TUMORS OF THE RETROPERITONEUM MESENTERY AND PERITONEUM

Lauren V. Ackerman, M. D.

ARMED FORCES INSTITUTE OF PATHOLOGY

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TUMORS OF THE RETROPERITONEUM MESENTERY AND PERITONEUM

by

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Lauren V. Ackerman

TUMORS OF THE RETROPERITONEUM, MESENTERY, AND PERITONEUM

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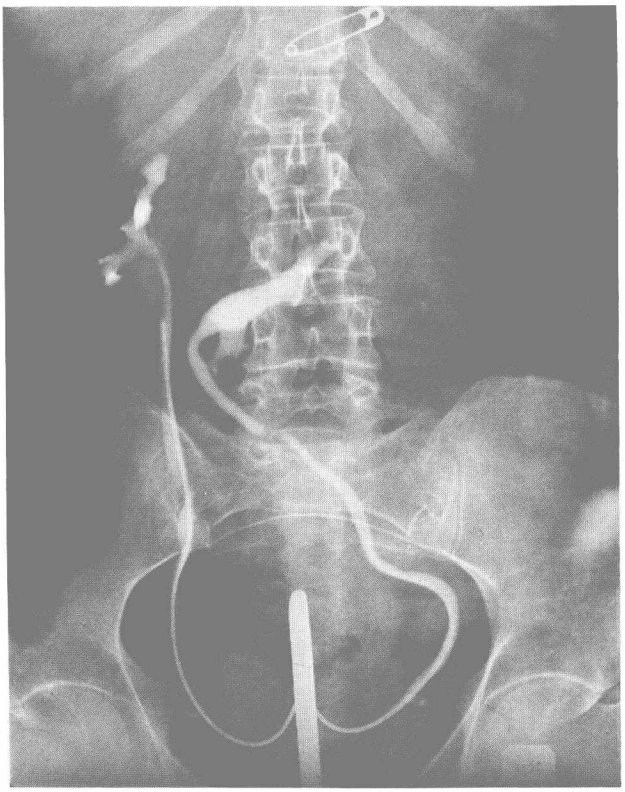

Fig. 1

LIPOSARCOMA

(Figures 1 and 2 are from the same case)

Figure 1. Retrograde pyelogram. Extreme displacement of stomach and ureter in a patient with a large retroperitoneal liposarcoma. A. F. I. P. Acc. No. 219023–33.

Figure 2. Lateral roentgenogram of the patient whose pyelogram is shown in figure 1. A. F. I. P. Acc. No. 219023–34.

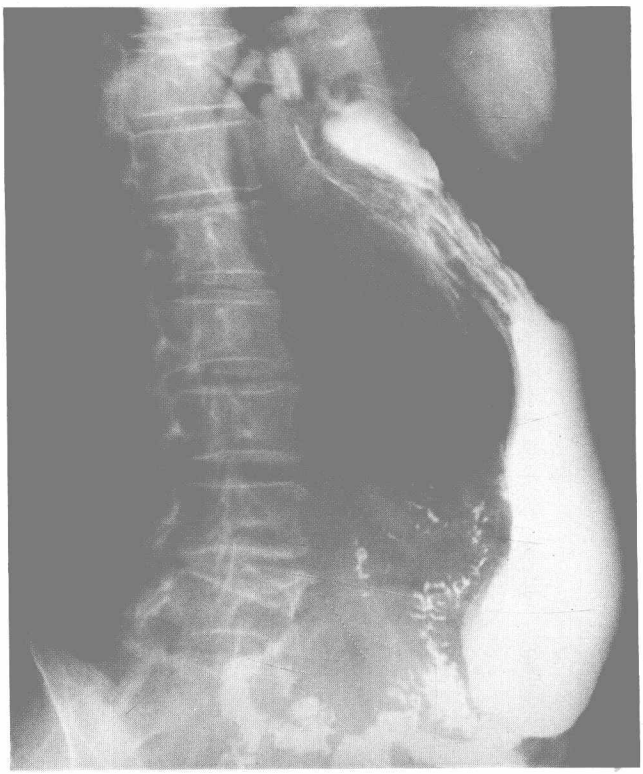

Fig. 2

TUMORS OF THE RETROPERITONEUM, MESENTERY, AND PERITONEUM

TUMORS OF THE RETROPERITONEUM

(Originally planned as Fascicle 24)

INTRODUCTION

Definition

The retroperitoneal space is that indefinite area in the lumbar and iliac region which lies between the peritoneum and the posterior parietal wall of the abdominal cavity. It extends upward to the twelfth rib and twelfth thoracic vertebra and downward to the base of the sacrum and the iliac crest. The lateral margins correspond to the lateral borders of the quadratus lumborum muscles. The floor of the space is formed by the quadratus lumborum and psoas major muscles, and extends from the lateral lumbocostal arch above to the iliolumbar ligament below. The space contains loose areolar fatty tissue, through which pass the ureters, nerves and ganglia, the renal vessels, and the spermatic vessels in the male and the ovarian vessels in the female. The retroperitoneal tissue abuts against the inferior vena cava on the right and the aorta on the left. There are numerous lymph nodes in this region: iliac lymph nodes in the iliac fossa and many lymph nodes along the aorta. The peritoneum covering the apposed surfaces of the rectum and bladder forms the rectovesical cul-de-sac (pouch of Douglas), which extends to the uppermost margin of the rectovesical fascia bridging the extraperitoneal portions of the rectum and bladder.

Scope

This fascicle includes tumors arising primarily from tissues in this retroperitoneal area; it does not include any discussion of primary neoplasms arising from organs that are partially or completely retroperitoneal such as the pancreas, kidneys, and adrenals, or of tumors primarily or secondarily involving bone and spinal cord. The fascicle, therefore, will not include ependymomas (Hundling), giant cell tumors, chondrosarcoma, and other neoplasms of this region that may invade the retroperitoneal zone. Chordomas of the sacrococcygeal region are included. The discussion briefly mentions metastatic tumors.

Primary retroperitoneal malignant neoplasms represent a variegated rare group. There is a vast literature on retroperitoneal tumors. Stout has collected the largest series of benign and malignant retroperitoneal tumors, based primarily on surgical material. The incidence of various types of tumors in his

series (tables I and II) is not necessarily representative of the true incidence, for a large proportion of these tumors were referred to Stout for diagnosis. It is unfortunate that in the literature gross and microscopic descriptions are often inadequate for diagnosis. There are many single cases reported, and usually there is no follow-up. At times the photomicrographs demonstrate an eloquent incorrect diagnosis. The rarity of these retroperitoneal neoplasms precludes any breakdown into relative frequency, and consistent lack of follow-up and too often incorrect diagnoses render it impossible for any predictions to be made concerning prognosis. This fascicle therefore represents merely an introduction to this complex area and, we hope, a stimulation to more careful classification of neoplasms arising there, as well as to more thorough follow-ups.

Table I

BENIGN RETROPERITONEAL TUMORS IN LABORATORY OF SURGICAL PATHOLOGY, COLUMBIA UNIVERSITY, 1905 to 1951 INCLUSIVE*

	Retroperitoneal	Mesenteric	Omental
Adenoma	1	.....	.....
Papillary cystadenoma	2	.....	.....
Lymphangioma	.....	3	.....
Lipoma	4	2	5
Xanthogranuloma	5	.....	.....
Neurilemoma	1	.....	.....
Neurofibroma	6	1	.....
Ganglioneuroma	4	.....	.....
Granular cell myoblastoma	11	.....	.....
Dermoid cyst	2	.....	.....
Leiomyoma	6	1	4
Paraganglioma (inactive)	2	.....	.....
Pheochromocytoma (active)	14	.....	.....
Cyst of caudal gut	17	.....	.....
Total	75	7	9

*Courtesy Arthur Purdy Stout, M. D.

General Characteristics

There are certain general characteristics common to many of the retroperitoneal tumors. Because the retroperitoneal space is rather extensive, and adjacent organs are easily displaced rather than invaded, many of the neoplasms grow to a large size before giving any clinical manifestations. Fre-

Table II

MALIGNANT RETROPERITONEAL TUMORS IN LABORATORY OF SURGICAL PATHOLOGY, COLUMBIA UNIVERSITY, 1905 to 1951 INCLUSIVE*

	Retroperitoneal	Mesenteric	Omental
Sympathicoblastoma	18	.....	.....
Malignant schwannoma	3	.....	.....
Lymphosarcoma	51	2	2
Hodgkin's disease	26	.....	.....
Plasmocytoma	.....	1	.....
Leukemia	2	.....	.....
Fibrosarcoma	4	6	.....
Liposarcoma	35	2	**2
Leiomyosarcoma	29	2	4
Hemangiopericytoma	18	1	2
Rhabdomyosarcoma	5	.....	.....
Mesenchymoma	5	.....	.....
Myxoma	3	.....	.....
Teratoma	6	.....	.....
Embryonal carcinoma	24	.....	.....
Suprarenal cortical carcinoma	1	.....	.....
Metastatic carcinoma of unknown origin	13	.....	.....
Malignant melanoma (metastatic)	1	.....	.....
Chordoma	2	.....	.....
Dysgerminoma	1	.....	.....
Undiagnosed tumor	18	1	4
Total	265	15	14

*Courtesy Arthur Purdy Stout, M. D.
**3 more attached to kidney capsule.

quently, when the patient is first seen, there is an indefinite abdominal mass the exact nature of which is obscure, and the diagnosis is not made until the time of surgical investigation. The operation is often complicated by the large size of the tumor and by its attachment to vital structures; therefore, operative mortality is high and complete removal difficult. It has been demonstrated in numerous instances, particularly by urologists (Sweetser), that certain radiographic findings may be helpful in demonstrating a tumor to be retroperitoneal (Windholz): these large masses displace the ureters, the bowel, and the stomach (figs. 1, 2). These pressure phenomena may result in various paresthesias, with

radiation of pain often to the thighs and legs, and with blockage and thrombosis of veins such as the iliacs or inferior vena cava, which in turn result in edema of the lower extremities and occasionally ascites. These tumors may extend into the mesentery and the large and small intestines, as well as over the promontory of the sacrum, into the pelvic connective tissues, and into the sigmoid mesocolon. They may grow through the inguinal canal into the labium and buttock (Delamater) and even through the obturator foramen. They frequently obstruct ureters and can cause hydronephrosis and renal failure.

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Sweetser, T. H. Retroperitoneal tumors influencing the kidneys and ureters. J. Urol., 47:619–631, 1942.

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Classification

There has never been a satisfactory classification of the retroperitoneal tumors. The following is suggested as a tentative classification:

I. Tumors of mesodermal origin
  A. Tumors arising from adipose tissue
    1. Benign—lipoma
    2. Malignant—liposarcoma
  B. Tumors arising from smooth muscle
    1. Benign—leiomyoma
    2. Malignant—leiomyosarcoma
  C. Tumors arising from connective tissue
    1. Benign—fibroma (not reported)
    2. Malignant—fibrosarcoma
  D. Tumors arising from striated muscle
    1. Benign—rhabdomyoma (not reported)
    2. Malignant—rhabdomyosarcoma
  E. Tumors arising from lymph vessels
    1. Benign—lymphangioma
    2. Malignant—lymphangiosarcoma (unknown)
  F. Tumors arising from blood vessels
    1. Hemangioma and angiosarcoma
    2. Hemangiopericytoma, benign and malignant
  G. Tumors arising from primitive mesenchyme
    1. Myxoma—myxosarcoma
  H. Tumors of uncertain origin
    1. Xanthogranuloma

II. Tumors of neurogenous origin
  A. Tumors of nerve sheath origin
    1. Nonencapsulated neurofibroma
    2. Encapsulated neurilemoma
    3. Malignant schwannoma
  B. Tumors arising from sympathetic nervous system
    1. Ganglioneuroma
    2. Sympathicoblastoma
    3. Neuroblastoma
  C. Tumors arising from heterotopic cortical adrenal and from chromaffin tissue
    1. Carcinoma arising from cortical adrenal tissue
    2. Malignant nonchromaffin paraganglioma
    3. Paraganglioma
    4. Functioning pheochromocytoma
III. Remnants of renal blastema
IV. Tumors arising from embryonic remnants
  A. Benign and malignant teratomas
  B. Chordomas
V. Metastatic malignant tumors

As can be seen from the classification, there are many different types of tumors which are naturally related to the great variation in the types of tissues found in the retroperitoneal space. Complete descriptions of practically all of these tumors may be found in individual fascicles: for instance, Stout has discussed the soft tissue and neurogenous sarcomas,* Karsner the adrenal neoplasms,** and Willis the teratoma.***

We have tried to avoid repetition of material presented by other authors, but the various tumors of this area are illustrated here, and their important gross and microscopic features briefly described; any difference in appearance because of location has been stressed.

TUMORS OF MESODERMAL ORIGIN

TUMORS ARISING FROM ADIPOSE TISSUE

Benign fatty tumors, **lipomas,** are infrequent. DeWeerd and Dockerty reported 43 retroperitoneal fatty tumors of which 28 were malignant and 15

*Fascicle 5, "Tumors of the Soft Tissues," and Fascicle 6, "Tumors of the Peripheral Nervous System."

**Fascicle 29, "Tumors of the Adrenal."

***Fascicle 9, "Teratomas."

PLATE I

LIPOSARCOMA

A. Enormous retroperitoneal bosselated liposarcoma, in situ. This case illustrates the difficulty of adequately resecting such a neoplasm. A. F. I. P. Acc. No. 219023–26.

B. Lobulated bright yellow liposarcoma. A. F. I. P. Acc. No. 219023–27.

were benign. Of 21 of the malignant group that had been surgically resected, only one was apparently cured.

Liposarcomas are probably the most common neoplasms of the retroperitoneal region. These tumors grow to an extremely large size: the record weight was reported by Delamater with a lipoma of about 275 pounds; Wells reported one of 69 pounds. These tumors occur about equally in males and females, usually in the older age groups, but they can appear in children (Kretschmer). These liposarcomas form lobulated masses of variable consistency (pl. I–A, B). Their surface may have the appearance of cerebral convolutions and on section may resemble brain tissue. More than half have a mucoid yellow slimy appearance so that grossly they are mistaken for myxoma. Such large neoplasms may form multiple masses; and, although they are frequently easy to shell out, they often have fine ramifications that are impossible to remove (figs. 3, 4). These fatty malignant tumors have a predilection for the perirenal area. Their intimate relation to the kidney may necessitate its removal. At times, liposarcomas of the retroperitoneal area are associated with other independent liposarcomas (Ackerman; Siegmund) or with benign fatty tumors (Starkloff, Saxton, and Johnson). Microscopically, these tumors show all the variations that can be seen in any group of liposarcomas (fig. 5). Some are well differentiated, others poorly differentiated; and the gradations from well differentiated to poorly differentiated may be found in the same tumor. The well differentiated tumors are frequently incorrectly diagnosed as lipomas, and often the medical literature has had reports of a large lipoma of the retroperitoneal space that was removed but later recurred and became malignant (Hosemann; von Wahlendorf). In practically all instances, however, if the tumor is malignant it is malignant from the start, and only under exceptional circumstances does it arise from a lipoma (Wright). The treatment of these tumors is surgical; but it is often difficult to remove them completely, and therefore recurrences appear. These liposarcomas, however, are to some extent radiosensitive (Stout, 1944); this may be related to their histogenesis (Wells).

TUMORS ARISING FROM SMOOTH MUSCLE

Leiomyomas are extremely rare, unless those of uterine origin are considered; the latter can form large retroperitoneal masses. Lindeman reported a 49-pound cystic leiomyoma arising from a pedicle behind the left uterine horn between the layers of the left broad ligament.

Leiomyosarcomas appear to be next in frequency to tumors of fatty origin. The malignant smooth muscle tumors often form large masses which may infiltrate and become fixed to surrounding structures, and may have pseudoencapsulation and central cystic degeneration (Lumb; figs. 6, 7). They are firmer than the lipsarcoma and may present a surface pattern suggesting smooth muscle