

Clinicopathological features and surgical management of pelvic soft tissue sarcoma

Thesis

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Abstract
Introduction,
and Aim of the work

Abstract

Background

Soft tissue sarcomas (STS) of the true pelvis are rare tumors and there is little information in the literature related to management of these tumors.

Purpose

The aim of this retrospective study was to review the literature regarding the clinicopathological features and current management of pelvic soft tissue sarcomas and to review the surgical experience of the National Cancer Institute in management of pelvic STS over the last 10 years.

Twenty seven consecutive patients presenting between January 1997 and December 2006 with soft tissue sarcomas involving the true pelvis had undergone surgical resection in the Surgical Department of the National Cancer Institute, Cairo University. The data of these cases were collected and analysed.

Results:

Of the 27 patients, sixteen cases were males and eleven cases were females. The median age was 53.7 (range 12-65) years at presentation. The mean duration between the onset of symptoms and diagnosis was 8 months (range 1 month-48 months). The most frequent clinical presentation was abdominal or perineal pain representing 77.7% of all cases. The commonest pathological 2 types in our study were MFH (7 cases) and liposarcoma (6 cases). Cross-sectional imaging was reviewed for each patient to determine the anatomical location of the lesions. Only six cases in our study were limited to the true pelvis. Most of the cases

were extending to the false pelvis (16 cases). Five cases were extending to the thigh. Only one case was extending to the perineum. External hemipelvectomy was performed in only one case, internal hemipelvectomy was done in twelve cases, lower partial sacral resection was done in two cases, resection of the external iliac artery was done in two cases and visceral resection was done in six cases. The most commonly used approach was the abdominoinguinal approach (51.8% of cases). No perioperative mortality was recorded and the incidence of perioperative complications was 44.4%. The commonest encountered complication was wound infection (6 cases). Adjuvant radiotherapy was given in 25 cases, while adjuvant chemotherapy was given in 15 cases. The median time between operation and date of last follow up was 39 months (range 10-84 months). At last follow-up, 16 patients had died, 4 cases were alive with evidence of disease, and 7 cases were free of disease. Ten patients developed local tumor recurrence (37% of all cases). Seventeen patients developed distant metastases after treatment. The overall 3 years survival was 59.2%, and the overall 3 years disease free survival was 25.9%.

Conclusions:

Soft tissue sarcoma of the pelvis is fortunately a rare disease. Anatomical consideration and the often extensive size of these tumors represent a challenge to proper treatment. It is generally agreed in the literature that complete surgical resection is the most important factor for cure or prolonged survival. Surgeons must be acquainted with the different approaches and reconstruction techniques to achieve the best results with least morbidity.

Introduction

Soft tissue sarcomas that arise in the retro peritoneum or in the pelvic cavity are relatively rare tumors that constitute approximately 20-25% of all soft tissue sarcomas. **(Van Dalen et al., 2004)**

Most of the studies in the literature present results of management of abdominal retroperitoneal sarcomas and few studies focused on the group of soft tissue sarcomas that affect the true pelvis. **(Clark et al., 2005)**

Patients commonly present with a large, non-tender abdominal mass. Often, the symptoms are non-specific abdominal discomfort and a feeling of fullness, and approximately half of the patients will report pain. Because of their anatomic location and the non-specific symptom, tumors in the retroperitoneum often grow to a considerable size before they are diagnosed. On the other hand, sarcomas arising in the gastrointestinal tract, urinary tract and gynecological organs are more likely to give earlier symptoms and patients often present with abdominal pain, bleeding or obstruction. **(Clark et al., 2005)**

Pelvic organ sarcomas are usually not grouped with retroperitoneal or intraabdominal sarcomas in the literature. **(Cormier& Pollock, 2004)** For most patients the prognosis of retroperitoneal and pelvic soft tissue sarcoma has historically been poor. Overall survival in most retrospective series is approximately 55% at five years with a range of 30-75%. Recurrence after 5 years is not uncommon and despite 5 years disease-free survival, 40% will recur in the subsequent 5 years **(Herman et al., 1999)**.

There are general agreement in the literature that complete surgical resection is the most important factor for cure or prolonged survival.

Furthermore, it has been shown that surgical treatment at a specialised centre, where a relatively greater number of similar cases are treated, confers the patients a significantly better prognosis. Complete resection can also be attained for recurrent disease, but the likelihood of achieving clear margins declines significantly as the number of recurrences increases. **(Lewis et al., 1996) (Cormier, Pollock , 2004)**

Due to anatomical considerations and the often extensive size of the tumors, it is often difficult to obtain adequate margins in the retroperitoneal compartment. Local recurrence is therefore more common than distant metastasis which occurs predominantly to the lung and liver in no more than one third of patients. Extensive locoregional tumors growth is usually the cause of death. **(Herman et al., 1999).**

In addition to complete surgical resection, also malignancy grade (high), tumor size (>10cm), histological subtype (leiomyosarcoma (unfavourable), liposarcoma (favourable), and age (>60 years), have been found to be related to survival (worst) by many groups. **(Pollock et al., 1996) (Pirayesh et al., 2001)**

Although adjuvant radiation reduces the local recurrence rate in extremity and superficial trunk sarcomas, gastrointestinal toxicity often limits the delivery of radiation to the retroperitoneum. Several retrospective studies suggest that adjuvant radiation improves local control after complete macroscopic resection is performed. **(Cody et al., 1981) (Catton et al., 1994).** Other investigators found no benefit in adjuvant radiation therapy for completely resected retroperitoneal sarcomas. **(McGrath et al., 1984) (Jaques et al., 1990) (Heslin et al., 1997) (Pirayesh et al., 2001)**

Retrospective studies had not demonstrated any benefit to neoadjuvant (**Storm et al., 1981**) or adjuvant chemotherapy (**Glenn et al., 1985**) for retroperitoneal or intra abdominal sarcomas. Some authors even suggest a negative effect of chemotherapeutic regimens on survival, with patients receiving treatment showing an increased risk of death. (**Singer et al., 1995**). As with extremity soft tissue sarcoma, there is no available data to support the routine use of adjuvant chemotherapy in these patients. (**Lewis et al., 1996**) (**Joensuu et al., 2001**)

Aim of the work

The aim of this work is to accomplish the following two objectives:

1-Review of the literature regarding the clinicopathological features ¤t management of pelvic soft tissue sarcoma (STS).

2-Retrospective study of cases of pelvic STS surgically treated in the NCI, Cairo University over the last 10 years.

The study will include analysis of the clinical presentations, diagnostic measures, pathological features, surgical approaches, perioperative complications as well as follow up data.

Review of literature

REVIEW OF PELVIC ANATOMY

A.Bony Wall of the pelvis

The **pelvis**, so called from its resemblance to a basin, is a bony ring, interposed between the movable vertebræ of the vertebral column which it supports, and the lower limbs upon which it rests; it is stronger and more massively constructed than the wall of the cranial or thoracic cavities, and is composed of four bones: the two **hip bones** laterally and in front and the **sacrum** and **coccyx** behind.

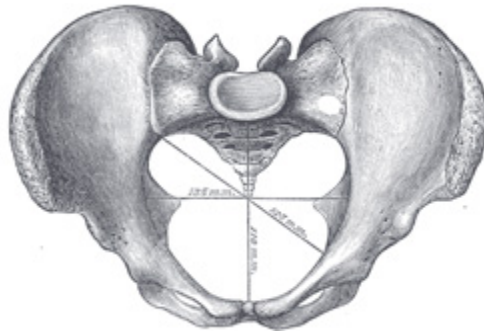
The pelvis is divided by an oblique plane passing through the prominence of the sacrum, the arcuate and pectineal lines, and the upper margin of the symphysis pubis, into the greater and the lesser pelvis. The circumference of this plane is termed the **linea terminalis** or **pelvic brim**.

The Greater or False Pelvis (pelvis major).—The greater pelvis is the expanded portion of the cavity situated above and in front of the pelvic brim. It is bounded on either side by the ilium; in front it is incomplete, presenting a wide interval between the anterior borders of the ilia, which is filled up in the fresh state by the parietes of the abdomen; behind is a deep notch on either side between the ilium and the base of the sacrum. It supports the intestines, and transmits part of their weight to the anterior wall of the abdomen.

The Lesser or True Pelvis (pelvis minor).—The lesser pelvis is that part of the pelvic cavity which is situated below and behind the pelvic brim. Its bony walls are more complete than those of the greater pelvis. For convenience of description, it is divided into an inlet bounded by the superior circumference, and outlet bounded by the inferior circumference, and a cavity.

The Superior Circumference.—The superior circumference forms the brim of the pelvis, the included space being called the superior aperture or inlet(fig 1) . It is formed laterally by the pectineal and arcuate lines, in front by the crests of the pubes, and behind by the anterior margin of the base of the sacrum and sacrovertebral angle. It has three principal diameters: antero-posterior, transverse, and oblique. The anteroposterior or conjugate diameter extends from the sacrovertebral angle to the symphysis pubis; its average measurement is about 110 mm. in the female. The transverse diameter extends across the greatest width of the superior aperture, from the middle of the brim on one side to the same point on the opposite; its average measurement is about 135 mm. in the female. The oblique diameter extends from the iliopectineal eminence of one side to the sacroiliac articulation of

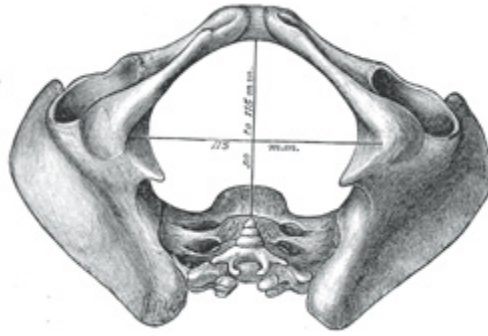
the opposite side; its average measurement is about 125 mm. in the female.



Fig(1)-Diameters of superior aperture of lesser pelvis (female).(quoted from **Gray anatomy: Anatomy of the human body**)

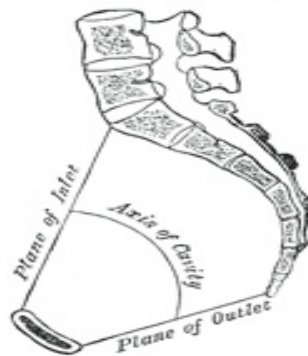
The cavity of the lesser pelvis is bounded in front and below by the pubic symphysis and the superior rami of the pubes; above and behind, by the pelvic surfaces of the sacrum and coccyx, which, curving forward above and below, contract the superior and inferior apertures of the cavity; laterally, by a broad, smooth, quadrangular area of bone, corresponding to the inner surfaces of the body and superior ramus of the ischium and that part of the ilium which is below the arcuate line.. It contains, in the fresh subject, the pelvic colon, rectum, bladder, and some of the organs of generation.. In the female the uterus and vagina occupy the interval between these viscera.

The Lower Circumference.—The lower circumference of the pelvis is very irregular; the space enclosed by it is named the inferior aperture or outlet(fig 2) ² and is bounded behind by the point of the coccyx, and laterally by the ischial tuberosities. These eminences are separated by three notches: one in front, the pubic arch, formed by the convergence of the inferior rami of the ischium and pubis on either side. The other notches, one on either side, are formed by the sacrum and coccyx behind, the ischium in front, and the ilium above; they are called the sciatic notches; in the natural state they are converted into foramina by the sacrotuberous and sacrospinous ligaments. When the ligaments are in situ, the inferior aperture of the pelvis is lozenge-shaped, bounded, in front, by the pubic arcuate ligament and the inferior rami of the pubes and ischia; laterally, by the ischial tuberosities; and behind, by the sacrotuberous ligaments and the tip of the coccyx.



Fig(2)– Diameters of inferior aperture of lesser pelvis (female).).(quoted from **Gray anatomy: Anatomy of the human body**)

The diameters of the outlet of the pelvis are two, antero-posterior and transverse. The antero-posterior diameter extends from the tip of the coccyx to the lower part of the pubic symphysis; its measurement is from 90 to 115 mm. The transverse diameter, measured between the posterior parts of the ischial tuberosities, is about 115 mm(fig 3).



Fig(3)-Median sagittal section of pelvis.).(quoted from **Gray anatomy: Anatomy of the human body**)

(Gray H, 1918. Anatomy of the Human Body, 20th ed.)

B.Ligaments of the pelvis

sacrotuberous and sacrospinous ligaments participate in the formation of the pelvic walls. They also serve to convert the greater and lesser sciatic notches into the greater and lesser sciatic foramina by their attachments to the sacrum and coccyx medially and the ischial tuberosity and ischial spine laterally

Sacrotuberous Ligament

The sacrotuberous ligament originates at the posterior superior iliac spine and the lateral border of the sacrum and coccyx. It inserts upon the ischial tuberosity

Sacrospinous Ligament

The origin of the sacrospinous ligament is the lateral border of the sacrum and coccyx. Insertion is upon the ischial spine.

(Skandalakis et al.,1996).