RETROPERITONEAL FIBROSARCOMA

REPORT OF EIGHT CASES

W. L. McNAMARA, M.D., H. D. SMITH, M.D., AND C. S. BOSWELL, M.D.

(From the Pathological Laboratory and Cancer Service, Veterans Administration Hospital, Los Angeles, Cal.)

Connective-tissue tumors are unusual in the retroperitoneal space. The finding of eight such neoplasms during 2500 routine autopsies forms the basis of this report.

Culver and Baker state that benign retroperitoneal tumors are not common, although over 200 cases appear in the literature; the majority of these are lipomatous in nature. Retroperitoneal fibromata have been reported by Schmid, Cope, Lind, Pritzi, Danheisser, and Ogilvie.

Magoun found in the records of the Mayo Clinic, from January 1907 to September 1919, 73 examples of non-metastatic retroperitoneal tumors. Fifty-three patients were operated upon and the growths were shown to be malignant in 29 instances and benign in 18 while in 6 the nature was undetermined. The malignant group showed the following tumors of connective-tissue origin: 4 spindle-cell sarcomas, 2 fibrosarcomas, and a fibroblastoma. In the benign group there was one fibroma.

Andrews studied 28 cases of retroperitoneal sarcoma, among which were the following of connective-tissue origin: 4 spindle-cell sarcomas, 4 fibrosarcomas, 2 mixed-cell tumors, 2 fibromyxomas, and a myxo-osteochondrosarcoma. Metastases occurred in 33 per cent of his cases: commonly to the liver, lungs, and lymph nodes; uncommonly to the spleen, kidney, skin, omentum, muscle, pleura, heart, bone, dura, spinal cord, and adrenals. Andrews noted that trauma was apparently not of etiologic importance. In our series only one patient gave a history of trauma.

Lind's retroperitoneal tumor showed only myxomatous and fibrous tissue. He terms it a fibromyxoma, and comments on the rarity of retroperitoneal myxomata unassociated with fat. Myxomatous tissue in our case we interpreted as a degenerative change, in accord with the opinion of Greco, who says: “Pure myxoma originates from embryonic tissue; in the myxosarcoma group only the minority can be considered pure malignant myxoma, the majority being sarcoma with a few islands of degenerated mucoid tissue.” In every instance sections stained with Sudan III failed to show the presence of fat cells.

SYMPTOMS AND CLINICAL COURSE

The symptoms of retroperitoneal growths are not characteristic. The fibrosarcomas are singularly devoid of symptoms and do not as a rule come to clinical attention until their size creates evidence of intra-abdominal pressure. The patients are free from the cachexia and signs of toxicity which so

1 Published under R & P, 6727, Veterans Administration.
characteristically accompany malignant neoplasms. Enlargement of lymph nodes and other evidences of metastasis are not seen. Metastasis occurred in only two of our cases; tumor nodules were found in the lungs in Case IV and in the skeleton, liver, and regional lymph nodes in Case VIII.

Danheisser reports the accidental discovery, during a general examination, of a large growth filling the abdomen of a man aged thirty-seven. It had occasioned no symptoms. An exploratory laparotomy and examination of the excised tissue showed it to be a fibroma. Cope’s patient, a girl of nineteen, had gastric symptoms. At operation a retroperitoneal fibroma was discovered which imprisoned and slightly compressed a portion of the posterior wall of the stomach.

**Diagnosis**

The diagnosis of retroperitoneal growths is difficult and is seldom made prior to operation or autopsy. Pritzi notes that in females these tumors are frequently diagnosed as of ovarian origin. In the case which he reports the tumor was considered to be most probably a metastatic ovarian carcinoma. In our series the preoperative diagnoses included retroperitoneal sarcoma, renal tumor, fibrosarcoma of the groin, tumor of the descending colon, and hypernephroma.

**Therapy**

The only rational therapy of these tumors is radical extirpation in the cases in which surgery is possible. Pritzi states that operation should be done at the earliest possible moment, as he believes that there is a tendency to malignant change, as in his case. He adds that the technic of the operation is often complicated because of adhesions to neighboring organs, and that intestinal resection may be necessary. The employment of combined roentgen and radium therapy, as advised by Andrews, is regarded by Pritzi as questionable, as it is almost impossible to make a diagnosis prior to operation, but postoperative roentgen therapy may be advantageous. He notes the frequency with which resection of retroperitoneal tumors is accompanied by shock. Schmid also stresses the necessity of early operation.

In Danheisser’s case the tumor was diagnosed by exploratory laparotomy and removed surgically.

**Prognosis**

The prognosis is rendered unfavorable by the fact that these tumors are seldom recognized until their size and adhesions to important structures render them inoperable. As pointed out above, they show little tendency to metastasis, and it may be assumed that if it were possible to detect the tumor at an early stage and to subject it to radical surgical extirpation recovery would result. Pritzi states that the prognosis is not unfavorable in operable cases. The result of surgical extirpation in Ogilvie’s case was good. Schmid states, however, that at least half of the solid tumors of the retroperitoneal space behave in a malignant manner even when they are benign histologically.
RETROPERITONEAL FIBROSARCOMA

REPORT OF CASES

Case I: G. S., white male aged forty-one, had noticed a gradual increase in the size of the abdomen for a year. During this time he had had increasingly severe abdominal pains of a colicky nature, a sense of fulness after eating, heartburn, and gastric distress. The family history and past history were not significant. There was no history of trauma.

Exploratory laparotomy revealed a large inoperable retroperitoneal mass, diagnosed from the biopsy specimen as fibroma. Deep roentgen therapy produced no results and the patient died eight months later, with evidences of circulatory failure.

Post-mortem Examination: On opening the abdomen a large nodular mass was found, entirely confined to the retroperitoneal space. The tumor, measuring 30 × 42 cm., had pushed forward and to the left, displacing the abdominal organs to the right. About 1000 c.c. of straw-colored fluid was found in the peritoneal sac. The lymph nodes were not en-

Fig. 1. Case I: Well Differentiated Long Spindle Cells Showing No Variation in Size or Staining Characteristics. × 300

larged and the liver, spleen, and pancreas were normal. The genito-urinary and gastrointestinal tracts showed no gross pathological lesions.

The tumor, which weighed 7230 gm., was grayish white in color and nodular. It apparently arose in the retroperitoneal fascia. Cut section revealed a glistening white surface with small areas of gelatinous change. Microscopic study showed a highly cellular spindle-cell growth, the cells of which were well differentiated and in places were undergoing myxomatous degeneration. The histologic diagnosis was fibrosarcoma undergoing myxomatous degeneration. There were no metastases.

Case II: J. V., white male aged fifty-six, stated that for the past six months he had not felt as well as previously. During that time a tumor the size of a grapefruit had appeared in his right flank; he had suffered with nausea and heartburn following meals, and there had been a decrease in appetite and loss of weight. The past history and family history were negative. There was no history of trauma.

Clinical examination showed a mass in the right side, and a neoplasm of the right kidney was suspected. Exploratory laparotomy revealed a tumor 9 cm. in diameter in the right renal region, not attached to the kidney. A biopsy was not obtained.

Following roentgen therapy the tumor decreased in size and the patient gained weight. Death occurred, however, four months after operation, with evidences of circulatory failure.
The growth was highly cellular, being composed of hyperchromatic spindle cells, as shown in the photomicrograph (× 150). The cut section of the aorta is to be seen in the upper left corner of the gross photograph.

**Post-mortem Examination:** On opening the abdomen a nodular mass covered with peritoneum, measuring 28 × 28 × 20 cm., was discovered, displacing the intestines and other abdominal organs. No excess fluid was present and the abdominal lymph nodes were not enlarged. The liver, spleen, and pancreas showed nothing of significance. No lesions were observed in the gastro-intestinal tract or in the kidneys.

The tumor, weighing 8000 gm., was retroperitoneal, apparently originating in the region of the right kidney. It was nodular and grayish yellow in some areas, white in others. Gross section showed a grayish white surface with scattered areas of caseous-like degeneration, small hemorrhagic areas, and gelatinous changes in portions. Grossly the appearance was that of a fibroma which was breaking down and undergoing myxomatous change. Histologic study showed the tumor to be made up of closely packed, well differentiated spindle-shaped cells. The nuclei were hyperchromatic and a mitotic figure was seen. The diagnosis was fibrosarcoma.

**Case III:** R. E. M., white male aged fifty-one, complained of masses in the abdomen and a weight loss of thirty pounds in eight months. Nocturia was the only subjective symptom. The onset of the illness was dated back to an accident fourteen months previously, when the patient was struck in the abdomen by the steering wheel of an automobile. No abdominal or genito-urinary symptoms resulted, save a slight hematuria which was observed three weeks after the accident and had not recurred. Otherwise the history was not significant.

The x-ray picture was that of an extrinsic mass causing displacement of the stomach and the left kidney to the right. The clinical diagnosis was retroperitoneal sarcoma. An exploratory laparotomy revealed a retroperitoneal tumor, and biopsy sections proved it to be a cellular fibroma, probably malignant. Deep roentgen therapy caused a slight regression in size, with some temporary improvement in the general well being of the patient. Death occurred after a few months.

**Post-mortem Examination:** The abdomen was found to be filled by an enormous retroperitoneal mass, measuring 65 × 55 × 25 cm., pushing the spleen and left kidney under the lower right border of the liver and crowding all the intestines to the right side. The transverse colon and the descending colon were seen on the anterior surface of the tumor.

The pericardial and pleural cavities showed no evidence of tumor invasion. The lungs were pushed upward, the diaphragm being crowded to the level of the 4th rib on each side. The lungs and heart showed nothing of special significance. The liver and spleen were small. The left kidney was rotated to the right by the tumor, but there were no significant renal changes. The bladder, prostate, and vesicles showed no gross lesions. The gastro-intestinal tract was pushed to the right of the tumor. The stomach was adherent to the region of
The gross photograph shows the lobulated tumor in situ. Note the glistening appearance. The spleen lies below the liver and the sectioned left kidney under the right costal margin. The photomicrograph (X 150) shows the loosely disposed cells and extensive myxomatous change.

In the lower portion of the gross photograph may be seen the invasion of the laparotomy scar. The photomicrograph (X 150) shows the hyperchromatic spindle cells which made up the tumor. Metastases were found in the lungs.

the hepatic flexure of the colon and the colon was adherent to the anterior surface of the tumor.

The tumor was lobulated, measuring 25 × 75 × 65 cm., and weighed 27,200 gm. On section it was fibrous in character, and showed areas of myxomatous change and of degeneration and softening. Histologic study revealed it to be a fibrosarcoma undergoing myxomatous change.

Case IV: S. R., white male aged forty-three, complained of pain in the back, colicky abdominal pains, and alternating periods of constipation and diarrhea, of seven months’ duration. The clinical diagnoses included left-sided hypernephroma and sarcoma of the left groin. An exploratory laparotomy revealed a large retroperitoneal growth, which was termed inoperable. It proved on biopsy to be a fibrosarcoma. The patient showed a severe secondary anemia and generalized edema. Roentgen therapy was administered, and the tumor became smaller but death occurred six months later.

Post-mortem Examination: On opening the abdomen a large retroperitoneal mass was found in the left abdomen and pelvis. It impinged on the ilium, which was eroded in places, extended into the inguinal region for a distance of about 6 cm. below the ligament, and
The tumor was composed of interlacing, pale-staining, elongated cells, and was abundantly vascularized. Photomicrograph X 150.

protruded subcutaneously in the left flank, where a superficial ulceration of the overlying skin, about 10 cm. in diameter, was noted. The peritoneum was not invaded. The heart showed nothing of importance.

Scattered throughout the right lung, which weighed 450 gm., were firm, grayish-white, well circumscribed nodules ranging in size from a pea to a kidney bean. A moderate amount of anthracotic pigment was observed and there were marked edema and pressure congestion at the base. The left lung weighed 350 gm. and showed practically the same gross characteristics. There were no gross pathologic changes in the mediastinal nodes. The liver, spleen and pancreas showed nothing of special interest.

The right kidney weighed 210 gm. and was normal. The left kidney weighed 200 gm., and on section showed a hydronephrosis which had destroyed about half of the medulla. The markings were hazy. The left ureter was dilated. It passed over the surface of the tumor, which compressed it in its lower portion. The bladder was normal; the prostate was not enlarged. The gastro-intestinal tract showed no gross pathological lesion. No evidence of metastases was found in the brain.

The tumor weighed 7200 gm. and measured $36 \times 20 \times 12$ cm. It was grayish white, with areas of gelatinous degeneration. Histologic study proved it to be a highly cellular new growth made up of spindle cells which showed some variation in size and staining characteristics. No metastases other than those in the lungs were found. A few mitoses were seen.

Case V: H. C., white male aged forty-six, complained of lumbago-like pains in the back and legs of five weeks’ duration. There was no history of constipation or abdominal pain. The family history and past history showed nothing of importance.

Physical examination revealed a mass in the left lower quadrant measuring $12 \times 15 \times 5$ cm. The patient refused treatment and left the hospital. An outside physician made a diagnosis of carcinoma of the sigmoid and performed an exploratory laparotomy, which revealed a retroperitoneal mass. No biopsy was obtained. The patient was brought back to the hospital and died a few days later.

Post-mortem Examination: On opening the peritoneal sac a retroperitoneal mass some 10 cm. in diameter was encountered about 6 inches above the brim of the pelvis on the left. It partially encircled the gut. A smaller mass was seen below this. The brain, heart, lungs, liver, spleen, pancreas and kidneys showed nothing of importance. In the gastro-intestinal tract was an old healed ulcer on the posterior wall of the duodenum. The bladder was distended and contained a large amount of gravel. The prostate and testes were normal.

The tumor weighed 2200 gm. and partially encircled the sigmoid portion of the colon but did not invade it. The colon remained patent. On section the tumor tissue was seen to be grayish white with small areas of hemorrhage and scattered gelatinous areas suggesting
In the gross photograph the tumor can be seen invading the laparotomy wound. Photomicrograph × 300.

myxomatous degeneration. The smaller mass showed the same gross characteristics. The histologic diagnosis was fibroma undergoing myxomatous change.

Case VI: L. L. R., white male aged fifty-seven, stated that about five months prior to admission he began to have a sensation of fullness in the upper right abdominal quadrant, and at about the same time noticed a large tumor in that area. Since then he had lost fifty pounds in weight. He had no pain, only discomfort due to pressure, and constipation. He had suffered no urinary disturbances.

About two weeks after the onset of symptoms the patient entered a hospital, where an exploratory laparotomy was done and an inoperable tumor was discovered, believed probably to be a left hypernephroma. He was transferred to the present hospital for deep roentgen therapy, but his condition was so poor that this was not carried out. Death ensued about seven months after the onset of symptoms. There was no history of trauma in this case.

Post-mortem Examination: A large tumor filled the left abdomen, extended beyond the midline, and protruded through the ulcerated operative incision. The lower extremities were edematous.

On opening the peritoneal sac a retroperitoneal mass arising in the left lower quadrant was observed, pushing the peritoneum forward and to the right. The descending and transverse colon were on the right of the midline. The tumor did not extend into the peritoneal sac. The mass growing through the old abdominal scar on the left side had followed the line of the surgical incision. The pleura, pericardium, heart, lungs, liver, spleen, pancreas, abdominal and mediastinal nodes showed nothing of importance. The genito-urinary tract was essentially normal save for a marked hydronephrosis of the left kidney due to pressure on the ureter by the retroperitoneal mass. Aside from displacement by the tumor, the gastro-intestinal tract was normal.

The tumor apparently arose from the fascia of the retroperitoneal region of the left lower quadrant. It was grayish white and nodular, with areas of gelatinous or necrotic change. The point of attachment measured about 7 cm. in diameter, and much of it was gelatinous in nature. The body of the tumor measured $32 \times 22 \times 27$ cm. and the weight was 7600 gm. In its growth it had rotated, pushing the entire intestinal tract to the right of the abdomen and displacing the spleen toward the midline. The left kidney was not displaced.

Microscopic examination showed a highly cellular fibrosarcoma; the cells for the most part were of spindle type with occasional round and polyhedral cells. An occasional mitotic figure was noticed. Small areas of myxomatous degeneration were present. There were no metastases.
CASE VII: B. A. G., white male aged forty-six, had been well until the onset of the present illness, in July 1935, when he experienced intermittent dull abdominal pains of a bearing-down type, radiating to the back and left thigh. The abdomen was enlarged but not tender. Save for constipation, there were no symptoms referable to the gastro-intestinal tract, and none to the genito-urinary tract.

Six months after the onset of symptoms the patient consulted a physician. At that time the abdomen was swollen and taut, and a large mass was palpable in the left side. Roentgen examinations of the gastro-intestinal and genito-urinary systems and of the chest revealed nothing of importance. A moderate secondary anemia was present, but there was no blood dyscrasia. A diagnosis of tumor either of the spleen or of the left kidney was made. An abdominal section (June 19, 1936) revealed a large inoperable retroperitoneal mass, which was thought to be either a retroperitoneal sarcoma or a hypernephroma.

The patient was admitted to this hospital July 26, 1936, having lost twenty pounds since the onset of symptoms. The abdominal tumor at this time was massive and nodular, extending from the symphysis to the ensiform cartilage. High-voltage x-ray therapy was administered through four portals, but no regression in the size of the mass was obtained. The patient rapidly lost weight, and showed progressive anemia. Death occurred Dec. 11, 1936, about eighteen months after the onset of symptoms.

Post-mortem Examination: A nodular retroperitoneal mass filled the abdomen. It was apparently fixed to an area left of the midline, displacing the spleen to the midline and crowding the small intestines to the right side of the peritoneal scar. The aorta was partially surrounded by the tumor, and the stomach and liver were displaced upwards. The left kidney lay behind the tumor. The regional lymph nodes were not involved.

The liver, spleen and pancreas showed no gross evidences of pathologic changes. Aside from marked displacement, the gastro-intestinal tract appeared normal, as did the genito-urinary organs. The heart showed softening. The right ventricle was hypertrophied and dilated. There was generalized arteriosclerosis. In the lungs pneumonoconiosis of marked degree was observed. There were healed tuberculous areas in the peribronchial area and mediastinal lymph nodes. No tumor metastases were observed.

The tumor weighed 15,500 gm. and measured $45 \times 32.5 \times 25$ cm. It was grayish in color, smooth and firm. The cut section disclosed a lobulated, grayish-pink mass with areas of gelatinous change. A number of small caseous-like areas were noticed.

On microscopic examination the new growth was seen to be composed of round, oval and spindle cells rather loosely disposed. These cells stained evenly and showed no tendency to variation in size. No mitotic figures were found. Sudan III stain revealed no fat in the cells. The diagnosis was fibrosarcoma.

CASE VIII: G. L. S., white male aged forty-eight, gave a history of diphtheria and smallpox during childhood. During military service (1917-1918) he had been told he had a weak heart but had never experienced symptoms suggestive of cardiac derangement.
FIG. 8. CASE VIII: LOBULATED SPINDLE-CELL TUMOR (3180 gm.), SHOWING MARKED VARIATION IN SIZE OF CELLS

Metastases were present in the bones, lymph nodes, and liver. Photomicrograph × 300.

The patient was in fairly good health until June 15, 1936, when he began to experience a dull abdominal pain, radiating to the back on the left side. The pain was constant, but never "unbearable." A physician was consulted two months after the onset. After roentgen examinations of the gastro-intestinal and genito-urinary tracts the patient was told that he had a tumor in the region of the left kidney and operation was advised. The symptoms continued, the pain assuming a pulling character. No symptoms referable to the urinary system were experienced. On Oct. 7, 1936, a left lumbar incision was made and a retroperitoneal tumor was found, apparently not connected with the kidney. The biopsy diagnosis was fibrosarcoma. Recovery was uneventful.

The patient was admitted to this hospital Nov. 22, 1936. He was somewhat emaciated, having lost twenty pounds since the onset of symptoms. A palpable mass was present in the left lumbar region. Treatment was limited to supportive measures. Death occurred Dec. 9, 1936.

Post-mortem Examination: On opening the peritoneal sac a retroperitoneal tumor the size of a football was encountered. It surrounded the left kidney and extended almost to the level of the diaphragm. The spleen was displaced toward the midline. Two metastatic nodules, 2.5 cm. in diameter, were found in the lower portion of the right lobe of the liver. All of the regional lymph nodes were involved, the largest being 5 cm. in diameter. The left kidney and adrenal were surrounded by the tumor but were not involved. The gastro-intestinal and genito-urinary tracts showed no gross pathological lesions. The cardiovascular system, aside from a mild atherosclerosis, was negative. The lungs revealed no gross lesion.

Metastases were present in practically all of the lumbar and thoracic vertebrae and in the ribs on both sides. There was a pathological fracture of the fourth rib on the left side in the mid-axillary line.

The tumor weighed 3180 gm. It was grayish-white in color and nodular. On section it had a grayish glassy appearance with small translucent areas of gelatinous change. Except in the immediate area of the operative wound there was little evidence of breaking down. The metastatic nodules in the liver and lymph nodes were firm, grayish white, and homogeneous. The bony metastases were soft and gray with marked destruction of bone architecture. There was no evidence of bone formation.

Microscopic examination revealed a highly cellular spindle-cell growth. The cells were of the same size, closely placed, and had oval, reticular, somewhat hyperchromatic nuclei. There were occasional mononuclear giant cells; a few mitotic figures were seen. The growth was quite vascular. In some areas myxomatous changes were observed. Sudan III stain failed to show fat. The diagnosis was fibrosarcoma with metastases to lymph nodes, liver, and skeleton.
SUMMARY

(1) Eight cases of retroperitoneal fibrosarcoma were encountered in approximately 2500 routine post-mortem examinations.

(2) These tumors were symptomless until they had reached inoperable proportions, and caused damage to vital structures. They were not accompanied by the degree of systemic toxicity or cachexia usually associated with malignant tumors of a corresponding size. They showed little tendency to metastasize.

(3) The tumors were radio-resistant.

(4) Surgical extirpation is the rational treatment in cases in which the tumor is diagnosed before its size and relation to neighboring organs render operation impossible.

BIBLIOGRAPHY