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UPDATE IN MANAGEMENT OF RETROPERITONEAL NEOPLASMS

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Thesis

Submitted in Partial Fulfillment for The Requirements Of
MD Degree *in General Surgery*

2003

الملخص العربى

تعتبر أورام ما خلف الغشاء البريتونى من الأمراض النادرة وتتميز بسرعة النمو وإرتفاع نسبة الإرتجاع بعد الأستئصال 0 والأعراض المصاحبة لها غالبا ما تكون مشابهة للأمراض الأخرى كأمراض الجهاز الهضمى والجهاز البولى والتناسلى والجهاز العضلى وهى كثيرا ما تخدع الأطباء عند التشخيص0

ويعتبر التشخيص عن طريق الأشعة المقطعية والرنين المغناطيسى هو الأمثل فى مثل هذه الحالات 0 ولقد أثبتت هذه الدراسة أن الأستئصال الجراحى الجذرى هو العلاج الأمثل لمثل هذه الأورام وفى بعض هذه الحالات نحتاج لأستئصال بعض الأعضاء القريبة من الورم مثل الكليتين والطحال وأجزاء من الأمعاء الدقيقة والغليظة حتى نحصل على أعلى قدر من الشفاء0 وأثبتت هذه الدراسة أيضا أن الحالات المتقدمة والحالات المرتجعه لمثل هذه الأورام لا يزال العلاج الجراحى سواء بالأستئصال الجذرى أو شبة الكامل هو أفضل الطرق للعلاج0 بينما وجد أن العلاج الكيماوى والإشعاعى لا يزال غير مجدى فى علاج هذه الأورام0

Anatomy of Retroperitoneal Space

The retroperitoneal space is the area of the posterior abdominal wall, which is located between the posterior parietal peritoneum and the posterior part of the transversalis fascia. Within this space are embryologically related organs such as the adrenal glands, kidneys, ureters, and pancreas. Also, within the retroperitoneal space is the neurovascular apparatus, formed by the aorta and its branches, the IVC (IVC) and its tributaries, the lymphatic vessels, lymph nodes, the lumbar plexus with its branches, and the sympathetic trunk *(Karakousis et al., 1995).*

The retroperitoneal space is covered anteriorly by parietal peritoneum and posteriorly by the transversalis fascia. The retroperitoneal space extends from the 12th thoracic vertebra and the 12th rib superiorly, to the base of the sacrum, the iliac crest, and the pelvic diaphragm inferiorly. Its lateral borders extend from an imaginary line drawn from the tip of the 12th rib down to the junction of the anterior and posterior halves of the iliac crest *(Neifeld et al., 1992).*

The parietal peritoneum is in continuity with the visceral peritoneum and vice versa. Because the parietal peritoneum is not fixed or fused, it can be readily dissected; only the transversalis fascia is fixed, as it is fused with the sub-diaphragmatic fascia superiolrly, and with the psoas fascia and the fascia of the quadratus lumborum muscle laterally to form the anterior lamina of the lumbosacral fascia. The transversalis fascia attaches medially to the vertebral spinous processes and inferiorly it continues with the iliac fossa and the fascia of the pelvic diaphragm *(Alvarenga et al., 2001).*

Several separate spaces or compartments can be defined within the retroperitoneal space. The fascial layers and the spaces in the renal area, from anterior to posterior, are the parietal peritoneum, the anterior pararenal space (fat), the anterior lamina of the renal (Gerota's) fascia, the perirenal space (fat, adrenal, kidney, ureter), the posterior lamina of Gerota's fascia, the posterior pararenal space (fat), and the lumbosacral fascia and the fascia of the psoas muscle *(Cohan et al., 1988).*

There are three distinct layers of structures posterior to the peritoneal cavity, each with its own blood and lymph vessels and nerves. From behind forwards these are:

1) The posterior abdominal wall consisting of the verterbal column and the muscles attached to it. It contains the lumbar vessels and nerves.
2) The kidneys.
3) The unpaired viscera (duodenum, pancreas, ascending and descending colon), which are adherent to the posterior wall of the peritoneal cavity.

THE RETROPERITONEAL VISCERA:

The Kidneys:

The renal fascia covers the fat of the anterior and posterior surfaces of the kidney, having some fixation medially with the adventitial coverings of the renal vessels, with extension to the aorta on the left and the IVC on the right. Superiorly and towards the adrenal glands and diaphragm, the anterior and posterior laminae unite and join the sub-diaphragmatic fascia. However, at the upper pole of the kidney, the fascia separates the adrenal gland from the kidney *(Cohan et al., 1988).*

Duodenum

It is approximately 25 cm long, bent in a C-shaped curve. Its concavity faces upwards and to the left and it is filled by the pancreas. It lies astride the vertebal column, and extends posteriorly to the medial aspect of the right kidney in the paraverterbal gutter. The superior part of the duodenum passes upwards, backwards, and to the right from the pylorus. Superior to the pancreas, it lies anterior to the portal vein and the IVC, and then crosses their right sides.

The descending part is 8 cm long and has no mesentery. It lies directly on the medial part of the right kidney and on the psoas major muscle (anterior to the renal vessels and ureter) down to the level of the third lumbar vertebra. Medially, it is applied to the head of the pancreas, and two-thirds of the way along its length, the bile and the main pancreatic ducts enter its posteromedial aspect together.

The horizontal part, also adherent to the posterior abdominal

wall, is nearly 10 cm long. It passes horizontally to the left, inferior to the pancreas and anterior to the right psoas muscle and verterbal column and the structures lying on them (i.e.; the right ureter and testicular or ovarian artery, the IVC, and the aorta with the inferior mesentric artery arising from it). The anterior and inferior surfaces are covered with peritoneum except where the root of the mesentry containing the superior mesentric vessels crosses it anteriorly.

The short ascending part passes upwards on the left psoas muscle to the left of the aorta and the head of the pancreas, lying anterior to the left sympathetic trunk and the testicular or ovarian artery. It bends anteriorly (duodenojejunal flexure) 2-3 cm to the left of the median plane at the level of the second lumbar vertebra *(Romanberg, 1989).*

Pancreas

It lies across the upper part of the posterior abdominal wall from the duodenum to the spleen, at the junction of the supracolic and infracolic compartments of the peritoneal cavity.

The expanded head lies in the concavity of the duodenum, overlapping the descending and horizontal parts. The head is anterior to the IVC, to the bile-duct and the aorta. The head is crossed by the transverse colon or its mesentery anteriorly, and by the first 2-3 cm of the duodenum superiorly, and joins the body, anterior to the formation of the portal vein. The body passes to the left across the aorta, the left crus of the diaphragm, the psoas major muscle, and the left renal vessels and the kidney. It lies posterior to the omental bursa and the stomach, but its tuber omentale is in contact with the lesser omentum immediately inferior to the celiac trunk, whence the splenic artery runs along its upper margin. The splenic vein lies on its posterior surface and it is joined by the inferior and the superior mesentric veins. The blunt end of the body, the tail, lies in the lienorenal ligament and it may touch the hilus of the spleen *(Karakousis et al., 1995).*

Ascending Colon

The ascending colon is 12-20 cm long. It begins in the right iliac fossa at the entry of the ileum. It ascends on the iliacus, the iliac crest, and the quadratus lumborum in the paraverterbal gutter, crossing the

lateral cutaneous nerve of the thigh, and the ilioinguinal and iliohypogastric nerves. It ends in the right flexure, which turns sharply to the left on the lower part of the right kidney, posterior to the liver. The pertioneum covers the front and sides of the ascending colon and binds it to the posterior abdominal wall *(Alvarenga et al., 2001).*

Descending Colon

The descending colon passes from the left flexure to the margin of the superior aperture of the pelvis near the inguinal ligament. It is attached by the peritoneum to the posterior abdominal wall in the left paraverterbal gutter and the iliac fossa. At first anterior to the lateral surface of the left kidney and medial to the diaphragm, it descends on the transversus abdominis and the quadratus lumborum to the iliac crest, anterior to the same nerves as the ascending colon. It continues in the left iliac fossa on the iliacus to the anterior superior iliac spine. Here, it turns medially, superior to the inguinal ligament, and lies on the femoral nerve, the psoas muscle, the gonadal vessels, and the genitofemoral nerve, and joins the sigmoid colon anterior to the external iliac vessels *(Karakousis et al., 1995).*

For all practical purposes, the psoas muscle extends from the posterior mediastinum to the thigh. On its way downwards, it is closely associated with the perirenal space and the posterior pararenal space *(Alvarenga et al., 2001).*

The iliac fossa is lined by peritoneum. It continues medially to the retroperitoneal space, then downwards to the pelvic wall and to the anterior abdominal wall. It is the home of iliac vessels, ureters, genitofemoral nerves, gonadal vessels, and lymphatic vessels and nodes. Its floor is the iliacus muscle covered by the iliac fascia attached to brim of pelvis or to the linea terminalis after covering the psoas major muscle, inferiorly it is attached to the inguinal ligament together with the transversalis fascia *(Karakousis et al., 1995).*

THE ABDOMINAL AORTA

It descends on the vertebral column from the aortic hiatus in the diaphargm at the level of the 12th thoracic vertebra to its bifurcation into the common iliac vessels at the level of the fourth lumbar vertebra.

The aorta lies on the anterior longitudinal ligament and on the left lumbar veins, which pass transeversely to the IVC on its right side. Superiorly, it lies between the crura of the diaphragm; inferiorly it lies between the sympathetic trunks with the ascending part of the duodenum and coils of the jejunum on its left side.

Anterior to the aorta, from above downwards, are **(1)** the pancreas and splenic vein, partly separated from the aorta by the superior mesentric artery; **(2)** the left renal vein that lies between the aorta and the superior mesentric artery; **(3)** the horizontal part of the duodenum; **(4)** the root of the mesentry; and **(5)** the peritoneum separating the aorta from the coils of the small intestine.

There are three sets of aortic branches that include **(1)** the unpaired ventral branches; **(2)** the paired lateral branches to the suprarenal glands, the kidneys, and the ovaries or testes; and **(3)** the paired posterolateral branches (lumbar arteries) to the abdominal wall.

The Unpaired Branches
The unpaired branches are the celiac trunk, the superior and inferior mesentric arteries, and the median sacral artery. The median sacral artery arises from the posterior surface of the aorta immediately above the bifurcation. It descends on the vertebral column, in the median plane, to end in a series of arteriovenous anastomoses in the cellular coccygeal body, on the front of the coccyx.

The Paired Branches
The Inferior Phrenic Arteries
It pass superolaterally over the crura of the diaphragm. They send many small superior suprarenal arteries.

The Middle Suprarenal Arteries
They arise near the origin of the superior mesentric artery. The suprarenal veins, one on each side, drain respectively into the left renal vein and the inferior vena cava. Occasionally the left vein passes across the anterior surface of the aorta to the inferior vena cava.

The Renal Arteries
They arise at the level of the upper part of the second lumbar

vertebra, and cross the corresponding crus of the diaphragm and the psoas muscle to the kidneys. The right renal artery passes posterior to the IVC and the right renal vein; the left is posterior to its vein.

An accessory renal artery is not uncommon. It usually arises from the lower part of the aorta, passes anterior to the ureter (and IVC if right-sided), and enters the anteroinferior part of the kidney.

Testicular and Ovarian Arteries
They arises from the front of the aorta a short distance inferior to the renal arteries. Between the ureter lying on the posterior abdominal wall and the intestines and mesentric vessels, to reach the corresponding deep inguinal ring and enter the inguinal canal.

Lumbar Arteries
The upper four pairs arise from the posterior surface of the abdominal aorta on the lumbar vertebral bodies. They curve over the vertebral bodies passing deep to the sympathetic trunk (and to the cisterna chyli and IVC on the right) and the fibrous arches of origin of the psoas muscle. The upper one or two arteries also lie deep to the azygos or hemiazygos veins and the crura of the diaphragm. At the root of the transverse process, each artery gives off a posterior branch, then passes posterior to the quadratus lumborum muscle to end as a number of small branches between the transversus abdominis and internal oblique muscles. The posterior branch accompanies the dorsal ramus of the spinal nerve. It sends a spinal branch through the interverterbal foramen to the contents of the vertebral canal, and ends in the erector spinae muscle and the overlying skin. A small fifth pair of lumbar arteries arise from the median sacral artery or from the iliolumbar arteries.

Common Iliac Arteries
These terminal branches of the abdominal aorta arise on the anterior surface of the fourth lumbar vertebra. Each passes inferolaterally to the superior surface of the sacro-iliac joint, where it divides into the internal and the external iliac arteries. Both common iliac arteries lie anterior to the corresponding sympathetic trunk.

External Iliac Arteries

They begin immediately anterior to the sacro-iliac joints as the direct continuation of the common iliac arteries. They pass inferolaterally along the margins of the superior aperature of the lesser pelvis, at first medial and then anterior to the psoas muscle. Each artery is at first anterior and then lateral to its vein, and it is crossed proximally by the ureter and distally by the ductus deferens (or the round ligament of the uterus) and the deep circumflex iliac vein.

Internal Iliac Arteries

They begins medial to the psoas major muscle and anterior to the sacro-iliac joint, at the level of the lumbosacral intervertebral disc. They pass posteriorly into the lesser pelvis medial to the external iliac vein and the obturator nerve, and immediately lateral to the pelvic peritoneum. It lies between the ureter inferiorly and the internal iliac vein superiorly and usually ends in the anterior and the posterior divisions at the upper margin of the greater sciatic notch. The posterior division gives off the superior gluteal artery, the iliolumbar artery, and the lateral sacral arteries. The anterior division gives off the inferior gluteal artery, the obturator artery, the internal pudendal artery, the umbilical artery, the inferior vesical artery, the uterine artery, and the middle rectal artery *(Romanberg, 1989).*

THE INFERIOR VENA CAVA (IVC)

It is the widest vein of the body. It drains venous blood from the lower limbs, most of the abdominal wall, the urogential apparatus, and the suprarenal glands. The venous blood from the remaining abdominal viscera drains to the liver in the portal vein, and reaches the IVC only after it has traversed the liver.

The IVC begins by the union of the two common iliac veins on the front of the fifth lumbar vertebra, posterior to the right common iliac artery. It ascends to the right of the median plane, at first between the aorta and the right ureter, and anterior to the vertebral column, and the right psoas muscle, the sympathetic trunk, and the lumbar arteries. It then arches forwards on the right crus of the diaphragm, anterior to the right renal artery, the right celiac ganglion, and the middle suprarenal artery, to reach a deep groove in the liver between the right

and caudate lobes. Here, it lies anterior to the medial part of the right suprarental gland and the posterior part of the diaphragm.

Anterior to the IVC as it ascends are: **(1)** the superior mesenetric vessels in the root of the mesentery; **(2)** the ileocolic and right colic vessels; **(3)** the horizontal part of the duodenum and the right testicular or ovarian artery; **(4)** the head of the pancreas and the bile duct; **(5)** the portal vein, at first posterior to the superior part of the duodenum, and then in the lesser omentum anterior to the epiploic foramen; **(6)** the liver tissue between the right and caudate lobes.

Renal Veins
They enter the IVC on the right side of the median plane. The right vein passes posterior to the descending part of the duodenum, and it may be overlapped by the right margin of the head of the pancreas. The left renal vein passes to the right, posterior to the inferior border of the pancreas and to the inferior mesentric vein as it ascends to the splenic vein, and crosses the median plane in the angle between the aorta and the superior mesentric artery.

Lumbar Veins
They accompany the lumbar arteries. The first and second veins join the ascending lumbar vein on the lumbar transverse processes. The third and fourth veins enter the IVC, the left vessels passing posterior to the aorta. The fifth vein enters the iliolumbar vein. The ascending lumbar vein unites the lateral sacral, iliolumbar, and upper four lumbar veins to the subcostal vein. It ascends in psoas major anterior to the lumbar transverse processes. The major tributaries are the communications through the interverterbal foramina with the internal verterbal venous plexuses.

Pelvic Venous Plexuses
The veins of the pelvis form a number of important intercommunicating plexuses, which are difficult to dissect. The rectal venous plexuses lie on the surface of the rectum and in its submucosa. They drain through the superior, middle, and inferior rectal veins. In the male, the vesical venous plexus is principally found on the base of the bladder and it drains through the inferior vesical veins to the internal iliac veins. The prostatic venous plexus lies on the front and

sides of the prostate within its fascial sheath. In the female, the vesical plexus surrounds the pelvic part of the urethra and the neck of the bladder. The uterine plexus lies principally at the sides of the uterus between the layers of the broad ligament.

External Iliac Veins

Each ascends with its artery, passing from its medial to its posterior surface, and being crossed by the same structures.

Common Iliac Veins

These veins begin on the medial surface of the psoas muscle by the union of the internal and external iliac veins. The right vein lies posterior to the corresponding artery. The left vein lies inferomedial to its artery and runs a longer course to reach the IVC *(Romanberg, 1989).*

RETROPERITONEAL LYMPH NODES

Retroperitoneal lymph nodes form a rich and extensive chain from the inguinal ligament to the diaphragm and the posterior mediastinal nodes. The nodes are classified as follows:

Aortic group of lymph nodes
1) One to three celiac nodes are located around the base of the celiac artery. They are closely related to the celiac ganglion and lymph nodes of the superior mesenteric artery. These nodes receive lymph from the stomach, liver, pancreas and superior mesenteric nodes.
2) Two or three superior mesenteric nodes receive lymph from the small bowel, right colon, part of the transverse colon, and the pancreas. They communicate with celiac and inferior mesenteric nodes.
3) The two nodes of the inferior mesenteric artery receive lymph from the left colon.
4) The right lateral para-aortic nodes along with the left para-caval nodes form the right lumbar chain of nodes, which may be found around the IVC.
5) The left para-aortic lymph nodes are a group of 5-10 lymph nodes that communicate with the common iliac nodes and drain into the thoracic duct *(Bevilacqua et al., 1991).*

Caval group of lymph nodes

1) Pre-caval lymph nodes are located at the anterior wall of IVC. Two of these nodes are at the level of aortic bifurcation and one at the level of termination of the left renal vein.
2) Retro-caval lymph nodes are located on the psoas muscle and right crus of the diaphragm.
3) The right para-caval nodes are found at the right lateral side of IVC.
4) The left para-caval nodes are in close association with right aortic nodes *(Bevilacqua et al., 1991)*.

Pelvic group of lymph nodes

1) There are 4-6 common iliac lymph nodes located around the iliac artery.
2) There are 8-10 external iliac lymph nodes located laterally, medially, and occasionally anteriorly.
3) The internal iliac lymph nodes are located around the internal iliac vessels.
4) There are one or two obturator lymph nodes, which are located at the obturator foramen close to the obturator neurovascular apparatus.
5) The sacral lymph nodes are located close to the median and lateral sacral vessels.
6) The cisterna chyli is formed at the level of approximately second lumbar vertebra by the confluence of the right and the left lumbar lymphatic trunks with the intestinal trunk *(Karakousis et al., 1995)*.

RETROPERITONEAL NERVES

Six nerves are present in the retroperitoneal space. All are branches of the lumbar plexus, which is formed by a branch of 12th thoracic nerve and by the anterior primary division of the first through fourth lumbar nerves.

1) **Iliohypogastric nerve** (T12 and L1 or L1 only). It emerges from the lateral border of the psoas muscle. After relating to the quadratus lumborum muscle, it travels downwards between the internal oblique and transversus abdominis muscles. It has two branches, the lateral cutaneous nerve that supplies the posterolateral skin of the gluteal area, and the anterior cutaneous nerve that supplies the skin over the symphysis pubis.

2) **Ilioinguinal nerve** (L1). It has the same pathway as iliohypogastric nerve. It transverses the inguinal canal together with the spermatic cord. In the thigh, it innervates the triangle of Scarpa, and part of the scrotal or labia major skin.

3) **Genitofemoral nerve** (L1–2). It pierces the psoas muscle anteriorly. Its genital branch, within the inguinal canal, is related to the iliopubic tract and supplies the cremaster muscle and part of the scrotal skin. Its femoral branch passes below the inguinal ligament and innervates the skin of the triangle of Scarpa.

4) **The lateral femoral cutaneous nerve** (L2–3). It emerges from the lateral border of the psoas muscle at the area of L4 vertebra. It perforates the inguinal ligament close to the anterior superior iliac spine and passes into the lateral aspect of the thigh.

5) **The obturator nerve** (L2–4). It emerges from the medial border of the psoas muscle. It enters the obturator foramen with the obturator vessels downwards to innervate the medial part of the thigh.

6) **The femoral nerve** (L2–4). It emerges from the lateral border of the psoas muscle. It passes under the inguinal ligament and is closely associated with the iliopsoas muscle.

7) **The lumbar sympathetic chain** lies on both sides along the medial border of the psoas muscle. It is located anterior to the lumbar vertebrae and covered by IVC on right and right para-aortic nodes on the left. It is formed by four ganglia, which vary in size and position. These ganglia communicate with each other and with thoracic trunk above and the pelvic trunk below. Each trunk consists of a series of ganglia united by nerve fibres. The cells in the ganglia send their postganglionic fibres principally to the spinal nerves (grey rami communicantes) for the supply of the body wall. Bundles of preganglionic fibres, which enter each trunk through the white rami communicantes, extend along it and end on the cells of the ganglia at various levels. They also pass directly through the trunk and descend in front of it, as splanchnic nerves, to the visceral plexuses and ganglia on the aorta and its branches at muh lower levels *(Karakousis et al., 1995)*.

Pathology

Retroperitoneal STS are uncommon tumors comprising 10% to 20% of all STS in adults. Of the malignancies arising in retroperitoneal space, STS represents 40% to 70%, lymphoma forms 15% to 30%, and less common tumors such as paragangliomas and malignant teratomas account for the remainder *(Lewis et al., 2002).*

HISTOLOGICAL CLASSIFICATION OF SOFT TISSUE TUMOR

The most frequent histologic types of retroperitoneal sarcoma are liposarcomas (41%) and leiomyosarcoma (27%) followed by malignant fibrous histiocytoma (7%), fibrosarcoma (6%), and haemangiopericytoma (4%). Less commonly, malignant peripheral nerve tumor (3%), lymphangiosarcoma, rhabdomyosarcoma, haemangioendothilioma. Rare types (14%) *(Lewis et al., 2002):*

Fibrous tumors
1) Benign tumors
   a. Nodular fasciitis (including intravascular and cranial types)
   b. Proliferative fasciitis and myositis
   c. Elastofibroma
   d. Calcifying aponeurotic fibroma
   e. Fibrous hamartoma of infancy
   f. Fibromatosis colli
   g. Infantile digital fibromatosis
2) Fibromatoses
   a. Intraabdominal fibromatosis (intraabdominal desmoid)
   b. Mesenteric fibromatosis (including Gardner's syndrome)
   c. Infantile (desmoid-type) fibromatosis
3) Malignant tumors (Fibrosarcoma)
   a. Adult fibrosarcoma
   b. Congenital or infantile fibrosarcoma
   c. Inflammatory fibrosarcoma (inflammatory myofibroblastic tumor)

Fibrohistiocytic tumors
1) Benign tumors

   a. Fibrous histiocytoma
      i. Deep fibrous histiocytoma