Clinics in Surgery

Research Article
Published: 12 Feb, 2024

Infrequent Pelvic Non-Visceral Soft Tissue Mesenchymal Tumors: Surgical Techniques for En Bloc Resection and Long-Term Surveillance

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Abstract

Purpose: To assess long-term resections consequences for the sporadic large nonvisceral pelvic soft tissue tumors in a designed series referred to the National Cancer Institute 1998-2020. Main outcome measures are disease free survival, recurrence pattern and salvage.

Patients and Methods: Thirty-one patients (17 females and 14 males) averaged 48 years presented with average 21 cm tumors including 17 (55%) extensions (10-paraanal ischiorectal spaces, 3-vulva, 3-gluteal region (sciatic notch) and 1-femoral triangle) plus 21 (68%) upper abdominal growths. Immunohistochemistry showed Aggressive Angiomyxoid tumors AA (10), fibromatosis (6), Peripheral Nerve Sheath Tumors PNST (6), Solitary Fibrous Tumors SFT (3), leiomyosarcoma (2), liposarcoma (2), one monophasic synovial sarcoma and one undifferentiated sarcoma. Abdominal approach was combined with special perineal incisions to widely en masse resect tumors (primary/recurrence) plus infiltrated viscera.

Results: Pelvic and perineal tumor resection extended to the viscera in 23 (74%) with 77% (R0) and low morbidity (CDC grade I-III). Following 50-m median surveillance, 22/31 (71%) were disease free with relapses in 4/10 of AA (40%) as local perineal and/or pelvic recurrences, amenable to curative salvage resections; while, 4/6 with fibromatosis died of repeated recurrences (19 m-33 m), only 1/3 with SFT died after 21-m, single PNST had resectable local relapse while other 5 were disease free, one liposarcoma patient had resectable recurrence and 2/2 of leiomyosarcomas had systemic spread. Undifferentiated sarcoma patient died of recurrence 24 month; meanwhile, synovial sarcoma male patient was disease free.

Conclusion: Combined approaches enable en block resection and offer alone safe long-term disease-free survival in a reliable percent even for recurrences.

Keywords: Pelvic soft tissue tumor resection; Pelvi-perineal tumor; Vulval tumor; Aggressive angiomyxoma; Pelvic fibromatosis; Pelvic PNST; Pelvic SFT; Pelvic soft tissue sarcoma

Abbreviations

CDC: Clavien Dindo Classification Tool Grading Postoperative Complications; AA: Aggressive Angiomyxoma Tumor; PNST: Peripheral Nerve Sheath Tumor; SFT: Solitary Fibrous Tumor; Abd: Intra-Abdominal Extension

Introduction

In 80% pelvic soft mesenchymal tumors develop in visceral organs such as uterus, vagina, bladder and occasionally rectum but rarely in the retroperitoneal mesenchyme present in the muscular, neural and fibro-vascular wall components. Such tumors are locally aggressive, highly recurrent but less frequently metastatic [1-4]. Nonvisceral tumors usually have large dimensions and sometimes extra pelvic insinuate or frank invasion through anatomical planes. Slowly expanding tumors in a confined pelvic space herniate through the pelvic floor around anorectal segment to reach the ischiorectal space, paravaginal to the labia or other defects as sciatic notch or obturator [1]. These extensions present a surgical challenge and most of the studies are case reports with no consensus regards resection techniques or neoadjuvant down-sizing regimens [1-3].

The high rate of local recurrence noted with the whole pelvic mesenchymal tumors frequently

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Received Date: *19 Jan 2024*
Accepted Date: *06 Feb 2024*
Published Date: *12 Feb 2024*

Citation:
Nassar OAH, Fahim MI, Farahat IG. Infrequent Pelvic Non-Visceral Soft Tissue Mesenchymal Tumors: Surgical Techniques for En Bloc Resection and Long-Term Surveillance. Clin Surg. 2024; 9: 3689.

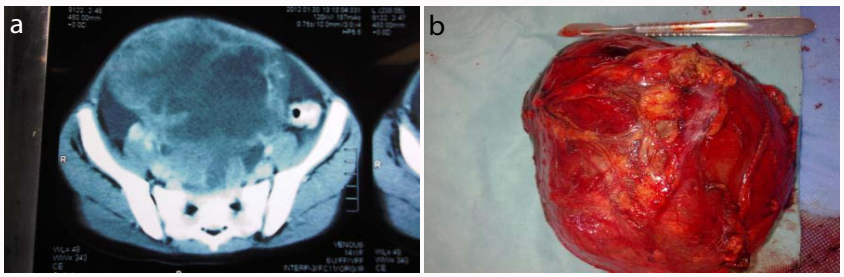

Figure 1: Patient 1 in Table 1. a) Pelvic CT shows schwannoma as heterogeneous pelvic abdominal mass in the rectovaginal space not adherent to uterus or rectum posterior. b) Resected tumor with margin and intact capsule.

necessitates combined surgical approaches and extensive encompass procedures that may involve visceral structures. High grade tumors require more debulking than benign and locally aggressive tumors [2,5]. Reported high grade tumors include Solitary Fibrous Tumor SFT (hemangiopericytoma), angiosarcoma, Malignant Peripheral Nerve Sheath Tumor MPNST, leiomyosarcoma, liposarcoma and synovial sarcoma [1-5]. Locally aggressive and benign tumors include Aggressive Angiomyxoma (AA) as the most frequent histology and albeit non-metastatic, 36% to 72% may relapse locally, low grade PNST, aggressive fibromatosis (desmoid) and low grade SFT. Aberrant extrapelvic herniation is frequently reported with AA. Other tumors; but hardly ever, may present similar clinical features as pelvic mucinous neurofibromas with less vascular component, low recurrence rate, fibromatosis (desmoid tumor) with high risk of local recurrence and liposarcoma [6-12].

Females are significantly more exposed than males and the bulk of tumor growth is often hidden within the pelvis and perineal fat reducing early compression symptoms [7-9]. Our aim was to assess surgical satisfactoriness and safety to offer long-term disease-free survival.

Patients and Methods

Starting with 1998 to 2020, 31 consecutive patients (17 females and 14 males) with pelvic nonvisceral tumors were surgically treated at the National Cancer Institute; Cairo University following an institutional review board approval. Cohort was carefully chosen out of 40 patients with similar tumors. Exclusion criteria were multiple tumors with visceral or bone invasion and synchronous systemic dissemination.

Patients were divided into two group local aggressive (Table 1 and Figures 1-3) and sarcoma (Table 2 and Figure 4, 5). Median age for the total was 48 (17-72) and 42 for the sarcoma group.

Diagnosis

Main presentations were bladder and rectal compression symptoms beside pelvic fullness and pain with average 14 m delay of seeking treatment (5-37). Palpable abdominal extensions were evident in 20 patients (10-20 cm above the pubic bone).

CT and/or MRI were the main localizing, diagnostic and metastatic work up procedure [13]. Working labs were within normal except for mild normocytic normochromic anemia in all ladies, mildly elevated Ca-125, Ca-19-9, CEA and normal levels of α-fetoprotein, β-HCG, Ca-72 & He-4.

Prior guided pathologic diagnoses either trans-abdominal or perineal were evident for 25-cases (spindle cell neoplasm). Other 6-patients had inappropriate perineal biopsy at the referring hospitals assuming reporting clinical diagnosis of lipoma, Bartholin's cyst or chronic infection.

Preparation

Renal back pressure changes and lower ureteric compression in 15 patients indicated ureteric stents; meanwhile, cystoscopies found neither mucosal bladder invasion nor edema. Similarly, colonoscopies didn't reveal mucosal invasion. Vascular embolizations were needed in 4 sarcomas and 7 local aggressive soft tissue tumors.

Sarcomas had neoadjuvant preoperative long-course 40 Gy to 50 Gy/25 fractions, /5 W to downsize. Postoperative pelvic and perineal external beam irradiation was tried in 6 patients with fibromatosis using 30 Gy to 35 Gy/4 W [14].

Technique

Surgery for primary and recurrent pelvic tumors started with abdominal phase to mobilize tumor adhesions away the intestine, sacrum, ureters, bladder prostate, seminal vesicles, vas, nerves and vessels, rectosigmoid and safeguards vascular connections legating main stem internal iliac artery uni/bilaterally according to tumor vascularity and depth of posteriolateral extensions. Resection included adherent pelvic organs and dissection extends to the pelvic floor around tumor to reach the pelvi-perineal tunnel for either perineal or sciatic notch extensions.

Perineal phase was tailored according to directions of tumor extension in Lithotomy or lateral position dissecting the ischiorectal fossa, incising the pelvic floor muscle, releasing anorectal adhesions plus blunt tunnel mobilization to deliver the mass back up to abdominal wound.

For vulval-labial extension, dissection started by wide elliptical labial skin incision around the mass extending up to paravaginal plus ipsilateral ischiorectal spaces ± adherent vaginal wall resection with immediate reconstruction [15]. Regards posterior perineal tumors and tumors extending to lower sacral S4 & S5 or neural plexus; approach was anterior coccygeal retroanal to encompass lower margin, dissect lower nerves, control bleeding and resect adherent sacral segment. For gluteal extension via greater sciatic notch; surgery commenced with gluteal incision (Kocher-Langenbeck) to identify sciatic nerve bundles and roots together with gluteal vessels. Abdominal phase mobilizes the mass out of adhesions with ligation of ipsilateral iliac vessels. Small tumor herniation may be pulled otherwise bone was widened of by Nippler to push the tumor inside the pelvis without mass disconnection [16,17]. Reconstruction included coloanal anastomosis with stoma cover in few cases, bladder wall reconstruction or ileal neobladder for cystectomy, vaginal

Table 1: Benign and local aggressive tumors: Clinicopathological features & Follow-up.

	Sex (age)	Tumor type (size)	Tumor extent (Delay period)	Surgical approaches Infiltration margins Perioperative Morbidity (CDC)	Rec. site (free period)	2nd surgery	Life + Follow-up
1 (Figure 1 A,B)	Female (19)	Schwannoma (8 cm)	Pelvic+Abd (9 m)	-Abdominal -No infiltration -(R0) CDC 0	-	-	Free (36 m)
2	Female (56)	Benign Solitary fibrous tumor (8 cm)	Pelvic (18 m)	-Abdominal -Rectum -(R0)	-	-	Free (75 m)
3	Female (59 y)	Aggressive angiomyxoma (35 cm)	Pelvic+Abd. + Lt para anal (37 m)	-Abdominal +perineal -No infiltration -(R0)	Perineal (16 m)	2 times	Free (58 m)
4	Male (34)	Fibromatosis (20 cm)	Pelvic+Abd (20 m)	-Abdominal -Bladder +ureter -(R0) CDCII	Pelvic (9 m)	2 times CDC- III	Dead (32 m)
5	Female (39 y)	Aggressive angiomyxoma (22 cm)	Pelvic+Abd+ Rt para anal (22 m)	-Abdominal+perineal -Ovary+sigmoid colon -(R0)	-	-	Free (101 m)
6	Male (61)	Fibromatosis (17 cm)	Pelvic+Abd (9 m)	-Abdominal -Intestine+bladder+ vascular -(R1)	Pelvic+peritoneu+ Abd. Wall (8 m)	-	Dead (19 m)
7 (Figure 6 A,B)	Female (22)	Schwannoma cellular version (16 m)	Pelvic (6 m)	-Abdominal -Rectum -(R0)	Pelvic+Abd (34 m)	2 times	Free (68 m)
8	Female (46 y)	Aggressive angiomyxoma (32 cm)	Pelvic +Abd + Lt para anal (19 m)	-Abdominal+perineal -Uterus+Ovary+Bladder -(R0) CDCI	Pelvic (27 m)	2 times CDC-III	Dead (51 m)
9	Female (43 y)	Aggressive angiomyxoma (28 cm)	Pelvic +Abd. + Lt para anal (10 m)	-Abdominal+perineal -Uterus+Ovary -(R0)	-	-	Free (43 m)
10 (Figure 2 A-C)	Female (68 y)	Aggressive angiomyxoma (26 cm)	Pelvic +Rt. Labia (9 m)	-Abdominal+trans labial -Vagina -(R0)	-	-	Free (26 m)
11	Female (18)	Fibromatosis (8 cm)	Pelvic (11 m)	-Abdominal -Uterus+rectum -(R0)	-	-	Free (63 m)
12	Male (71 y)	Fibromatosis (32 cm)	Pelvic+Abd+Rt. para anal (30 m)	-Abdominal+perineal -Iliac artery -(R1)	Pelvic+Peritoneal (8 m)	1time CDC-II	Dead (19 m)
13	Female (72 y)	Aggressive angiomyxoma (30 cm)	Pelvic+Abd. + Lt. labia (11 m)	-Abdominal+trans labial -Vagina -(R1)	-	-	Free (44 m)
14	Male (65 y)	Aggressive angiomyxoma (38 cm)	Pelvic+Abd. + Post anal (9 m)	-Abdominal+trans coccygeal retroanal -lumbosacral n+obturator n. -(R0) CDCIII	Perineal+Pelvic (13 m)	3 times	Free (67 m)
15	Female (33)	Fibromatosis (12 m)	Pelvic (14 m)	-Abdominal -Iliac a+uterus+bladder -(R0)	Pelvic+inguinal	3 times	Dead (33 m)
16	Female (50 y)	Neurofibroma (31 cm)	Pelvic+Abd+Rt. Labia (20 m)	-Abdominal+trans labial -No infiltration -(R0)	-	-	Free (64 m)
17	Female (61 y)	Aggressive angiomyxoma (26 cm)	Pelvic+Rt. para anal (10 m)	-Abdominal+perineal -Uterus -(R0)	-	-	Free (52 m)
18	Male (50)	Aggressive angiomyxoma (30 cm)	Pelvic+Abd+gluteal	-Abdominal+Rt gluteal+trans coccygeal retroanal -Sacrum -(R0) CDCI	-	-	Free (67 m)
19 (Figure 3 A-C)	Female (51 y)	Fibromatosis (37 cm)	Pelvic+Abd. + Lt. para anal (11 m)	-Abdominal+perineal -No infiltration -(R0) CDCI	-	-	Free (55 m)
20	Male (61 y)	Neurofibroma (18 cm)	Pelvic+Rt. para anal (12 m)	-Abdominal+perineal -No infiltration -(R0)	-	-	Free (47 m)
21	Female (60 y)	Aggressive angiomyxoma (19 cm)	Pelvic+Lt. para anal (20 m)	-Abdominal+perineal -Uterus -(R0) CDCII	Perineal (16 m)	1 time	Dead (25 m)
22	Male (42)	Neurofibroma (16 cm)	Pelvic+gluteal (25 m)	Abdominal+ Rt gluteal lumbosacral root n + rectum (R0) CDCIII	-	-	Free (19 m)

-Tumor extension: Abd (abdominal), para-anal (para-anal perineal extension)
-Histopathology: PNST (peripheral nerve sheath tumor or neurofibroma +ve S-100), Aggressive angiomyxoma (+ve CD34, SMA, ER, PR and vimentin). Fibromatosis (desmoid tumor) with -ve CD117 and +ve β-Catenin
-Rec (local recurrence site), free period (disease free period)

reconstruction and vascular grafts.

Patients were scheduled for regular follow-up 6-monthly with abdominal and pelvic MRI. Data analysis applies SPSS win statistical package version-12, median (minimum-maximum) and qualitative as frequency (%).

Results

Perioperative

For 24 operative specimens margins of resection were free (R0) and (R1) for [7]. Resected adherent organs were the uterus, tubes & ovaries in 10-ladies, external iliac vessels in 3+ common femoral in 1, lateral lower vaginal wall in 2-patients, full thickness bladder wall in 6 situations plus 1-total cystectomy and recto-sigmoid colon segment for 7 patients. Ligature transfixions of ipsilateral internal iliac artery were mandatory for 10 and bilateral for other 4-cases to control extensive parasitic neovascular adhesions.

Collectively histological proven tumor infiltrations are, 4/10 (40%) for hysterectomies, 3/10 (30%) ovariectomies, 2/2 vaginectomy, 4/7 equal for both bladder and colorectal resections. Resected bladder and colon segments were superficially infiltrated and resected vessels

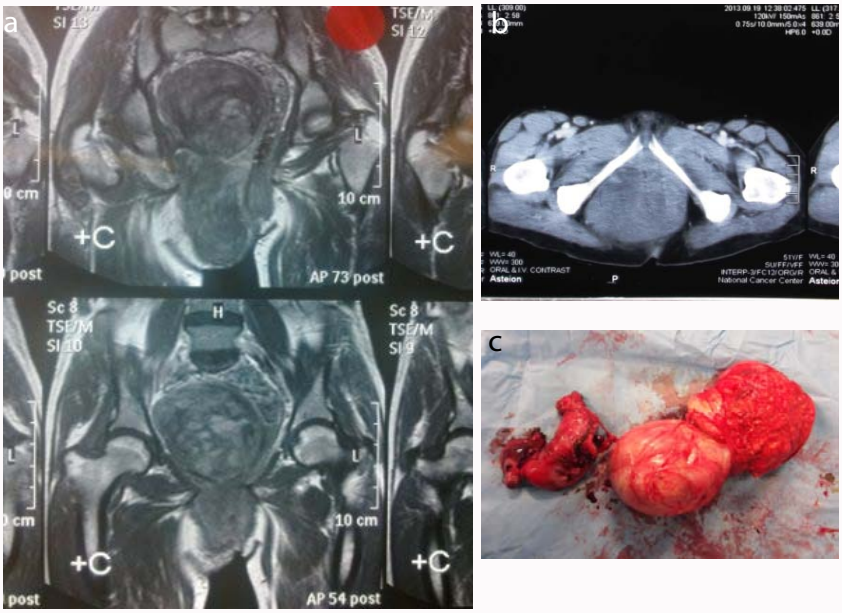

Figure 2: Patient 10 in Table 1. a) MRI of a 68-year lady with AA and Rt. Labial extension. b) CT of tumor herniating through paravaginal pelvic floor. c) Specimen with hysterosalpingo-oophorectomy and vaginal wall.

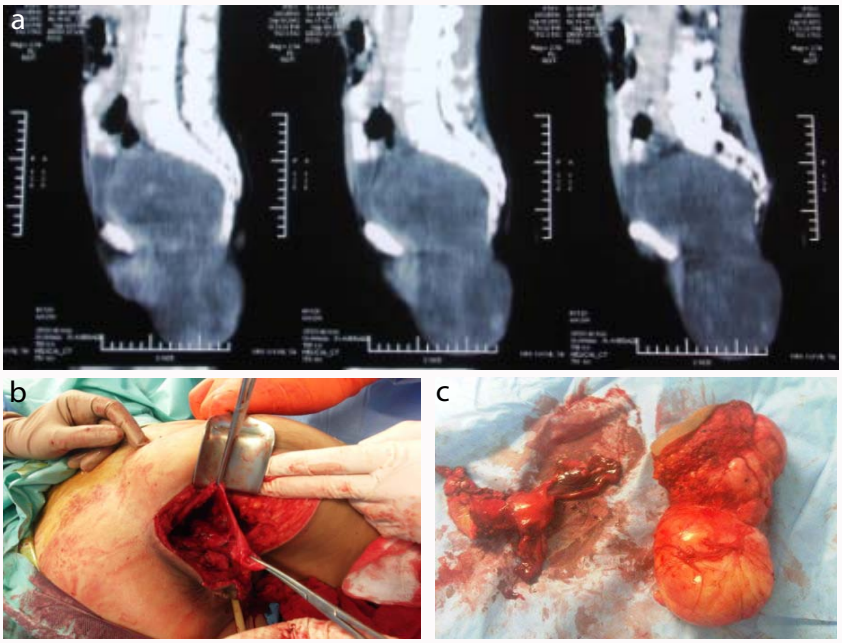

Figure 3: Patient 19 Table 1. a) Helical CT of pelvic perineal tumor to the Lt. Ischiorectal para-anal space. b) Perineal wound at the Lt ischiorectal fossa resecting overlying skin and fat with the tumor in close contact to the anorectal segment. Tumor mass is delivered up to the pelvis. c) Specimen of 37 cm Desmoid (Fibromatosis) with hysterosalpingo oophorectomy and vaginal cuff.

Table 2: Malignant pelvic soft tissue tumors and follow-up.

	Sex (age)	Histopathology Immunohistochemistry Tumor (size)	Tumor extent	Pelvic Rth-	Surgical approach Morbidity (CDC)	Infiltration & Margins	Rec. +(DF)	2nd surgery	Life+ (Follow-up period)
1	Male (72)	Solitary fibrous tumor grade 2 +ve CD34, -ve EMA & S-100 protein (11 cm)	Pelvic+Abd		Abdominal	Intestine+bladder (R0)	-	-	Free (61 m)
2	Female (57)	Liposarcoma, Well differentiated (18 cm)	Pelvic+Abd		Abdominal	No infiltration (R0)	-	-	Free (50 m)
3	Male (61)	Leiomyosarcoma +ve SMA, Desmin (15 cm)	Pelvic+Abd	Rth	Abdominal CDC-I	No infiltration (R0)	Lung (34 m)	metastatectomy	Free (43 m)
4 (Figure 7)	Male (61)	Malignant PNT (neuroepithelioma) +ve S-100 (22 cm)	Pelvic+Abd +Gluteal	Rth	Abdominal+Rt gluteal CDC-II	Obturator n Bladder (R1)	-	-	Free (44 m)
5	Male (17)	Synovial sarcoma +ve Cytokeratin, EMA (9cm)	Pelvic		Abdominal CDC-III	Rectum (R0)	-	-	Free (51 m)
6	Male (34)	Solitary fibrous tumor grade 3 +ve CD34, -ve EMA & S-100 protein (21 cm)	Pelvic+Abd	Rth	Abdominal	Intestine+rectum +vascular (R1)	Lung+pelvic +Liver (11 m)	unresectable	Dead (21 m)
7 (Figure 4 A, B)	Male (40)	Liposarcoma Well differentiated (19 cm)	Pelvic+Abd		Abdominal	No infiltration (R0)	Pelvic (22 m)	1 time	Free (65 m)
8	Male (34)	Leiomyosarcoma +ve SMA, Desmin (10 cm)	Pelvic+Abd		Abdominal+trans coccygeal retroanal CDC-III	Rectum+lower S4&5 (R1)	Peritoneum+liver (9 m)	unresectable	Dead (25 m)
9 (Figure 5 A, B)	Male (30)	Undifferentiated sarcoma (28 cm)	Pelvic+Abd+ femoral area	Rth	Abdominal + Inguino- femoral CDC-II	Bladder+Ureter +ileofemoral vessels (R1)	Pelvic (15 m)	1 time CDC-I	Dead (24 m)

CDC: Clavien-Dindo Complications grades [18];
I-Complication requiring allowed therapeutic regimens without surgical, endoscopic or radiological interventions.
II- More therapeutic regimens ± blood transfusions ± parenteral nutrition.
III-Required surgical, endoscopic or radiological interventions.
IV-Life threatening complication requiring IC/ICU management.
V-Death

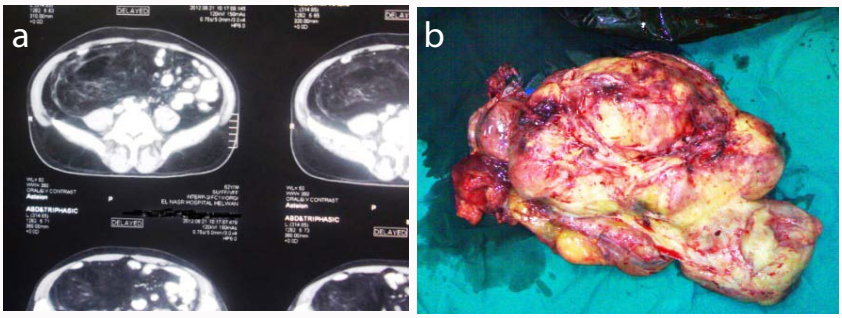

Figure 4: Patient 7 in Table 2. a) CT- 19 cm pelvic abdominal liposarcoma. b) Resected operative specimen of well differentiated liposarcoma.

revealed encasement rather than actual invasion. No evidence of lymphovascular or node invasions appears in the resected specimens.

Mean operative time was 185 minute (147-380) and blood loss averages 550 ml ± 460 ml with 3 (2-6) transfuse units. In-patient stays were 5 days (3-17); with minor CDC grade I & II abdominal wound sepsis in 3 patients [18].

Surveillance

After a median follow up of 50 m (19-101), local aggressive soft tissue tumor patients revealed 9/ 22 (41%) local recurrences after a median disease-free period of 15 months (9-34). In 6 cases local relapses were repetitive including 4 fibromatosis who eventually died in spite of repeated salvage surgery.

Four out of 10 AA tumors (40%) had repeated recurrences in the perineal scar and pelvis 13 m, 16 m & 27 m post-primary surgery. Wide perineal resection for two cases and pelvic tumor resection together with partial cystectomy and segmental rectal resection for another two controlled later recurrences. One AA male patient had (5 cm) perineal recurrence excision 13 m. post original surgery, 7 m later he had the 2nd (6 cm) pelvic recurrence excision and 12 m afterward the 3rd (2 cm) perineal scar reexcision.

All AA recurrences have the same histopathology with some mitotic figures and all resected organs were superficially invaded. Benign PNT tumors developed relapse (1/5) 34-month post-resection and recurrences were amenable to repeated curative salvage resections (Figure 6).

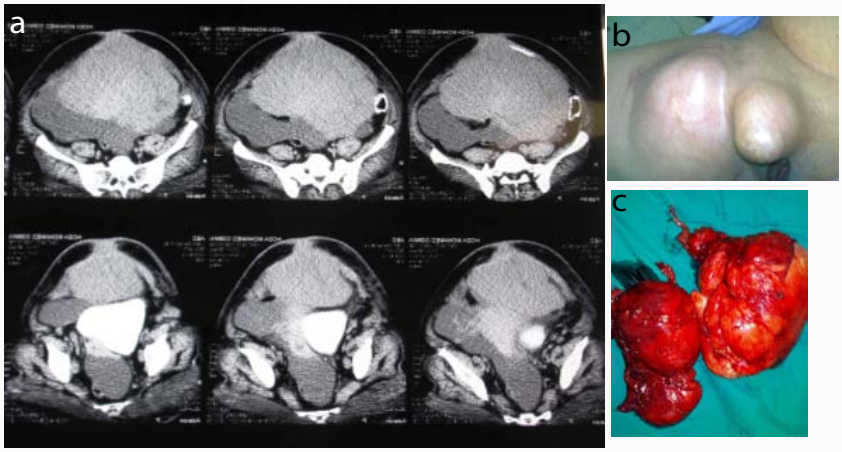

Figure 5: Patient No. 9 in Table 2. a, b) Pelvic undifferentiated sarcoma extending to the Rt. inguinal and femoral triangle area encasing iliofemoral vessels. c) Resected 2 components pelvic with cystectomy and inguinofemoral tumors and distal 5 cm of external iliac and common femoral artery segment with vein plus two cutaneous femoral branches and superficial part of floor muscles.

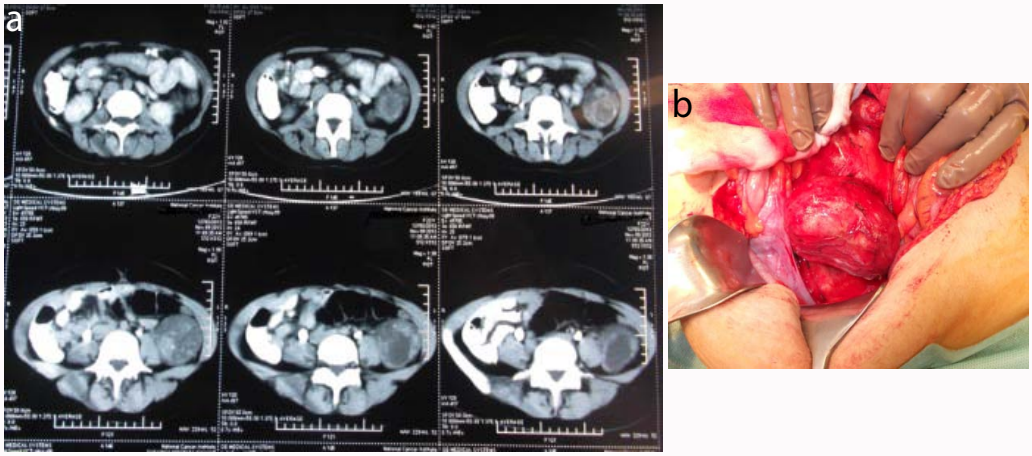

Figure 6: Patient 7 in Table 1. a) Surveillance CT lower abdomen of the lady 49 month after surgery shows the second recurrence of schwannoma in the left paracolic gutter. b) 3rd abdominal exploration to resect 9 cm recurrence.

Nine pelvic sarcomas developed 4 local pelvic recurrences (9–22-month post resection) and systemic spread (lung, Liver) in 3 patients. Lung metastatectomy was curative for one patient; local pelvic resection was also curative for another one with recurrent low grade liposarcoma. Salvage resection was not possible for two patients (SFT & leiomyosarcoma) with multiple pelvic relapses and synchronous systemic metastases.

Discussion

Pelvic anatomy is always disturbed with the large tumors dimensions and extensions rendering meticulous anatomical experience of the pelvis; critical to safely execute en bloc margin negative resection and maintain important viscera and vessels, also to differentiate the different tumors and to plan preoperative radiation treatment [2,3].

Since tumors grow deep seated gradually, it is difficult to detect them early and pathological examination revealed that the maximum diameter ranged up to 60 cm, mostly more than 20 cm. Tumors are commonly partially or non capsulated taking the shape of elongated, dumbbell or irregular large mass. Most tumors are lobular, heterogeneous in consistency with unclear invasive borders. They usually adhere to muscles, fat and nearby organs by firm dissectible fibrovascular tissues [19-21].

AA is locally aggressive because of local invasiveness plus recurrence and requires wide removal with firmly adherent organs, tissues and underlying perineal skin to get negative margins. Eighty five percent (85%) of relapses appear within 5 years and 2 m is the earliest reported recurrence time whereas the latest reported was 20 years. Pelvic desmoids (fibromatosis) is more invasive and highly recurrent. Benign PNST on the contrary; although reaches huge size, have more innocent behavior with low risk of recurrence [21-24].

Present series; although with limited cases gives description of possible surgical methods for these difficult and massive tumors with encouraging cure rate. Through preoperative assessment and meticulous operative technique with sufficient pelvic surgery experience were essential but, more problematic for the situation of recurrence in the previously operated pelvis. It was tough and critical to dissect nearly amalgamated fibrosed viscera to encompass the recurrent area which were sometimes multiple and intimately adherent to iliac arteries and veins. Pelvic recurrence salvage surgeries are lengthier, bloody and needs stoma cover frequently. Results for recurrence resection in local aggressive tumors were worth more than

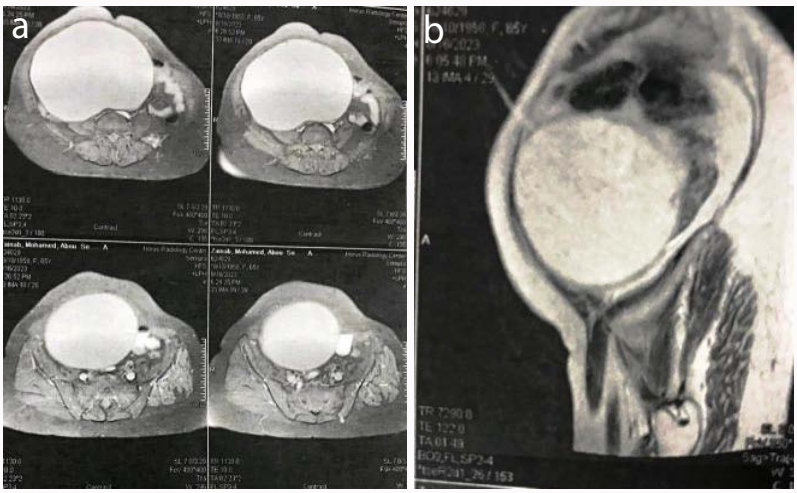

Figure 7: Patient 4 in Table 2. a, b) 61year patient with MPNST pelvic tumor and sciatic notch tumor extension.

for relapsed sarcoma [25]. Utmost local perineal relapses resections were straightforward except in the situations of anal, urethral and vaginal adherent tumor.

AA local recurrence was delayed but amenable for reexcision with cure but pelvic fibromatosis was aggressive with short disease-free period. Benign PNST (neurofibroma) were safe to resect without apparent recurrence over long observation time [26]. Even though schwannoma does not herniate extrapelvic but has a risk of local recurrence.

Local aggressive tumors have low mitotic activity excluding chemoradiation as an appropriate neoadjuvant treatment option. Certain studies suggested that AA tumors are hormone dependent because of +ve ER&PR in most patients and advised Gonadotropin Releasing Hormone agonist (GnRH-α), aromatase inhibitors or ant estrogen drugs as neoadjuvant shrinking treatment before surgery [27-29]. In a retrospective multicenter study on 13 advanced AA, first line GnRH-α treatments lead to overall response rate of 62% and progression-free survival of 24.6 m. Combining with aromatase inhibitors resulted in response in two non-responders and authors suggested the combined regimens with surgery [29].

Unluckily, hormone receptors were not available in our group of AA because of infrequency of cases in spite of use of other immunohistochemical markers for precise histologic diagnosis and no pre or post-operative hormonal or radiation treatment was given. Desmoid tumors had postoperative external pelvic radiotherapy; however, 4/6 developed ultimately killing multiple local pelvic recurrences. All 4 cases developed multiple peritoneal fibromatosis with multilevel intestinal obstructions and obstructive uropathy.

Limited patient's number makes difficulty to consider significant clinicopathologic prognostic factor that affects tumor recurrence risk. Involved surgical margins (R1) recurred and eventually died; however, other R0 patients recurred. Age and sex effects can't be considered because of different tumor histology and similarly extra tumor tissue infiltration.

3/4 large sarcomas had downsizing neoadjuvant external radiotherapy with partial regression in 2 and no regression for the third malignant SFT. Perioperative morbidity was similar to non radiation patients but, 2/4 had local pelvic relapse within first year.

Six-month radiological surveillance intensity for the first 5-year postoperative is adequate to detect recurrences that were more frequent in the first 24 month [30].

Both liposarcomas were well differentiated (R0) with no distant systemic relapse however one recurred in the pelvis. Recurrence histology showed dedifferentiated type; however, resection was curative.

Two-Leiomyosarcoma had no relation to gross iliac vessels and both recurred by distant spread with curative lung metastatectomy in only one. One MPNST (Figure 7) had preoperative downsizing 50 Gy radiation and R1 resection with neural and bladder invasion; however, luckily, he is free of recurrence for 44-month post surgery.

Conclusion

A consecutive series appraises clinical, pathologic and natural history of the sporadic pelvic soft tissue tumors. Surgery is the mainstay modality for these infrequent neoplasms with acceptable cure and morbidity rates, even for recurrent tumors. Management is dictated by histologic type and grade. Surgical resection with wide margins is the cornerstone of treatment for pelvic sarcomas, although this is often challenging due to anatomic constraints of the pelvis. Minimal invasive surgery in the face of huge complex pelvic and highly recurrent tumors are questionable and open resection with multiple approaches are valid with minimal morbidity.

Acknowledgment

Team of the Surgical Oncology and Surgical Pathology Departments, National Cancer Institute, Cairo University.

References

1. Mullen JT, VanHoudt W. Soft tissue tumors of the pelvis: Technical and histological considerations. J Surg Oncol. 2017;117(1):48-55.
2. Sambri A, Fiore M, Rottoli M, Bianchi G, Pignatti M, Bortoli M, et al. A planned multidisciplinary surgical approach to treat primary pelvic malignancies. Curr Oncol. 2023;30(1):1106-15.
3. Sarre-Lazcano C, Dumitra S, Fiore M. Pelvic soft tissue sarcomas. Eur J Surg Oncol. 2023;49(6):1102-10.
4. Fletcher C, Bridge J, Hogendoorn PCW, Mertens F. World Health Organization classification of tumors of soft tissue and bone. 4th Ed. IARC

Press, Lyon: 2013.

5. Gronchi A, Miceli R, Shurll E, Eilber FC, Eilber FR, Anaya DA, et al. Outcome prediction in primary resected retroperitoneal soft tissue sarcoma: Histology- specific overall survival and disease-free nomograms built on major sarcoma center data sets. J Clin Oncol. 2013;31(13):1649-55.
6. Mariani A, Nascimento AG, Webb MJ, Sim FH, Podratz KC. Surgical management of desmoid tumors of the female pelvis. J Am Coll Surg. 2000;191(2):175-83.
7. Chen H, Zhao H, Xie Y, Jin M. Clinicopathological features and differential diagnosis of aggressive angiomyxoma of the female pelvis. Medicine (Baltimore). 2017;96(20):e6820.
8. Li W, Chen J, Zhang, Chen W, Hu Y, Miao C, et al. Characteristics and outcomes of patients with primary abdominopelvic aggressive angiomyxoma: A retrospective review of 12 consecutive cases from a sarcoma referral center. BMC Surg. 2023;23(1):88.
9. Zou R, Xu H, Shi Y, Wang J, Wang S, Zhu L. Retrospective analysis of clinicopathological features and prognosis for aggressive angiomyxoma of 27 cases in a tertiary center: A 14-year survey and related literature review. Arch Gynecol Obstet. 2020;302(1):219-9.
10. Hsieh F, Chuang KT, Wu YT, Lin CH. Aggressive angiomyxoma-report of a rare male buttock lesion. Plast Reconstr Surg Glob Open. 2018;6(8):e1897.
11. Celik SU, Hesimov I, Kutlu B, Erkek AB. Aggressive angiomyxoma: A rare tumor of male pelvic cavity. Acta Med Port. 2018;31(11):693-6.
12. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: Diagnostic overview and update on selected diagnostic problems. Acta Neuropathol. 2012;123(3):295-319.
13. Kumar N, Goyal A, Manchanda S, Sharma R, Kumar A, Bansal VK. Aggressive pelvic angiomyxoma and its mimics: can imaging be the guiding light? Br J Radiol. 2020;93(1111):20200255.
14. Seoud M, Abbas J, Kaspar H, Khalil A, Geara F. Long-term survival following aggressive surgery and radiotherapy for pelvic fibromatosis. Int J Gynecol Cancer. 2005;15(6):1112-4.
15. Nassar OAH. Primary repair of rectovaginal fistulas complicating pelvic surgery by Gracilis myocutaneous flap. Gynecol Oncol. 2011;121(3):610-4.
16. Reyell MP, Grimer RJ. How to remove a dumbbell tumor of the sciatic notch. Sarcoma. 2000;4(1-2):61-2.
17. Spinner RJ, Endo T, Amrami KK, J Dozois E, Babovic-Vuksanovic D, Sim FH. Resection of benign sciatic notch dumbbell-shaped tumors. J Neurosurg. 2006;105(6):873-80.
18. Mitropoulos D, Artibani W, Biyani CS, Jensen BJ, Rouprêt M, Truss M. Validation of the Clavien-Dindo grading system in urology by the European Association of Urology Guidelines Ad Hoc panel. Eur Urol Focus. 2018;4(4):608-13.
19. Figueiredo G, O'shen A, Neville GM, Lee SI. Rare mesenchymal tumors of the pelvis: Imaging and pathologic correlation. Radiographics. 2022;42(1):143-58.
20. Messiou C, Moskovic E, Vanei D, Morosi C, Benchimol R, Strauss D, et al. Primary retroperitoneal soft tissue sarcoma: Imaging appearances, pitfalls and diagnostic algorithm. Eur J Surg Oncol. 2017;43(7):1191-8.
21. Choi H, Park C, Ji YI. Alternative surgical approaches for aggressive angiomyxoma at different sites in the pelvic cavity. Obstet Gynecol Sci. 2015;58(6):525-9.
22. Strauss DC, Qureshi YA, Hayes AJ, Thomas JM. Management of benign retroperitoneal schwannomas: A single center experience. Am J Surg; 2011;202(2):194-8.
23. Mehta GU, Huynh H, Lekovic GP. Peripheral nerve sheath tumors in Neurofibromatosis Type 2: Surgical and histopathologic features. Clin Neurol Neurosurg. 2020;190:105649.
24. Uerschels AK, Dengler NF, Chihi M, Lenkeit A, Dinger TF, Jabbarli R, et al. Benign peripheral nerve sheath tumors: An interdisciplinary diagnostic and therapeutic challenge. Neurosurg Rev. 2023;18;46(1):205.
25. Mor E, Assaf D, Shemia S, Ben-Ami E, Mor-Hadar D, Halfon M, et al. Resection of recurrent pelvic soft tissue sarcoma: Is the risk worth the reward? J Surg Res. 2023;283:914-22.
26. Nassar OAH. The infrequent large pelvi-perineal tumors as a surgical dilemma: en bloc resection and long-term results. Eur J Gynecol Oncol. 2023;44(4):22-7.
27. Fine BA, Munoz AK, Litz CE, Gershenson DM. Primary medical management of recurrent aggressive angiomyxoma of the vulva with gonadotropin-releasing hormone agonist. Gynecol Oncol. 2001;81(1):120-2.
28. Chan IM, Hon E, Ngai SW, Ng TY, Wong LC. Aggressive angiomyxoma in females: is radical resection the only option? Acta Obstet Gynecol Scand. 2000;79(3):216-20.
29. Fuca' G, Hindi N, Roy-Coquard I, Colia V, Tos APD, Martin-Broto J, et al. Treatment outcomes and sensitivity to hormone therapy of aggressive angiomyxoma: A multicenter, international, retrospective study. Oncologist. 2019;24(7):e536-41.
30. Glasbey JC, Bundred J, Tyler R, Hunt J, Tattersall H, Gourevitch D, et al. The impact of postoperative radiological surveillance intensity on disease free and overall survival from primary retroperitoneal, abdominal and pelvic soft-tissue sarcoma. Eur J Surg Oncol. 2021;47(7):1771-7.