	2 yrs., bleeding	Mass and anemia	Filling defect	5 x 2.5 cm., ulcerated	Pancreaticoduodenectomy	Died 16 mos. postop. of liver metastases
Schwartz <i>et al.</i> (1951)	2	55 F	Pain, bleeding, and diarrhea	Anemia	Obstruction of duodenum	5 x 7 x 10 cm., tumor with ulceration; lymph node metastases	Segmental resection with choledochojejunostomy and pancreaticojejunostomy	Well 15 mos.
Swartz and Eckman (1951)	2	56 M	8 mos., weight loss, fever, jaundice	Jaundice	Filling defect	2.5 cm.	Pancreaticoduodenectomy	Well 3 mos.
Ripstein and Flint (1952)	2	44 M	3 yrs., bleeding	Anemia	Normal	2 cm.	Pancreaticoduodenectomy	Died at operation, cardiac arrest
Ripstein and Flint (1952)	3	53 F	3 yrs. pain and bleeding	Mass and anemia	Not stated	Large tumor, invading ascending colon	Segmental resection and right colectomy	Died 7 wks., bowel obstruction
Ripstein and Flint (1952)	1	45 M	5 mos., pain, bleeding and weight loss	Mass and anemia	Not stated	15 cm.	Gastrectomy	Died 11th postop. day, peritonitis
Ayers and Cunningham (1952)	2	80 F	5 mos., bleeding	Anemia	Normal	3 cm.	Segmental resection	Well 9 mos.
Halligan <i>et al.</i> (1952)	3	Not stated	Bleeding	Not stated	Not stated	Not stated	Segmental resection	Not stated
Weinstein and Roberts (1953)	3	54 M	6 mos., pain, bleeding, weight loss	Mass and anemia	2 fistulas and abscess	20 cm., invading ileum and right colon; central necrosis and fistulas	Segmental resection, right colectomy	Well 1 yr.

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<i>Author and year reported</i>	<i>Location (part of duodenum)</i>	<i>Age, yrs. sex</i>	<i>Symptoms</i>	<i>Physical findings</i>	<i>Gastro-intestinal x-ray film</i>	<i>Gross pathology</i>	<i>Operation</i>	<i>Result</i>
Richardson (1953)	2	47 F	Bleeding	Mass	Negative, barium enema showed extrinsic colon deformity	Pedunculated extramural tumor with central necrosis and pseudo diverticulum formation	Local excision, gastroenterostomy	Well, duration not stated
Richardson (1953)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Died of perforation and peritonitis
Joergenson <i>et al.</i> (1953)	3	Not stated	7 mos.	Not stated	Duodenal obstruction; mucosal irregularity	Not stated	None	Died
Judd and Hill (1954)	2	31 M	5 mos., bleeding	Anemia	Not done	7 cm., ulcerated tumor invading pancreas	Pancreaticoduodenectomy	Well 11 mos.
Zintel (1954)	4	44 M	8 mos., bleeding, weight loss	Anemia	Filling defect with ulceration and fistula	4 x 3 cm., with ulceration and fistula into center of tumor	Segmental resection	Died 3 yrs. 8 mos. of metastases
Elias <i>et al.</i> (1954)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	None	Died
Starr and Dockerty (1955)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Starr and Dockerty (1955)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Starr and Dockerty (1955)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Starr and Dockerty (1955)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Starr and Dockerty (1955)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Martin (1955)	2	37 M	4 yrs., bleeding	Anemia	Filling defect with ulceration	5 cm. diameter, ulcerated	Local excision	Well 4 yrs.
Lopez (1956)	2	61 F	Pain and jaundice	Pneumonia, jaundice	Not stated	Metastases generally	Laparotomy	Died 1.5 yrs. after laparotomy
Burgerman <i>et al.</i> (1956)	2	Not stated	Not stated	Not stated	Not stated	Tumor with metastases to abdominal wall and lymph nodes	None	Died
Burgerman <i>et al.</i> (1956)	2	Not stated F	Not stated	Not stated	Not stated	Ulcerated small tumor without metastases	None	Died
Icgoren (1956)	2	Not stated	Not stated	Not stated	Not stated	3.5 x 4.5 cm., invading gallbladder, central necrosis	Resection—no details	Not stated

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<i>Author and year reported</i>	<i>Location (part of duodenum)</i>	<i>Age, yrs. sex</i>	<i>Symptoms</i>	<i>Physical findings</i>	<i>Gastro-intestinal x-ray film</i>	<i>Gross pathology</i>	<i>Operation</i>	<i>Result</i>
Riveros (1956)	1	25 F	Pain, bleeding, vomiting	Mass and anemia	Not stated	15 x 20 cm., free perforation	None	Died of peritonitis due to perforation
Riveros (1956)	2	40 M	6 mos., pain, weight loss, fever	Mass	Normal, barium enema showed extrinsic pressure defect	15 x 25 cm., attached to colon; peritoneal and omental metastases	Segmental resection	Died 10 mos. with sarcomatosis
Riveros (1956)	2	34 F	Pain, weakness, weight loss	Mass	Not stated, barium enema showed extrinsic pressure defect	Large mass	Pancreaticoduodenectomy	Died 2.5 mos. postop. of pulmonary embolus
Riveros (1956)	2	58 F	Pain, bleeding	Mass and anemia	Not stated	Lymphatic metastases	Pancreaticoduodenectomy	Died 6 mos., recurrence
Ochsner and Kleckner (1957)	2	Not stated	Not stated	Not stated	Filling defect	Not stated	Resected—no details	Not stated
Ochsner and Kleckner (1957)	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated	Not stated
Kelley <i>et al.</i> (1957)	3	34 M	7 yrs., pain, chills, weight loss	Tenderness, questionable mass	Fistula	7 x 5 x 6 cm., with invasion of pancreas, central necrosis, and fistula	Pancreaticoduodenectomy	Well 6 mos.
Kwiatkowski (1957)	2	48 F	5 mos., pain, vomiting, diarrhea, weight loss	Mass and anemia	Normal	Size of fist	Pancreaticoduodenectomy	Well 6 mos.
Brombart and Meuris (1958)	4	45 M	1 mo., pain, weight loss	Abdominal tenderness and rigidity	Duodenal obstruction, mucosal irregularity	Free perforation of tumor	Segmental resection	Living 5 mos., with recurrence
Pilati (1958)	1	45 F	3 mos., pain, anemia, diarrhea, weight loss	Mass and anemia	Ulcer	Size of orange, ulcerated	Pancreaticoduodenectomy	Well 2.5 yrs.
Wilber and Pollard (1958)	3	45 F	2 yrs., bleeding	Anemia	Filling defect with ulceration	4 x 2 cm., ulceration	Local excision	Well 8 mos.
Wilber and Pollard (1958)	3	21 F	6 yrs., bleeding, pain, wt. loss, fever	Anemia	Filling defect	3 cm., ulcerated	Local excision	Well 2 wks.
Wilber and Pollard (1958)	2	26 F	6 yrs., pain, bleeding, vomiting	Mass and anemia	Filling defect	11 x 5 x 5 cm., ulceration and fistula leading to necrotic center	Pancreaticoduodenectomy	Well 4 yrs.
Starzl <i>et al.</i> (1960)	4	42 M	9 mos., bleeding	Anemia	Filling defect with ulceration	4.5 x 4 x 3 cm., ulceration	Segmental resection	Well 18 mos.
Starzl <i>et al.</i> (1960)	2	55 M	4 mos., pain, vomiting, weight loss, diarrhea	Tenderness, right upper quadrant	Duodenal obstruction with mucosal irregularity	7 x 6 x 6 cm., invasion of pancreas	Pancreaticoduodenectomy	Well 14 mos.

#### LOCATION

In 51 cases in which the location was given, 27 or 53 per cent were in the second part of the duodenum, 14 or 27 per cent were in the third part, and 5 or 10 per cent each in the first and fourth segments. It is interesting that the high incidence of these tumors in the second part of the duodenum, 53 per cent, is paralleled by a similar preponderance of duodenal adenocarcinomas, 59 per cent, in this area (6).

#### SYMPTOMATOLOGY

Abdominal pain was the most frequent symptom, being noted in 27 cases, usually in the upper abdomen, but occasionally in the lower quadrants. This pain was characterized in a number of ways; it was described as ulcerlike, cramping, constant, boring, or dull and vague in quality. Also common was gastrointestinal bleeding (26 cases). This ranged from massive hemorrhage requiring emergency surgical control (23) to slow bleeding with associated anemia, weakness, and pallor. Weight loss, the third most common symptom, was a prominent complaint in 19 cases.

Less frequent symptoms were vomiting, 7 cases, diarrhea, 6 cases, fever, 6 cases, and jaundice, 5 cases. It is reasonable to believe that diarrhea was related to pancreatic malfunction, since the tumor was located in the second part of the duodenum in five instances (25, 33, 40, 42, 45) and in the first portion in the other (36). Of the 6 patients who complained of fever, there was central necrosis of the tumor with abscesses or fistula formation in 4 (24, 29, 41, 48).

#### PHYSICAL EXAMINATION

Abnormalities related to anemia were the most common findings (26 cases) with pallor, dyspnea, and hemic murmurs. The presence of a mass was also found frequently, in 23 cases. The mass was usually palpated in the epigastrium or the right upper quadrant, but masses in the left upper quadrant and mid and lower abdomen were also noted. The size varied from that of a lemon to a baby's head. The masses were variously described as tender, nontender, smooth, and irregular.

Jaundice was an infrequent finding, 5 cases, and when present was always due to a tumor originating in or involving the second part of the duodenum (26, 31, 41, 46, 47). It should be

emphasized, however, that jaundice was usually not present with tumors in this location. Twenty-two of the 27 cases in which leiomyosarcoma was in the second portion did not have jaundice. Abdominal tenderness was noted in 4 cases.

#### ROENTGEN FINDINGS

Not unexpectedly, the gastrointestinal series was the single most useful diagnostic tool. Of the 34 cases studied by this method, 24 had some abnormality and the remaining 10 were interpreted as normal. The commonest roentgenographic abnormality was the presence of a filling defect which occurred as an isolated finding in 9 cases, with associated ulceration within the filling defect in 7 cases, and with concomitant duodenal obstruction in 4 instances. The salient roentgen finding in 4 other cases was the presence of a fistula (8, 24, 32, 49) directed either into the retroperitoneal space or into other viscera. In 2 cases the mass was seen by means of a plain roentgenogram of the abdomen. In 4 cases in which the result of the gastrointestinal series was normal, an extrinsic pressure defect of the colon was demonstrated by barium enema.

Although the correct pathologic diagnosis was almost never reached preoperatively in the cases analyzed, certain characteristics encountered in the present review and previously described for smooth muscle tumors elsewhere in the gastrointestinal tract (3, 27, 35) should make it possible to increase the index of accuracy. These features are (1) filling defect with sharp borders, (2) ulceration on the surface of an otherwise regular filling defect, and (3) fistula as an isolated finding or emanating from the central area of a filling defect. As previously described by Baker and Good, the presence of a fistula is a particularly ominous portent of malignancy. Other findings such as duodenal obstruction and mucosal irregularity are common to a variety of tumors. Similarly, extraluminal defects detected on barium enema examination were seen in 4 cases in the present review and simply pointed to the general problem of an intra-abdominal mass.

#### PATHOLOGY

Concerning the pathogenesis of leiomyosarcoma, it is not clear in the cases reviewed whether the tumors developed *de novo* or as the result of growth changes in pre-existing leio-

myomas. Such evidence would be particularly hard to obtain since there is some lack of precision in making a microscopic diagnosis in the smooth muscle groups of tumors. Golden and Stout and Starr and Dockerty have emphasized the difficulty of histologic differentiation of benign from malignant neoplasms. Some myomatous tumors that appear harmless under the microscope grow rapidly and metastasize, and others with similar cell structure have benign clinical behavior. In most of the instances reviewed here, evidence of local invasion made the decision of malignancy an easy one. In other patients, invasion had not taken place and the decision was made on the sometimes unreliable basis of the investigation of the cellular anatomy.

One of the common microscopic features of duodenal leiomyosarcoma deserves special attention since it explains several facets of the clinical picture. The tumor has a tendency to outgrow its blood supply with consequent central necrosis and ulceration. This may result in pain, fever, or hemorrhage, and it may produce the characteristic roentgen sign of a filling defect with ulceration at the tip. With further necrosis, the ulcer may be transformed into a fistula with communication into an abscess cavity (1, 24, 29, 32, 37, 41, 48, 49, 50, 52) or into the free peritoneal space (7, 34, 37, 39, 51).

Because of the importance in planning surgical therapy, data were accumulated from 42 cases in which the status regarding metastases was noted (Table II). At the time of surgery or autopsy, only 9 of the 42 patients had metastases beyond the local area. The most common sites of distant metastases were liver, 6 cases, peritoneum and omentum, 3 cases, lymph nodes, 3 cases, abdominal wall, 2 cases, and central nervous system, 1 case.

TREATMENT

From studies on the behavior of leiomyosarcoma in other portions of the small intestines (19, 44) and from consideration of the pathologic findings in the present review, the essence of rational surgical treatment seems to be radical local removal. Procedures designed to remove extensive regional lymphatics would presumably have little beneficial effect on the prognosis, since lymphatic metastases are rare (Table II).

Because of the complex anatomy of the duo-

TABLE II.—INCIDENCE AND LOCATION OF METASTASES IN CASES AT AUTOPSY (NO OPERATION) AND AT OPERATION

	<i>No. of cases</i>	<i>Location of metastases</i>
AUTOPSY		
Without metastases.	5	
With metastases....	3	Liver (2), lymph nodes (1), abdominal wall (1)
OPERATION		
*Without metastases.	28	
With metastases....	6	Liver (2), intestine (1), lymph nodes (2), peritoneum and omentum (2), wound scar (1)

\*In 4 of these patients metastases developed subsequently. There were 2 instances of metastases to the liver, 1 to the brain and cervical spine, and 1 to the peritoneum and omentum.

denum, radical local extirpation in its different portions has necessitated procedures which vary enormously in their magnitude. In the third and fourth parts, segmental resections or local excision with repair of the duodenal defect have usually been possible, occasionally in conjunction with removal of adjacent structures such as the colon which have been invaded. Since the first report of Shackelford (42), lesions in the first and second portions of the duodenum have most often been treated with pancreaticoduodenectomy. Indeed, pancreaticoduodenectomy under these circumstances has seemed to be an acceptably safe procedure since 11 of 12 patients so treated have survived surgery (Table III).

TABLE III.—RELATION OF SITE OF TUMOR AND TYPE OF OPERATION TO MORTALITY

<i>Type of operation</i>	<i>Portion of duodenum</i>				<i>Operative deaths</i>		
	1	2	3	4	<i>No. of operations</i>	<i>No. of deaths</i>	<i>Per cent</i>
Local excision	0	3	4	0	7	2	28
Segmental resection	2	4	5	3	14	4	29
Pancreaticoduodenectomy	1	10	1	0	12	1	8
Resection (no details)	0	2	0	0	2	0	0
Gastroenterostomy, palliative	0	0	1	0	1	0	0
Laparotomy only	0	3	0	2	5	3	60
Totals	3	22	11	5	41	10	24
No. of operative deaths	2	4	3	1			
Percentage of total	67	18	27	20			

The relation of mortality to the location of the tumor and to the type of operative procedure employed is shown in Table III. The figures indicate a high surgical death rate in all locations and with various procedures. Since the evolution of safer methods of pancreatic and duodenal surgery has occurred comparatively recently, analysis of the recent cases would provide a more meaningful estimate of the operative risk. Prior to 1949, 14 patients were operated on and 8 of these died during or shortly after operation. In the last decade, 23 patients have been treated surgically with only 2 deaths.

Recognition of the nature of the tumor at the operating table will not be difficult in some cases, especially if the tumor is small, and if the features described under the pathology section are present. On the other hand, many of the tumors, notably in the second part of the duodenum, have been invasive and extraordinarily large. Since the identifying characteristics were obliterated these have commonly been thought to be carcinoma of the pancreas. These tumors have frequently been successfully removed. It has been pointed out by Bowden that the large size of tumors in this location should not preclude an attempt at resection if other factors are favorable, and this would seem to be a particularly applicable point of view in respect to leiomyosarcoma.

#### PROGNOSIS

There is not yet enough available clinical material to make an authoritative statement about prognosis. In 24 of the cases in which the patient survived surgical therapy (Table IV), 16 were alive and free of metastases at the time of report. Since 9 of the 16 cases had been traced for less than a year, it is likely that recurrent tumor was present but not obvious in some of these.

One pathologic feature of leiomyosarcoma that makes long term follow-up necessary for

evaluation of surgical results is the protracted natural history sometimes observed. One patient in the present review died of sarcomatosis almost 4 years after operation (52) and another lived for 18 months after laparotomy at which no definite procedure was carried out (26).

Despite the uncertainty of prognosis, an aggressive surgical attack on these lesions is certainly indicated. Even when the procedure fails to eradicate the tumor, considerable symptomatic relief can be expected, and in view of the handful of long term survivors (Table IV) some cures may be expected.

#### SUMMARY

1. Sixty-one cases of leiomyosarcoma of the duodenum have appeared in the world literature.

2. Duodenal leiomyosarcoma occurs in both sexes with approximately the same frequency. Peak incidence is in the 40 to 49 year age bracket.

3. More than one-half of the tumors were in the second portion of the duodenum, and the rest were in the third, first, and fourth parts in that order of frequency.

4. The symptoms most frequently encountered were abdominal pain, intestinal bleeding, and weight loss. In a small number of cases diarrhea, vomiting, fever, and jaundice were present.

5. On physical examination, abnormalities caused by gastrointestinal hemorrhage were the most common findings. In a large number of cases an abdominal mass was detected during preoperative examination.

6. The gastrointestinal series was the single most useful diagnostic tool. The most common roentgenographic abnormality was a filling defect in the central portion of which ulceration or fistula formation had often occurred.

7. One common gross pathologic finding has been the propensity of the tumor for central necrosis, a characteristic which explains many of the clinical features and the roentgen appearance of the neoplasm.

8. Metastases from duodenal leiomyosarcoma occur almost exclusively by local invasion.

9. The most effective surgical therapy has been radical local excision of the tumor. Extensive removal of regional lymphatics would not seem to be a fruitful pursuit, since nodal metastases are rare. Since the techniques of

TABLE IV.—FOLLOW-UP AT TIME OF REPORT IN SURGICALLY TREATED PATIENTS WHO SURVIVED OPERATION

Status of patient	Duration of follow-up				Totals
	0-1 yr.	1-2 yrs.	2-3 yrs.	3-4 yrs.	
Well . . . . .	9	3	1	3	16
Dead or with recurrence . . . . .	5	2	0	1	8
Totals . . . . .	14	5	1	4	24



pancreatic and duodenal surgery have been refined, excisional therapy has been possible with a low mortality.

10. In general, follow-up reports after surgical removal of these tumors have been of short duration. Because of this and because the tumor sometimes follows a protracted course even without therapy, it is not possible to make an authoritative statement about the prognosis in the treated patients. However, aggressive surgical therapy is indicated both for relief of symptoms and because of the definite chance for cure in some cases.

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