LEIOMYOSARCOMA OF THE DUODENUM

REPORT OF CASE

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Only three cases of leiomyosarcoma of the duodenum have been reported to date, the first by von Salis (1) in 1920, the second by Andersen and Doob (2) in 1933, and the third by Silverstone (3) in 1934. The following case is the fourth known instance of leiomyosarcoma arising in the duodenum.

REPORT OF CASE

J. S., a white male aged fifty-four, a machinist by occupation, was admitted to the William J. Seymour Hospital on Nov. 11, 1935, and died Dec. 1, 1935.

The chief complaints on admission were pain in the abdomen, loss of weight, itching and jaundice. Up to three months previously the patient had felt perfectly well. At that time he noticed a gradual onset of pain in the upper abdomen below the ribs. He could not take food and vomited frequently after meals. The pain was sharp or burning in character, and occasionally radiated to the back or up into the left chest. He had had no previous stomach trouble. He complained, also, of itching all over his body for the past week, and was told that his skin was yellow. Since the onset of his illness he had had dryness of the mouth and skin and increased thirst, but no increased hunger and no frequency of urination. The appetite was poor. Although the patient stated that he had lost 60 pounds in weight in the last six months, he appeared well nourished.

The patient had had an appendectomy in 1930 for a ruptured appendix. In 1920 he lost his left thumb in an accident. An injury by a piece of steel in 1920 caused loss of sight in the right eye, which was enucleated. A similar injury to the left eye in 1927 caused total blindness. Alcohol was used in moderation.

On physical examination the skin showed marked icterus and numerous scratch marks. A few moist rales were heard at the base of the right lung. The heart was enlarged to the left and downward. The sounds were regular but distant and of poor quality. The blood pressure was 128/80. The abdomen was tender in the right upper quadrant and to a lesser extent in the left upper quadrant. The liver was enlarged and palpable, reaching to within one inch of the umbilicus. There was an enlarged, soft, cystic, gallbladder palpable in the right upper quadrant. In the epigastrium was a tender irregular mass about the size of a grapefruit. The kidneys and spleen were not palpable. There was an incisional hernia in the right lower quadrant. The lymph nodes in the inguinal regions were small and discrete. The patellar reflexes were sluggish, the abdominal reflexes absent.

A roentgenogram of the chest (Nov. 15) showed calcified nodes in both hila with many old scattered calcified foci in the lung fields suggesting a healed miliary tuberculosis. The pleura was thickened on both sides, more so on the right. There were no recent areas of infiltration.

The report on the gastro-intestinal series (Nov. 12) by Dr. J. M. Grace, roentgenologist, was as follows: "The esophagus and stomach are negative. Within the terminal portion of the duodenal bulb and just beyond the bulb are two rounded filling defects (Fig. 1). There is questionable widening of the duodenal loop but no evidence of pressure on its inner margin. Diagnosis: Intra-duodenal tumors or one lobulated tumor, whether polyp or malignancy we are unable to say."
Laboratory findings (Nov. 6) were as follows: Urine: sp. gr. 1.026; albumin, trace; sugar xxxx; acetone, negative. Blood count: Hemoglobin 13.0 gm.; red cells 4,050,000; white cells 8800. Blood glucose 296 mg.; icterus index 80; van den Bergh immediate direct, bilirubin 18 mg. Feces: occult blood weakly positive, no bile present. The blood Kahn and Kline tests were negative. There was a progressive rise of serum bilirubin with no remissions; two days before expiration the bilirubin was 26.5 mg.

The patient was placed on a diabetic diet with 40 units of insulin, and the diabetes was easily controlled. The sleep was interrupted by severe itching. Nausea and vomiting occurred, and there was severe pain in the abdomen. From Nov. 14 on, the temperature and pulse were elevated. The temperature varied from 100° to 102.5°, the pulse from 112 to 130. Death occurred on Dec. 1, 1935. The autopsy was performed by Dr. J. J. Kraus, resident surgeon in pathology, on Dec. 4.

**Fig. 1. Leiomyosarcoma of the Duodenum**

**Autopsy:** The body was that of a well nourished and well developed white male, 160 cm. in length. The skin and conjunctivae were icteric and there were small areas of subcutaneous hemorrhage scattered over the extremities and the trunk.

The abdominal cavity contained no fluid. The panniculus adiposus was yellow and measured 2.5 cm. in thickness.

The right pleural cavity contained 1200 c.c. of clear yellowish fluid.

The heart weighed 375 grams. The musculature was pale, yellowish brown in color, and very flabby. There was a moderate amount of coronary sclerosis. The left ventricle measured 14 mm. in thickness, the right ventricle 4 mm. in thickness. The aorta was negative.

The left lung weighed 525 grams, the right lung 575 grams. Both were moderately congested throughout. Scattered over the pleural surfaces and in the lung tissue itself were numerous small, calcified, miliary nodules.

The stomach was negative.

The liver weighed 1700 grams. It was large, soft, and deeply bile-stained. The cut
surface showed a yellowish-green color with a diffuse punctate to linear mosaic of darker brownish color.

The spleen weighed 225 grams. It was large, deep red, soft, and mushy.

The gallbladder, the cystic duct, and the common bile ducts were distended, their walls somewhat thickened. No stones were present. The gallbladder contained two ounces of dark green bile. The cystic and common bile ducts were compressed near the ampulla of Vater by an abscess located between the first portion of the duodenum and the head of the pancreas. The papilla of Vater was situated 7 cm. below the pyloric ring and its lumen was patent.

One centimeter below the pyloric ring, in the first portion of the duodenum, on the posterior wall, was a soft, fragile, grayish polyp 1 cm. in length and 1 to 1.5 cm. in diameter. Its base measured 8 to 10 mm. in diameter and was not movable. Contiguous with the base of the polyp, immediately below it and to either side, was an irregular area of ulceration which measured 3 to 4 cm. transversely and 2 to 3 cm. in the longitudinal axis of the duodenum. The borders of the ulcer were irregular and the base was of a dirty gray color, ragged in contour, and of varying depth. In the central region was a small perforation which communicated with the abscess mentioned above. The abscess measured approximately 4 cm. in diameter, was situated between the posterior aspect of the duodenum and the anterior surface of the body of the pancreas just to the left of the head of the pancreas, and had thickened walls of a dirty greenish color while its interior was filled with a light greenish pus. The pancreas was yellow with yellowish-white areas of necrosis in the periphery. Upon section it showed marked increase in fat. Its interior was not involved by the abscess.

The adrenals were autolyzed.

The kidneys weighed 225 grams each. There was an increase in the pelvic fat and marked congestion of the parenchyma. The markings were poorly differentiated. The pelvic organs were negative.

Gross Diagnosis: Neoplasm of posterior wall of first portion of duodenum with ulceration, fistula formation, and abscess; compression of common bile duct by retroduodenal abscess; obstructive jaundice; biliary cirrhosis; fatty infiltration of pancreas; myocardial degeneration; coronary sclerosis; right hydrothorax; pulmonary congestion; old healed miliary tuberculosis of pleura and lungs; diabetes mellitus.

Microscopic Examination: The peripheral portions of the duodenal polyp were necrotic and contained a number of dilated blood capillaries. The central portion revealed numerous loosely arranged single cells which varied in diameter from about 20 to 85\mu, the majority of the cells measuring about 35\mu. Most of the cells were round or oval, while others were spindle-shaped or polygonal. The nuclei were large, varying in diameter from about 8 to 40\mu; the majority measured 15 to 20\mu. These cells were arranged loosely as single cells or groups of cells which seemed ready to fall out of the section. With hematoxylin and eosin stain, the cytoplasm appeared abundant and bluish pink. The nuclei stained well, contained granules or clumps of deep blue-staining material and many small vesicular structures having a ground glass or spongy appearance. Most of the nuclei had a coarse granular or a fine mesh-like structure, a few contained distinct nucleoli, and a number showed mitotic figures. Many amitotic division forms were also seen, a number of cells containing two nuclei and a few having three. Many bizarre nuclear patterns were present. The stroma was scant and delicate and capillaries were not prominent. Some groups of cells were surrounded by delicate strands of young fibrous tissue containing young capillary structures and a few scattered lymphocytes. A few faded erythrocytes or their remnants and an occasional phagocytic cell containing blood pigment could be seen.

Other sections taken from the duodenal ulcer showed necrosis and ulceration of the mucosa, lymphoid tissue and new growth present within the submucosa, while the muscular layer was largely replaced by new growth. The cells were pleomorphic, round, oval, or spindle-shaped with large bizarre-shaped nuclei, many of which presented atypical mitotic and other division forms.

Sections of duodenum beyond the area of ulceration showed loss of mucosa, fatty infiltration of the submucosa, atrophy of the muscularis, and fibrosis of all the layers.

Sections stained with del Rio-Hortega’s silver nuclear stain and counterstained with
FIG. 2. SECTION OF LEIOMYOSARCOMA OF THE DUODENUM STAINED WITH DEL RIO HORTEGA’S SILVER STAIN WITH Picro-indigo. × 105

FIG. 3. SECTION OF LEIOMYOSARCOMA OF DUODENUM, STAINED WITH DEL RIO HORTEGA’S SILVER NUCLEAR STAIN. × 210
Note origin of sarcoma cells from muscle fibers.
picro-indigo or picrofuchsin brought out the nuclear detail and served to differentiate the fibrous connective-tissue elements from the muscle fibers. They also showed strikingly the origin of the tumor cells from the muscle fibers (Figs. 2 and 3). With this stain the muscle fibers and the tumor cells appeared yellow, the nuclei dark brown, the connective tissue green with the picro-indigo and red with the picrofuchsin counterstain. Other differential stains employed were the van Gieson, Bielschowsky-Foote reticulum, Mallory aniline blue, and the phosphotungstic acid. With the latter stain, fibrillae could be seen within many of the tumor cells.

The sections of pancreas showed necrosis of the peripheral portions with localized abscess formation and hemorrhage, marked increase in perilobular fibrous tissue with scattered lymphocytic infiltration and mild fatty infiltration. The parenchyma showed areas of necrosis and marked atrophy of the acini. There was mild increase in the perilobular fibrous tissue with some fibroblastic proliferation, and scattered as well as dense focal lymphocytic infiltration. A number of vessels showing obliterating endarteritis were present. Practically all of the duct structures were greatly distended. Many of the ducts showed loss of their lining epithelium, with distention of the lumen by desquamated cells and amorphous pinkish-staining material. Some of the ducts had undergone necrosis. The islands of Langerhans were obscured and definitely decreased in number, but in some areas a few were left behind intact. Small areas of hemorrhage and blood pigment deposits were seen.

The sections of liver showed slight increase in subcapsular and perilobular connective tissue with some lymphocytic infiltration, marked passive congestion with distention of the interlobular veins, severe parenchymatous degeneration with necrosis and mild fatty degeneration affecting chiefly the central portion of the lobules, staining of the necrotic areas by bile pigment, and congestion of the bile capillaries. The bile capillaries and ducts contained deposits of fine granular golden pigment.

The regional lymph nodes showed no evidence of new growth.

Microscopic Diagnosis: Leiomyosarcoma of duodenum; peripancreatic abscess; necrosis and atrophy of pancreas; chronic pancreatitis with obliterating endarteritis; biliary cirrhosis; parenchymatous degeneration of kidneys and adrenals; fatty infiltration of myocardium; purulent bronchitis; chronic miliary pulmonary and pleural tuberculosis; general passive congestion; jaundice.

Review of Literature

Von Salis' (1) patient was a man of forty with symptoms of ulcer for seven years. Seven months before death he had fever and an abdominal mass. A lobulated tumor about the size of a baby's head was found in the duodenum just above its junction with the jejunum. A fistula and an abscess were also present. The neoplasm arose from the muscle layer, did not produce stenosis or dilatation, and was found to be a leiomyosarcoma. No mitoses were seen. von Salis believed that the tumor arose as a benign leiomyoma, possibly on the basis of a chronic ulcer.

Andersen and Doob's (2) patient was a male machinist of thirty-seven who had had symptoms of peptic ulcer five years previously. Symptoms recurred along with the presence of a lump in the right side of the abdomen six weeks before death. The x-ray showed a mass in the right upper quadrant, believed to be an abdominal tumor pressing on the colon. At autopsy a neoplasm 15 x 18 x 12 cm. was found in the second and third portions of the duodenum involving chiefly the posterior wall. Two ulcerations were present leading into cavities present within the mass. The tumor revealed rare mitotic figures, scanty non-vascular stroma, and tumor cells having delicate protoplasmic fibrillae demonstrated by Mallory's phosphotungstic acid and Masson's trichrome stains.
Silverstone’s (3) patient was a housewife of fifty-one with epigastric pain for two years, which became severe during the last two months. A mass 2 × 1 inches was felt in the right hypochondrium. Roentgenograms of the gastro-intestinal tract were negative. At autopsy a new growth measuring more than 5 × 4 × 3 cm. was found in the third portion of the duodenum. No ulceration was noted. The pathologist reported “a borderline growth of unstriped muscle—myosarcoma.”

In the case here reported the patient was a male of fifty-four, a machinist, who complained of loss of sixty pounds of weight over a period of six months. During the last three months there occurred loss of appetite, nausea and vomiting, epigastric pain radiating to the left chest and back, progressive jaundice, and symptoms of diabetes mellitus. The roentgenologist reported “intra-duodenal tumors or one lobulated tumor due to polyp or malignancy.” Autopsy revealed a new growth of the first portion of the duodenum, which projected as a polyp into the lumen and was continuous with an area of ulceration measuring 4 × 3 cm. In the central portion of the ulcerated area was a perforation, resulting in the formation of a fistula and a retroduodenal abscess which overlay the pancreas and compressed the common bile duct. Obstructive jaundice, biliary cirrhosis, and a moderately severe diabetes mellitus resulted. The microscopic picture revealed a leiomyosarcoma which was apparently more malignant than the tumor in any of the cases previously reported. Numerous mitotic and other division forms were present. No metastases were found.

Prey, Foster, and Dennis (4) also list cases of myosarcoma reported by Ghon and Hintz (5), Kathe (6), and Wesener (7), but in these cases it is not clear that the duodenum was the primary site. In Kathe’s case the tumor arose in the duodenojejunal flexure. Andersen and Doob mention a possible case by Hüttl (8), which “infiltrated” the pancreas, but the latter author stated that the tumor was a myoma and that there were “intimate adhesions” to the pancreas. Andersen and Doob analyzed 18 cases, exclusive of their own, of leiomyosarcoma of the small intestine which have been reported in the literature. In two of these cases (9, 10) the sarcoma was successfully removed.

Most tumors of the duodenum are benign (11) and occur in the first and second portions. Of the malignant tumors, carcinoma is commoner than sarcoma. Among the sarcomata, the commonest type is lymphosarcoma. The latter is usually limited to the mucosa and submucosa and tends to grow longitudinally. It is regularly associated with metastasis. The myosarcoma, arising from the muscular layer, tends to grow externally. It is frequently associated with ulceration, fistula formation, and abscess, but does not often set up metastasis. Myosarcoma of the stomach is said to be more benign than lymphosarcoma. According to Prey, Foster, and Dennis, no patient with sarcoma of the duodenum has been reported as cured either as the result of surgery or radiotherapy. Feyrter (12) states that perforation of myogenic tumors of the stomach or intestinal tract into the peritoneal cavity appears to represent a complication of a malignant tumor.

La Roque and Shiflett (13) believe that the diagnosis of tumor of the duodenum by x-ray should be made invariably. A filling defect, especially with
dilatation of the duodenum, is said to constitute a reasonable basis for a
diagnosis of tumor, as distinguished from ulcer and diverticulum. The defect
is best shown with the patient in the right oblique position. A vacuolar defect
suggests adenoma or sarcoma.

SUMMARY

The literature containing three known cases of primary leiomyosarcoma
of the duodenum is briefly reviewed. A fourth case is reported, with roentgen
findings.

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