

Experience with Surgical Treatment of Retroperitoneal Soft Tissue Sarcomas at a University Hospital in Thailand

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Background: Retroperitoneal soft tissue sarcomas (RSTS) are rare malignant tumors with a distinguishing feature of slow growth in the silent retroperitoneal space. The patients usually present late with a large retroperitoneal mass surrounded by the major vascular structures and visceral organs rendering curative resection an extremely difficult and risky operation. The purpose of the present study was to demonstrate surgical experience and results of treatment of RSTS at King Chulalongkorn Memorial Hospital. Operative techniques of these complex surgical procedures were also described.

Material and Method: A retrospective study was performed in patients who had RSTS and underwent surgical resection between June 2003 and November 2011 at King Chulalongkorn Memorial Hospital, Bangkok, Thailand. All patients were followed after the operations until death or last follow-up at the out-patient clinic in October 2012. Data collection included demographic data, details of operations, operative complications, neoadjuvant and adjuvant chemoradiation therapy, local recurrence, treatment of local recurrence, and 5-year overall survival rate. Factors associated with local recurrence were also examined.

Results: During the 9.4-year period, 18 patients entered into the present study. Fourteen (77.8%) were female and four (22.2%) were male. The age ranged from 44 to 80 years (median 53.5 years). Duration of symptoms ranged from one week to 24 months (median 3.5 months). The tumor size ranged from 10 to 48 cm (median 27 cm) in greatest dimension. All patients underwent preoperative CT scan. Preoperative core needle biopsy was performed in one patient. One patient had preoperative radiation therapy. Sixteen patients (88.9%) underwent complete gross resection (CGR) (R0 or R1 resection) and two (11.1%) had palliative resection (R2 resection). All patients who had CGR (n = 16) had one or more contiguous organ resection (kidney 87.5%, colon 50%, or adrenal gland 43.7%). The operative time ranged from 120 to 360 minutes (median 330 minutes). The operative blood transfusion ranged from 0 to 12 units (median 2.5 units). Postoperative bleeding complication requiring reoperation occurred in three patients (16.7%). One patient had postoperative uncomplicated pancreatic fistula. There was no perioperative mortality. The final pathological reports were liposarcoma in 15 patients (83.3%). Other histology were atypical lipomatous tumor, malignant fibrous histiocytoma, and unspecified spindle cell tumor in one patient each. Two patients who had palliative resection died at six and 16 months after the operations. Local recurrence occurred in five patients who had CGR (31.3%). One of them died at 60 months after the operation. The median follow-up time in patients who underwent CGR was 39.5 months (range 12-114 months). The 5-year overall survival of the entire cohort was 73.5% (95% CI: 44.3-88.4%). The 5-year overall survival of patients who had CGR was 83.3% (95% CI: 53.5-98.5%). Univariate analysis of the tumor size, tumor grading, status of the surgical margins, and primary operation or re-resection revealed no statistical significance in patients who had CGR with and without local recurrence.

Conclusion: Acceptable outcomes after complete surgical resection of the RSTS were achieved from this small but important case-series. The authors have demonstrated that CGR with concomitant resection of the contiguous organs can be safely performed in patients with large RSTS. Preoperative CT scan was invaluable for diagnosis and treatment plan. Preoperative core needle biopsy was not necessary when preoperative CT scan was diagnostic. Intention for curative resection should be attempted whenever possible to minimize chance of local recurrence and improve survival. Experience of the surgical team is an important factor for successful results when conducting these technically demanding operations.

Keywords: Retroperitoneal soft tissue sarcomas, Complete gross resection, Contiguous organ resection, Liposarcoma

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Retroperitoneal soft tissue sarcomas (RSTS) are rare malignant tumors arising from the retroperitoneal adipose and connective tissue of the mesenchymal origin. It has been estimated that soft

tissue sarcomas are found in less than 1% of all malignancies and approximately 15% of these cases are RSTS^(1,2). The annual incidence of RSTS has been reported to be 2.7 cases per one million population⁽³⁾. Such uncommon occurrence may explain the difficulty to achieve the optimal results of treatment in those hospitals or institutions not dedicated for this disease. The importance of clinical and surgical experience in dealing with these aggressive tumors cannot be

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overemphasized. Some investigators have demonstrated that soft tissue sarcoma patients treated at high-volume centers have significantly better outcomes than those treated at low-volume centers and patients with large tumors (>10 cm in maximum diameter) should be treated only in a high-volume center⁽⁴⁾. Comparing with other malignancies, publications concerning RSTS are scant and most of them were reported from large cancer centers in North America and Europe⁽⁵⁻¹⁰⁾. Literature regarding surgical treatment of RSTS from the Asian countries is still relatively lacking⁽¹¹⁻¹³⁾. In Thailand, the authors are scarcely able to obtain adequate information specific to RSTS from the current available English language database while knowledge of other more common diseases is increasingly disclosed. One case report of gigantiform retroperitoneal liposarcoma was published in Journal of the Medical Association of Thailand 30 years ago⁽¹⁴⁾. Until now, the publications regarding RSTS in Thailand have been very limited.

It is well-known to surgeons practicing in this field of surgery that prognosis of RSTS is formidable with the dismal natural history of local recurrence after resection, on-going enlargement of recurrent or unresectable tumors, distant metastasis, and eventually death. One important factor that may be responsible to poor outcomes is the delay in diagnosis resulting in common presentation of patients with large tumor size. Large tumor size has been reported to be associated with higher rate of incomplete resection and hence, higher rate of local recurrence compared to smaller one^(15,16). Unfortunately, the retroperitoneal location of these rare tumors with silent slow growing and quiet expansion in nature render the patients asymptomatic until the tumors become enormous in size and complete resection may be compromised by nearby vascular structures such as aorta or inferior vena cava (IVC) or may require resection of contiguous organs such as kidney and adrenal gland, colon, spleen, etc. to achieve adequate oncologic margins^(6,7,9,10,17-19). However, even what seems to be a satisfactory operation by successful removal of all bulk of tumor or complete gross resection (CGR) with or without resection of the contiguous organs, chance of local recurrence still remains regardless the microscopic status of the surgical margins^(5,17). This unpredictable tumor behavior explains the difficulty in management of RSTS and encourages further studies and researches for more understanding and better outcomes of treatment.

Surgery on patients with RSTS is technically demanding especially in huge or giant tumors^(4,14,16). However, only a few publications described practical viewpoints of surgical techniques. Furthermore, there have been some controversies in the literature regarding safe surgical approaches starting from the incision to details of operation⁽²⁰⁻²²⁾. Appropriate surgical techniques are extremely important during resection of these tumors in a limited surgical field, especially when the tumors are very large. Failure to proceed cautiously with proper manner of dissections and along the correct surgical planes may result in exsanguinating hemorrhage and/or incomplete resection of the tumors. The ultimate goal of surgery is CGR with microscopically negative surgical margins, which is essential for the best outcomes^(5-7,9,10,16).

The purpose of the present study was to examine the authors' surgical experience in patients with RSTS at a University Hospital in Bangkok, Thailand. During such period of study, CGR with or without concomitant resection of the contiguous organs whenever possible was our policy of management of RSTS. Details of patients, operative procedures, postoperative complications, tumor characteristics, and pathology of the tumors were summarized. The surgical techniques are presented and discussed. Results of treatment, local recurrence, factors associated with local recurrence, and survival analyses were studied. Comparisons of the outcomes to other previous reports were also selectively made when appropriate.

Material and Method

All patients who had RSTS and underwent surgical resection by the first author between June 2003 and November 2011 at King Chulalongkorn Memorial Hospital, Bangkok, Thailand were examined and analyzed. The patients were followed postoperatively until death or last follow-up at the out-patient clinic in October 2012. The study was approved by the Institutional Review Board of the Faculty of Medicine, Chulalongkorn University. Patients who presented with retroperitoneal mass with the final diagnosis of other malignancies apart from RSTS were excluded from the study. These exclusions were lymphomas, sarcoma of the pancreas, sarcomas arising from the gastrointestinal tract, and genito-urinary malignancies. Almost all patients presented to us with a large abdominal mass with varying period of vague abdominal discomfort and/or pain. Some patients had had previous surgical resection of the RSTS elsewhere

and were transferred to King Chulalongkorn Memorial Hospital because of local recurrence. One patient already had tissue diagnosis of RSTS from core needle biopsy before transferring to us. When the patient first came to us with an abdominal mass and/or symptoms of vague abdominal discomfort and/or pain, CT scan of the abdomen was routinely performed and the preliminary diagnosis of RSTS was made by the radiologists according to the specific radiologic criteria (Fig. 1-3). After that, surgical resection was planned without any attempt to obtain preoperative diagnosis by fine needle aspiration or core needle biopsy. Before September 2011, all patients underwent surgical resection without preoperative radiotherapy. After September 2011, one patient who was the last patient

in our case-series had preoperative radiotherapy at the tumor bed (mostly posterior abdominal wall muscles) for 40 Gy followed by surgical resection five weeks later. Since complete surgical resection of the tumor usually included removal of ipsilateral kidney, preoperative assessment of the contralateral kidney was also necessary. Normal serum BUN and creatinine levels and normal radiologic appearance of contralateral kidney and ureter on preoperative CT scan indicated safe postoperative renal functions after removal of the tumor and kidney of the affected side. The patients were also informed preoperatively about the possibility of simultaneous removal of the kidney and/or other visceral organs. Preoperative mechanical bowel preparation was also routinely performed owing to a

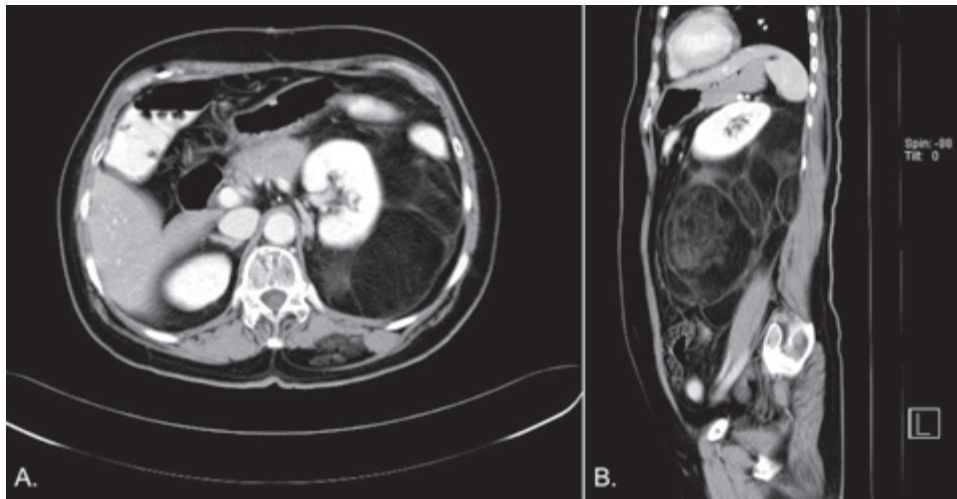


Fig. 1 CT scan of a 63-year-old female patient with left RSTS. The patient underwent en-bloc resection of RSTS, left nephrectomy, left colectomy, and left salpingo-oophorectomy. The final pathological report was well differentiated liposarcoma. A. Horizontal view, B. Sagittal view.

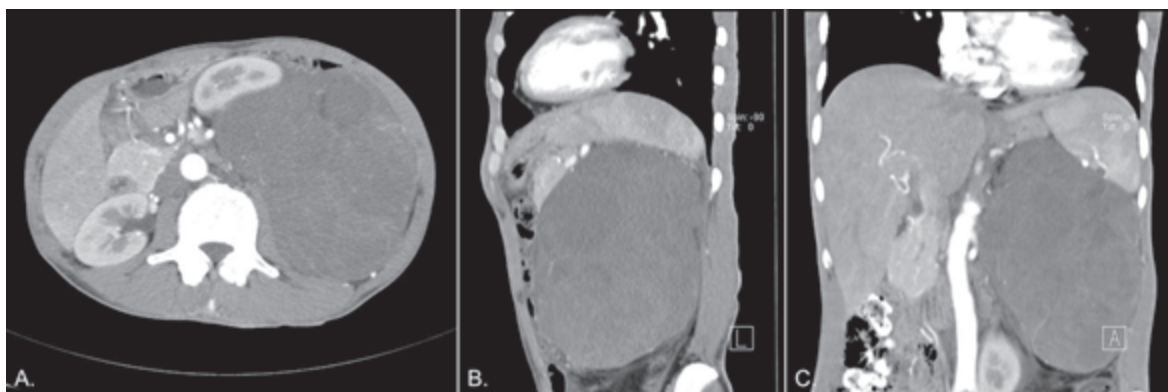


Fig. 2 CT scan of a 54-year-old male patient with left RSTS. The patient underwent en-bloc resection of RSTS, left nephrectomy, and left adrenalectomy. The final pathological report was liposarcoma, mixed type, grade 3. A. Horizontal view, B. Sagittal view, C. Coronal view.

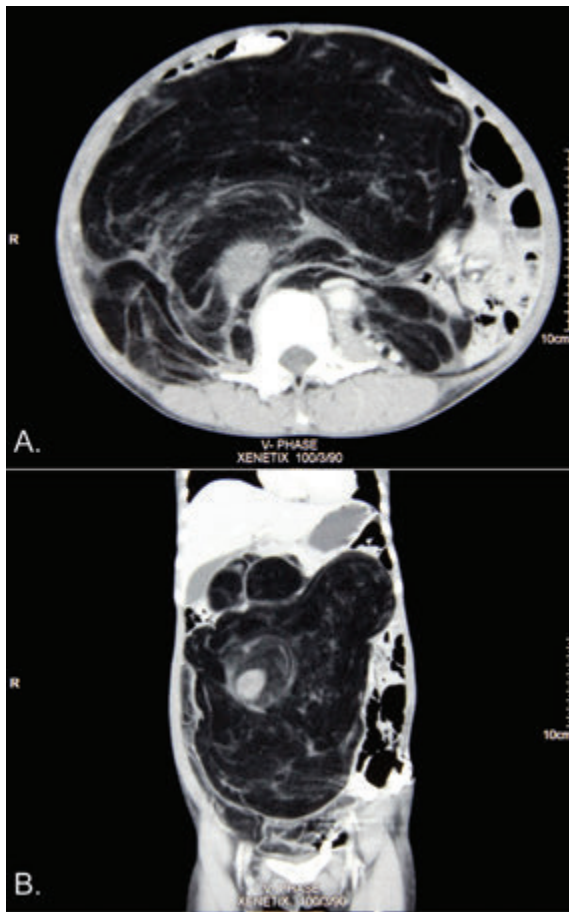


Fig. 3 CT scan of a 51-year-old female patient showing a large recurrent liposarcoma. The patient had undergone previous excision of the right RSTS 6 years before coming to us with a large recurrence. The final pathological report after resection of this recurrence was dedifferentiated liposarcoma. A. Horizontal view, B. Coronal view.

relatively high possibility of resection of the right or left colon in patients with large tumors.

During the operation, CGR of the tumor was the primary goal. Palliative resection was performed in only two patients; both of them had multiple sites of tumors, one of them also had multiple recurrence and encasement of the superior mesenteric artery. CGR was defined as total removal of the RSTS without entering its surrounding capsule. To accomplish this purpose, the contiguous organs were also removed en-bloc, i.e. left or right kidney and adrenal gland, left or right colon, spleen, distal pancreas, and diaphragm, etc. Most of the time, the kidney and adrenal gland were removed en-bloc with the RSTS (Fig. 4, 5). Since the tumors were frequently quite large in size at

the time of surgery, CGS was usually difficult and sometimes extremely dangerous from exsanguinating hemorrhage of inadvertent injury to the IVC or aorta. The authors would like to present our surgical approaches for removal of RSTS that had been proved to be safe in our patients. Such approaches were successfully performed in large RSTS of our case-series with the largest tumor measuring 48 cm in greatest dimension and weighing 25 Kg without perioperative mortality (Fig. 6).

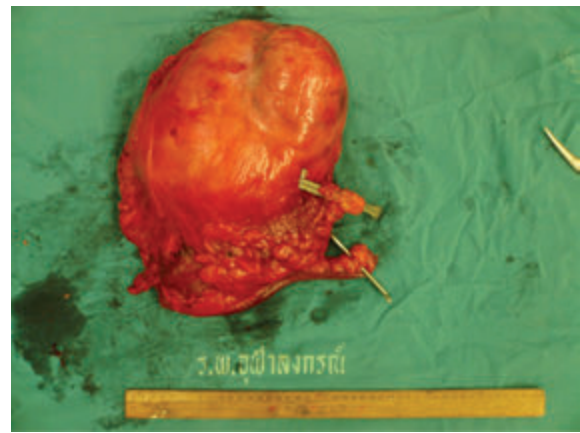


Fig. 4 Photograph showing surgical specimen after CGR with contiguous organ resection. The resected mass included the tumor, the right kidney, and the right colon. The final pathological report was dedifferentiated liposarcoma, grade 3.



Fig. 5 Photograph of the surgical specimen showing surgical specimen after CGR with contiguous organ resection. The resected mass included the tumor, the left kidney and adrenal gland, the distal pancreas, the spleen, and the left colon. The final pathological report was well differentiated liposarcoma.

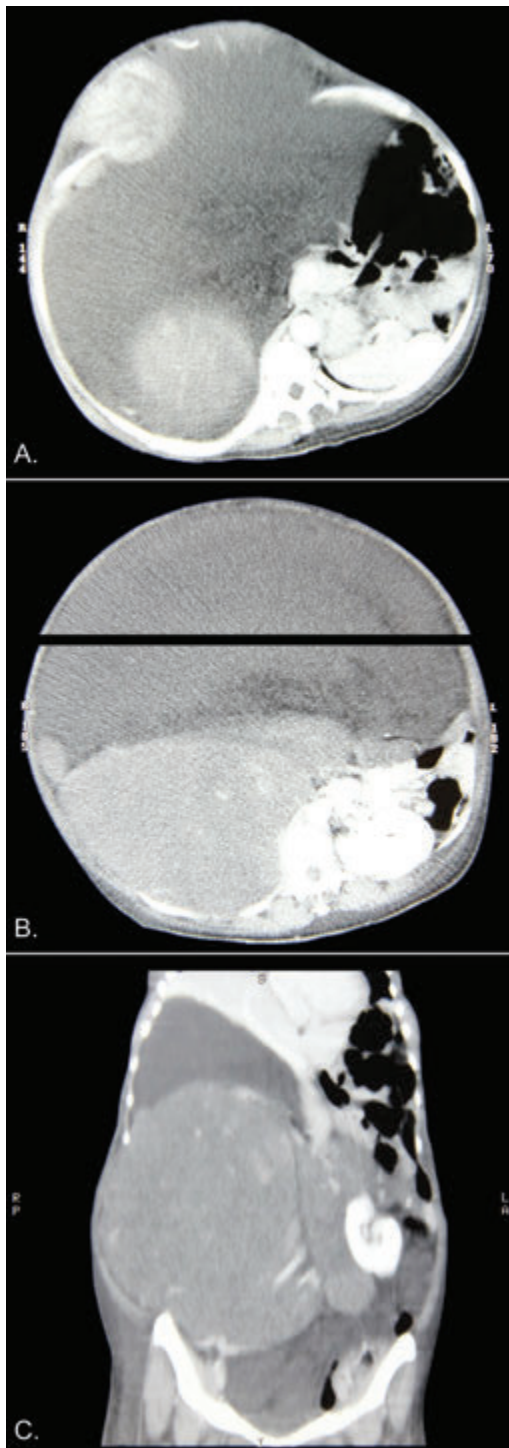


Fig. 6 CT scan of a 47-year-old female patient showing a very large right RSTS. The tumor was the largest RSTS in our case-series, measuring 48x45x17 cm and weighing 25 Kg. The final pathological report was dedifferentiated liposarcoma, grade 3. A. and B. Horizontal views, C. Coronal view.

Surgical techniques

The authors' preferred surgical approach for tumor resection is a long midline incision. The length of incision depends on the tumor size. In very large tumors, we usually use an incision from xiphoid cartilage to pubic symphysis. No thoracic extension of incision was necessary in our patients. Upon entering the abdominal cavity, the necessity for resection of the right colon (for tumor in the right retroperitoneal area) or the left colon (for tumor in the left retroperitoneal area) is determined. This decision is dependent largely on the tumor size. A very large tumor is associated with the need for right or left half colectomy since mobilization of the right or left colon from the tumor in such a situation is not possible or may take the risk of entering into the tumor capsule (Fig. 7, 8). When the tumor is in moderate size and the surgeon thinks that mobilization of the right or left colon from the tumor behind is safe, the lateral peritoneal attachment of the right or left colon to the lateral abdominal wall is incised and the right or left colon is mobilized with sharp and blunt dissection medially until the right or left colon and its mesocolon is free from the medial aspect of the tumor. Dissections are then made from the infero-medial aspect of the tumor upwards. The tumor is lifted up from the posterior and midline of the abdomen showing the plane between postero-medial aspect of the tumor and the anterior wall of the iliac vessels, IVC, and aorta. During this step, care should be taken not to lift the tumor too strong since it may cause hemorrhage from avulsion of the small branches of vessels from the midline IVC and aorta.

When the avascular plane between the postero-medial aspect of the tumor and the anterior wall of the IVC and aorta cannot be safely developed owing to large tumor size causing a very limited space for dissections, the tumor should be partially mobilized from the lateral and posterior abdominal wall muscles just to allow more space for safe dissections of the tumor from the midline IVC and aorta. Separation of the tumor from the lateral and posterior abdominal wall muscles should not proceed too much without complete clearance of the tumor from the midline IVC and aorta since uncontrollable hemorrhage may occur in every step as long as the midline IVC and aorta are not free from the tumor. Divisions of the ureter and gonadal vessels also enhance the mobility of the tumor and should be performed early at this step when the ipsilateral kidney is planned to be removed (87.5% of our patients who had CGR) (Fig. 9). When the avascular plane is

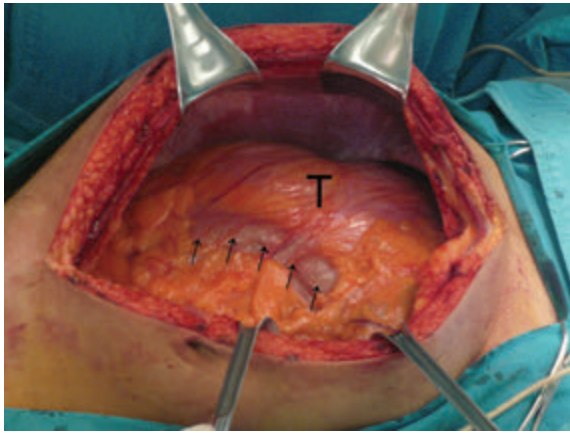


Fig. 7 Operative photograph of a patient with left RSTS showing appearance of the tumor (T) behind the left colon (arrows) when the abdomen was first entered.

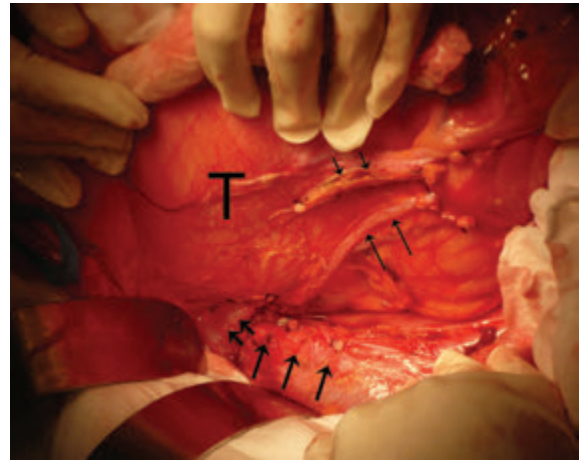


Fig. 9 Operative photograph showing surgical techniques by lifting the tumor (T) out of the posterior abdominal wall after ligation and division of the left ureter (long, thin arrows) and the left ovarian vein (short, thin arrows). The dissection was performed along the plane of the tumor (T) and the abdominal aorta (long, thick arrows) from the aortic bifurcation cephaladly until the left renal vein (short, thick arrows) was identified.

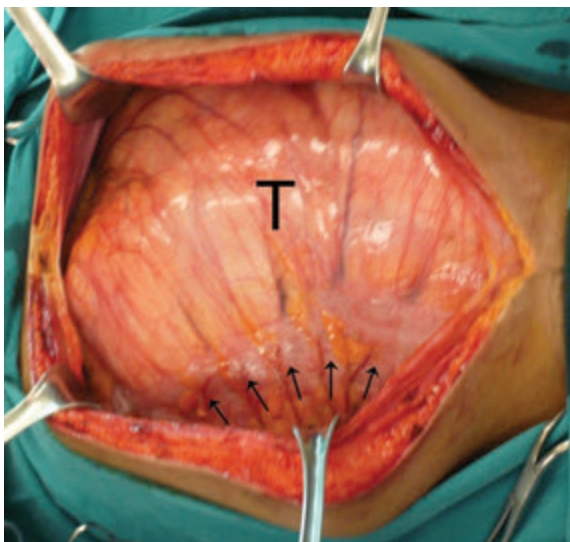


Fig. 8 Operative photograph of a patient with right RSTS showing appearance of the tumor (T) behind the right colon (arrows) when the abdomen was first entered.

developed from lifting the tumor upwards, dissections continue along this plane separating anterior wall of the IVC and aorta from the postero-medial aspect of the tumor until reaching the suprarenal IVC and aorta. The renal artery and vein of the affected side are then identified, isolated, double ligated, and divided. In our patients, we have observed that the renal vessels are the major connections of the tumor to the midline IVC and aorta. After division of renal vessels, further dissections upwards along the suprarenal IVC and

aorta (depending on the size of the tumor) can be performed without difficulty. At this step, the postero-medial aspect of the tumor is totally separated from the midline vascular structures and ready to be extensively mobilized from the posterior and lateral abdominal wall muscles. The tumor is lifted from the posterior and lateral abdominal wall muscles and the fascia connecting the tumor to the lateral and posterior abdominal wall muscles are cut with a pair of long scissors. Dissections move towards the diaphragm until the tumor is totally separated from the surrounding tissue without entering the tumor capsule. If the tumor is densely adherent to the diaphragm and the liver on the right or to the diaphragm, the pancreas, and the spleen on the left; all these structures should be resected en-bloc with the tumor too.

When removal of the right or left colon is considered necessary in patients with large tumor, the right or left colon is not mobilized away from the tumor as previously described for situation when the right or left colon is spared from resection. Instead, the transverse colon is dissected from the greater omentum and point of transection at the transverse colon is selected. The transverse colon is then transected between two clamps. For removal of the right colon, the transverse and right mesocolon is then divided to the mesentery of the terminal ileum, followed by

transection of the terminal ileum between two clamps. For removal of the left colon, the mesocolon of the transverse, left and sigmoid colon is divided with subsequent transection of the sigmoid colon between two clamps. At this stage, the right or left colon is ready to be removed en-bloc with the tumor and dissections commence from the postero-medial aspect of the tumor by lifting the tumor and the right or left colon away from the midline IVC and aorta as described earlier.

After the tumor with contiguous organs has been removed, complete hemostasis is of the utmost importance since the large raw surface area of the lateral and posterior abdominal wall muscles can cause massive postoperative hemorrhage (Fig. 10, 11). All the bleeding points from the muscles and lumbar arteries should be suture ligated with No. 2-0 silk. Other oozing of blood from the raw surface may be stopped with electrocoagulation. If the right or left colon has been removed, the intestinal anastomosis is subsequently performed. Two vacuum drains are placed at the raw surface and the abdomen is closed in usual manner. The perioperative mortality in the present study was defined as death within 1 month after the operation.

The surgical specimen was sent for the pathological examination for definite pathological diagnosis. It includes tumor size, tumor histology, grading of the tumor, and microscopic margins. After obtaining the final pathological reports, the radiotherapists and oncologists were consulted for appropriate adjuvant therapy. All patients were followed and searched for local recurrence and distant metastasis by interval CT scan of the chest and abdomen (every 3 to 6 months during the first 2 years and once a year thereafter). If local recurrence was detected, resection was considered whenever possible. Follow-up was done until the patient died or last seen in October 2012 at the out-patient clinic. The last patient in our case-series was followed-up to 12 months after surgical resection.

Univariate analysis was performed to evaluate factors associated with local recurrence by using Unpaired t-test and Fisher's exact test. A *p*-value of <0.05 was considered statistically significant. Survival analyses were performed by using Kaplan-Meier method.

Results

During the 9.4-year period, 18 patients entered into the present study. Fourteen (77.8%) were female

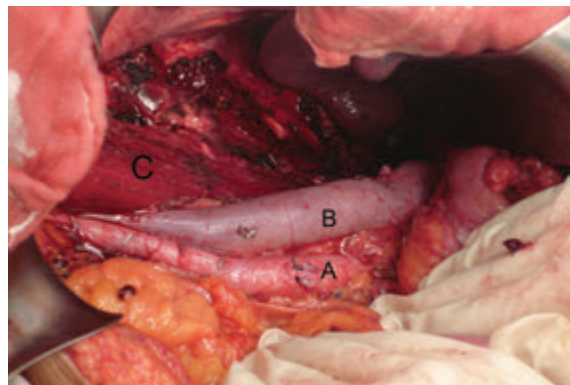


Fig. 10 Operative photograph showing appearance of the posterior abdominal wall after CGR with contiguous organ resection of the right RSTS. A = abdominal aorta, B = inferior vena cava, C = psoas muscle.

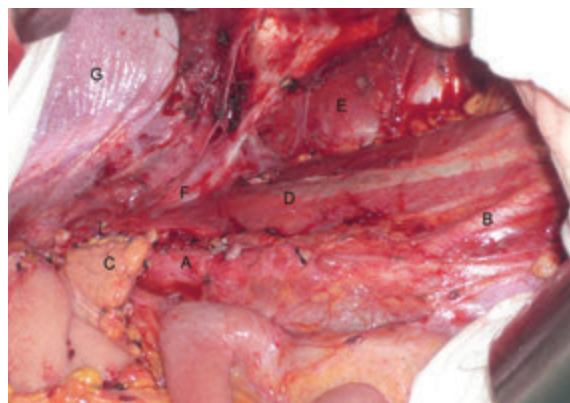


Fig. 11 Operative photograph showing appearance of the posterior abdominal wall after CGR with contiguous organ resection of the left RSTS. A = abdominal aorta, B = left common iliac artery, C = pancreatic stump, D = psoas muscle, E = transversus abdominis muscle, F = quadratus lumborum muscle, G = diaphragm.

and 4 (22.2%) were male. The age ranged from 44 to 80 years (median 53.5 years). Duration of symptoms of abdominal distension and/or discomfort and/or pain ranged from one week to 24 months (median 3.5 months). The tumor size ranged from 10 to 48 cm (median 27 cm) in greatest dimension measuring from the surgical specimens. Data about the tumor weight was available in nine patients ranging from 1,159 gm to 25 Kg (median 5,000 gm) (Table 1). All patients had preoperative CT scan of the abdomen that had led to preoperative diagnosis and decision for surgical resection. Seventeen patients (94.4%) underwent surgical resection without preoperative tissue

Table 1. Details of patients

Patient No.	Age	Gender	Tumor size (cm)	Tumor weight (gm)	Duration of symptoms (month)	Previous operation for RSTS	Pathological diagnosis
1	47	Female	15x14x9	1,159	2	No	Mixoid LS (grade 1-2)
2	54	Female	35x25x9	NA	2	No	Atypical lipomatous tumor
3	48	Female	30x23.5x15	6,999	4	No	LS, mixed type, grade 3
4	73	Female	21x11x5.5	NA	NA	No	LS, low grade dedifferentiation
5	63	Female	19x16x11	NA	NA	Yes (12 months)	LS, well differentiated
6	80	Male	10x8x7	NA	20	No	Unspecified spindle cell tumor
7	47	Female	48x45x17	25,000	10	No	LS, dedifferentiated, grade 3
8	68	Female	34x24x9	3,785	3	No	LS, mixed type, grade 2
9	49	Female	15x13x10	NA	1 week	No	MFH grade 3
10	68	Male	37x31x15	8,000	6	No	LS, mixed type, grade 3
11	53	Female	33x25x14	8,000	NA	Yes (76 months)	LS, dedifferentiated
12	48	Female	24x15x11	NA	NA	Yes (4 times)	LS, mixoid/round cell, grade 3
13	52	Male	25x24x11	5,000	3	No	LS, mixed type, grade 3
14	44	Female	29x20x16	NA	24	Yes (5 times)	LS, mixed type (mixoid/round cell), grade 3
15	54	Female	30x22x12	NA	2	No	LS, pleomorphic, grade 3
16	54	Male	19.2x16.9x14	NA	12	No	LS, mixed type, grade 3
17	55	Male	17x15x10	4,000	12	No	LS, dedifferentiated, grade 3
18	46	Female	31.8x24.3x7.5	4,400	3	No	LS, well differentiated

RSTS = retroperitoneal soft tissue sarcomas; NA = not available; LS = liposarcoma; MFH = malignant fibrous histiocytoma

diagnosis. One patient (5.6%) had had previous core needle biopsy with the diagnosis of spindle cell tumor before transferring to King Chulalongkorn Memorial Hospital and the final pathological diagnosis after tumor resection was malignant fibrous histiocytoma (MFH). Four patients (22.2%) had had previous surgical resection and were transferred to us because of recurrence. Among these patients with recurrence, two had one previous surgical resection and the remaining two had four and five previous surgical resections. One patient (11.1%) had preoperative radiotherapy (40 Gy) before undergoing surgical resection five weeks later. Sixteen patients (88.9%) had CGR (R0 or R1 resection), of these, nine (56.3%) had R0 (microscopically negative margins) and seven (43.7%) had R1 (microscopically positive margins). CGR was performed in three patients with recurrent tumors from previous operations elsewhere, two had one previous resection and one had five previous resections. Of the two patients who had incomplete gross or palliative resection (R2 resection) at King Chulalongkorn Memorial Hospital, both had multiple

sites tumors, one of them had multiple previous surgical resections (4 times).

All of the patients who had CGR (n = 16) had one or more contiguous organ resection. Among the adjacent organs removed in CGR, kidney was the most common (14/16 or 87.5%), followed by right or left colon (8/16 or 50%), adrenal gland (7/16 or 43.7%), psoas muscle (3/16 or 18.8%), uterus and both ovaries, ovary only, spleen (2/16 each or 12.5% each), distal pancreas, and diaphragm (1/16 each or 6.3% each). Microscopic involvement of the resected contiguous organs were found in eight out of 16 patients or 50%. Eleven out of 16 patients (68.8%) had high-grade RSTS (histologic grade 2 and 3). The operative time ranged from 120 to 360 minutes (median 330 minutes). The operative blood transfusion ranged from 0 to 12 units (median 2.5 units) (Table 2).

Postoperative bleeding complication occurred in three patients (16.7%). All of them underwent successful reoperation with hemorrhagic control by suturing the bleeding points from the

Table 2. Details of operations

Patient No.	Type of resection	Microscopic resection margins	Contiguous organ resection (only in CGR)	Pathological involvement of the resected contiguous organs	Operative time (minute)	Operative blood transfusion (unit)
1	CGR	R1	Kidney	No	150	0
2	CGR	R0	Kidney	No	240	1
3	CGR	R1	Kidney, adrenal gland, liver, uterus and both ovaries	Adrenal gland, liver	540	12
4	Palliative	R2	-	-	480	8
5	CGR	R0	Kidney, adrenal gland, ovary, left colon	Colon	400	4
6	CGR	R0	Psoas muscle	No	120	0
7	CGR	R1	Kidney	No	240	5
8	CGR	R1	Kidney, adrenal gland, spleen	Perinephric fat, periadrenal and perisplenic tissue	360	1
9	CGR	R0	Left colon	No	300	0
10	CGR	R1	Kidney, adrenal gland, left colon	Perinephric fat, periadrenal fat, serosa and pericolonic fat	420	4
11	CGR	R1	Kidney, right colon, psoas muscle	Kidney capsule, psoas muscle	480	7
12	Palliative	R2	-	-	270	3
13	CGR	R0	Kidney, right colon, diaphragm	Perinephric fat, pericolonic fat	540	3
14*	CGR	R1	Kidney, right colon, ovary, psoas muscle	Perinephric fat, pericolonic fat, psoas muscle	540	5
15	CGR	R0	Kidney	Perinephric fat	300	0
16	CGR	R0	Kidney, adrenal gland	No	270	1
17	CGR	R0	Kidney, adrenal gland, right colon	No	400	1
18	CGR	R0	Kidney, adrenal gland, spleen distal pancreas, left colon, uterus and both ovary	No	270	2

CGR = complete gross resection, R0 = free resected margins, R1 = microscopically positive resected margins, R2 = incomplete gross resection

* Concomitant removal of 5.8x4.0x4.0 cm liposarcoma at left buttock

lumbar arteries and psoas muscle with silk No. 2-0. One patient who had preoperative radiation therapy followed by CGR with distal pancreatectomy developed uncomplicated pancreatic fistula, which was spontaneously closed by conservative management. There was no perioperative mortality in the present study.

Liposarcoma was the most common histology of RSTS in the present study (15/18 or 83.3%). The histology of the remaining three patients was atypical

lipomatous tumor, MFH, and unspecified spindle cell tumor. Eleven out of 15 patients (73.3%) who had liposarcoma had grade 3 differentiation.

Eight patients (44.4%) had postoperative radiotherapy only. One patient (5.6%) had postoperative chemotherapy only. Three patients (16.7%) had postoperative radiation and chemotherapy. One patient (5.6%) had preoperative radiation therapy only (Fig. 12). Five patients (27.8%) had no adjuvant or neoadjuvant radiation or chemotherapy (Table 3).

Table 3. Neoadjuvant, adjuvant therapy and results in patients with CGR (R0 or R1) (n = 16)

Patient No.	Microscopic involvement of the resected margins	Preoperative radiotherapy	Postoperative radiotherapy	Postoperative chemotherapy	Postoperative chemotherapy	Recurrence (months after CGR)	Treatment of recurrence	Outcomes at last follow-up (October 2012)	Follow-up time (month)
1	R1	-	50 Gy	-	-	No	-	Alive	114
2	R0	-	-	-	-	No	-	Alive	102
3	R1	-	50 Gy	Yes	Chemotherapy	Yes (24)	-	Dead (June 2010)	60
4	R2	-	-	-	-	-	-	-	-
5	R0	-	50 Gy	-	-	No	-	Alive	66
6	R0	-	-	-	-	No	-	Alive	61
7	R1	-	-	-	-	No	-	Alive	61
8	R1	-	-	-	Resection+XRT (54 Gy)	Yes (27)	-	Alive with recurrence (no further resection)	46
9	R0	-	50 Gy	-	Resection + chemotherapy	Yes (26)	-	Recurrence at 40 months with successful re-resection	46
10	R1	-	57.2 Gy	-	-	No	-	Alive	33
11	R1	-	-	Yes	Resection (only)	Yes (15)	-	Alive	32
12	R2	-	-	-	-	-	-	-	-
13	R0	-	50 Gy	Yes	-	No	-	Alive	25
14	R1	-	50.4 Gy	-	Resection	Yes (22)	-	Alive	24
15	R0	-	54 Gy	-	-	No	-	Alive	24
16	R0	-	50 Gy	-	-	No	-	Alive	17
17	R0	-	-	-	-	No	-	Alive	14
18	R0	40 Gy	-	-	-	No	-	Alive	12

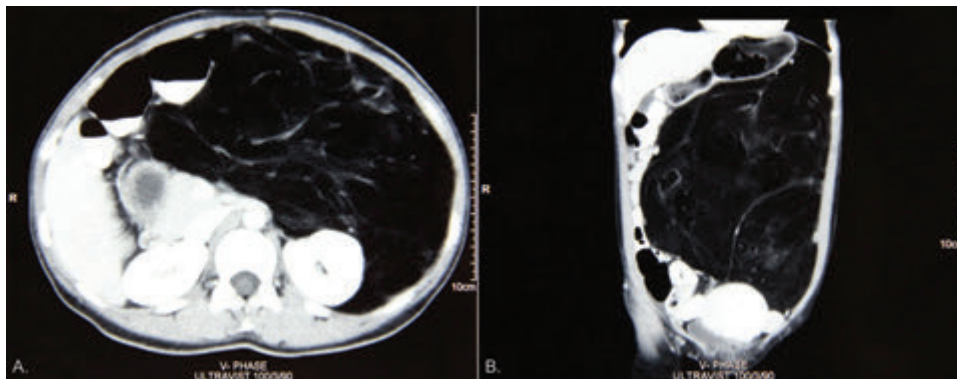


Fig. 12 CT scan of a 46-year-old female patient showing a left RSTS. This is the last and the only patient in our case-series who had preoperative radiation therapy before surgical resection. The final pathological report was well differentiated liposarcoma. A. Horizontal view, B. Coronal view.

During postoperative follow-up, both patients who had incomplete gross (R2) resection had died six and 16 months after the operations from progression of disease. Of the 16 patients who had CGS (R0 or R1 resection), five (31.3%) had local recurrence at 15, 22, 24, 26, and 27 months after CGR at King Chulalongkorn Memorial Hospital. Four recurrences occurred in patients who had R1 resection ($n = 7$) (57.1%) and only one (11.17) occurred in patients who had R0 resection ($n = 9$) ($p = 0.101$) (Table 4). Of these recurrences, one who had recurrence at 24 months postoperatively, had progression of disease, and died at 60 months postoperatively without re-resection. The remaining four patients had re-resection of the recurrences; two still well with no evidence of recurrence, and two had another recurrence (one had successful re-resection and one refused further surgical resection). All four patients were still alive with normal lifestyle at the time of preparing this manuscript. The remaining 11 patients who had CGR with no recurrence were all alive and well.

In patients who had CGR, the recurrence rate was 31.3% (5 out of 16) with median follow-up time of 39.5 months (range 12-114 months). Univariate analysis of the tumor size, tumor grading, microscopic margins of the resected tumors, and primary operation vs. re-resection to evaluate factors associated with local recurrence failed to demonstrate statistical significance in patients who had CGR with and without local recurrence (Table 4). Survival analysis by Kaplan-Meier method revealed a 5-year overall survival of 73.5% (95% CI: 44.3-88.4%) of the entire cohort (Fig. 13). When analysis was performed only in patients who had CGR, the 5-year overall survival was 83.3% (95% CI: 53.5-98.5%) (Fig. 14). The median survival was not reached.

Discussion

Patients with RSTS are usually presented late with a considerable size of abdominal mass. Although it is difficult to pinpoint the exact onset of symptoms, most patients can roughly estimate the duration of

Table 4. Univariate analysis of factors associated with local recurrence in patients undergoing CGR ($n = 16$)

Variable	Recurrence ($n = 5$)	No recurrence ($n = 11$)	<i>p</i> -value
Tumor size (cm) (mean \pm SD)	28.2 \pm 7.66	26.1 \pm 11.36	0.7135
Tumor grading			
Low grade (grade 1)	0 (0%)	6 (54.5%)	0.093
High grade (grade 2, 3)	5 (100%)	5 (45.5%)	
Resection margins			
R0	1 (20%)	8 (72.7%)	0.101
R1	4 (80%)	3 (27.3%)	
Primary surgery vs. re-resection			
Primary surgery	3 (60%)	10 (90.9%)	0.214
Re-resection	2 (40%)	1 (9.09%)	

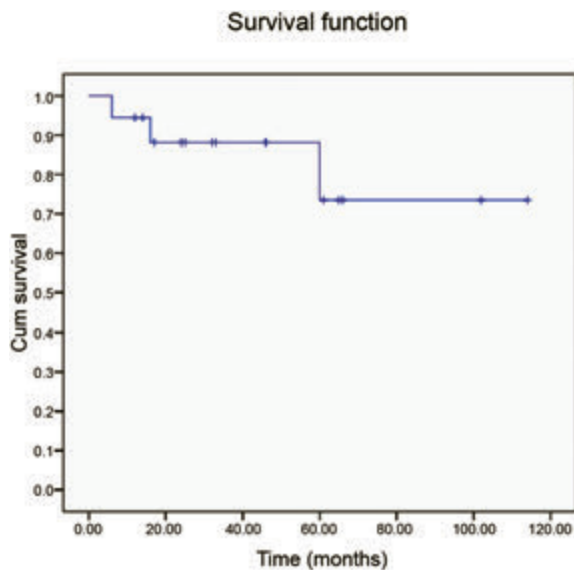


Fig. 13 Survival analysis of the entire cohort (n = 18) by Kaplan-Meier method. The 5-year overall survival was 73.5%.

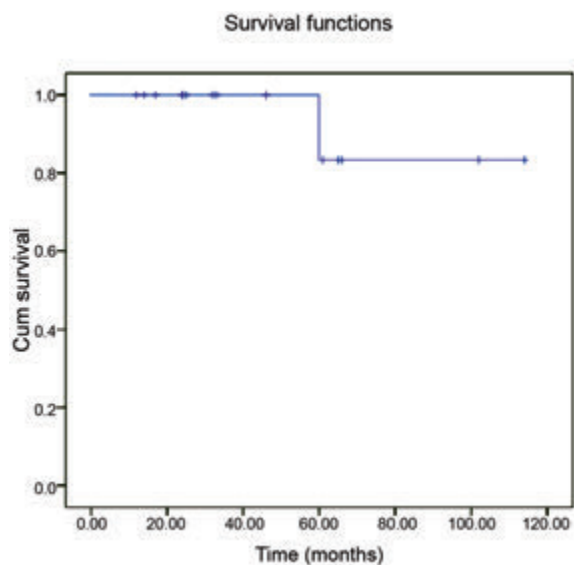


Fig. 14 Survival analysis of patients who had CGR (n = 16) by Kaplan-Meier method. The 5-year overall survival was 83.3%.

vague abdominal discomfort and/or distension and/or pain. The mean duration of symptoms of 3.5 months in our patients is not different from those reported previously^(23,24). In our experience, the reasons for late presentation of such large retroperitoneal mass may be one or more of the following. Firstly, RSTS grew slowly in the silent retroperitoneal area without notice until it became sizable. Secondly, some patients had

local recurrence after previous surgical resection elsewhere and did not recognize their recurrence until symptoms occurred. Thirdly, some patients underwent extensive investigations that took several months without obtaining definite diagnosis before transferring to us. Lastly, some patients came to us by themselves after being considered of having an unresectable tumor from the other hospitals. In our current practice, we usually make preoperative diagnosis of RSTS from abdominal CT scan and proceed to surgical resection without obtaining preoperative tissue diagnosis by fine needle aspiration or core needle biopsy. Diagnosis of RSTS can be made with a high certainty from the current available multidetector computed tomography (MDCT). We agree with several investigators that preoperative fine needle aspiration or core needle biopsy is not necessary when surgical resection of RSTS is planned^(7,25,26). Some investigators recommend preoperative core needle biopsy only when complete surgical resection is not possible and radiation and/or chemotherapy are the first line of therapy or when diagnosis of RSTS is inconclusive⁽²⁵⁻²⁷⁾. Some investigators also recommend percutaneous core needle biopsy to obtain tissue diagnosis when neoadjuvant therapy is under consideration⁽²⁶⁾.

Surgical resection of the RSTS is generally accepted as the most effective treatment modality for curative purpose^(5,9,10). Unfortunately, the common presentation of large RSTS at the time of diagnosis frequently prohibits a microscopically negative margin of the resected tumor and adjuvant radiation and/or chemotherapy are under intensive investigation in order to control local recurrence and possible improvement of survival. Our policy is to performed CGR en-bloc with the contiguous organs every time we operate on the patient to minimize the chance of local recurrence with subsequent long-term survival. CGR was employed in almost all patients in our case series. The only two patients who had palliative resection of the tumor had had multiple sites tumors (sarcomatosis) in one and had previous multiple resections elsewhere with tumor involvement of the superior mesenteric artery in the other. In spite of aggressive en-bloc CGR with multivisceral organ resection in our patients, nine out of 16 (56.3%) had microscopically uninvolved margins and seven (43.7%) had microscopically involved margins. These findings indicate the limitation of surgery in patients with RSTS. We believe that we had performed adequate CGR since the rate of multivisceral organ resection in our patients was relatively high (all patients had one or

more contiguous organ resection, 87.5% nephrectomy, 50% right or left colectomy) compared with those previously reported (16-78.5% nephrectomy, 12-65% colectomy)^(6,7,10,17,19,28). Our study confirmed the necessity of contiguous organ resection for oncologic point of view since 50% of the resected organs had tumor involvement. The reasons explaining CGR with microscopic margin positive (R1 resection) in our patients may be, firstly, our patients had relatively large size tumors (median 27 cm, range 10 to 48 cm), and secondly, 68.8% of our patients who had CGR had high-grade (grade 2 and 3 tumors). In our study, four out of seven patients (57.1%) who had CGR with microscopically positive margins had local recurrence. Of the nine patients who had CGR with microscopically negative margins, only one (11.1%) had recurrence. However, no statistical significance of the local recurrence in both groups of patients could be demonstrated ($p = 0.101$). A longer follow-up period and larger number of patients are required for more precise conclusion of the occurrence of recurrence RSTS which is well-known for its mysterious nature.

CGR with resection of contiguous organs has now become a recommended surgical procedure for RSTS^(9,10,28). Although the surgical margins may be microscopically involved in large tumor and recurrence may occur even in R0 resection, CGR seems to be oncologically reasonable since the contiguous organs are not peeled out from the tumor but are resected instead. We support such en-bloc resection and have shown that the surgical complications are low without perioperative mortality. However, the operations are technically demanding and the surgical procedures are deserved to be discussed. The operation should be carried out with a long midline incision as suggested by Strom and Mahvi⁽²⁰⁾. The decision to do colectomy should be made early during the operation. Dissection should be started at midline of the abdomen by carefully separate the IVC, aorta and iliac vessels from the tumor. The tumor becomes relatively free from the surrounding structures when the renal artery and vein are ligated and divided. If the tumor is quite large and the space to dissect midline IVC and aorta from the tumor is compromised, partial mobilization of the tumor from the lateral and posterior abdominal wall muscles will solve this problem and allows more space for midline dissections. Aggressive mobilization of the tumor from the lateral and posterior abdominal wall muscles before complete vascular control at the midline vessels should be avoided because it may result in uncontrollable hemorrhage from the IVC and aorta or

their branches. This precaution has been previously mentioned⁽²⁰⁾. After the tumor has been removed from the operative field, complete hemostasis with No. 2-0 silk suture ligatures should be performed to the lumbar arteries and arterial hemorrhage from the posterior and lateral abdominal wall muscles. Postoperatively, if there is evidence of intraabdominal hemorrhage, reoperation should be performed expeditiously since the large area of raw surface after tumor removal may contain several liters of blood coming out from the lumbar arteries and muscles of the lateral and posterior abdominal wall. Failure of early detection and control these bleeding points may lead to disastrous consequences.

Postoperative adjuvant radiation and/or chemotherapy are under consideration of our radiotherapists and oncologists. The decision for radiation or chemotherapy depends on microscopic margins of the tumor, tumor size, tumor grading, and general physical health of the patients. Recently, preoperative radiotherapy has been extensively studied^(8,26,27,29). The advantages of preoperative radiotherapy are, firstly, avoidance of toxicity of radiation to the visceral organs since they are displaced by the tumor bulk out of the radiation field and secondly, potentially prevention of local recurrence at the tumor bed of the posterior abdominal wall^(8,26,27,29). We started administration of preoperative radiotherapy in September 2011 to the last patient in our case-series. During operation, five weeks after radiotherapy, we observed that there was a loose plane between the tumor and structures of the posterior abdominal wall including the aorta and posterior abdominal wall muscles possibly due to responsive of the tissue at the radiated area. Dissections along this loose areolar plane could be easily performed with minimal blood loss. This patient underwent en-bloc resection of the retroperitoneal liposarcoma including total mass removal, left colectomy, left nephrectomy and adrenalectomy, splenectomy, distal pancreatectomy, and total abdominal hysterectomy with bilateral salpingo-oophorectomy. Postoperatively, the patient recovered well but developed uncomplicated pancreatic fistula, which was successfully treated by conservative management. Although we have limited experience with preoperative radiotherapy patients, our observation described above is quite attractive in the surgical point of view.

Surveillance of local recurrence and distant metastasis in our patients was performed by CT scan of the chest and abdomen every three to six months.

Four out of five patients (80%) with local recurrence without distant metastasis in our study underwent re-resection. While preparing this manuscript, all four patients are still well. Re-resection of the local recurrence in patients with RSTS has been reported but the outcome is under investigation owing to the rarity of the disease⁽³⁰⁾. However, re-resection of the local recurrence in selected cases was recommended by several investigators^(31,32). In patients who had local recurrence after the first resection elsewhere and underwent subsequent CGR at King Chulalongkorn Memorial Hospital (three patients), all are still well at last follow-up.

Our local recurrence rate after CGR was 31.3%. The present results are not different from those reported from the cancer centers in Europe and North America^(5,7,9,10,13). The 5-year overall survival is also comparable to other experienced cancer centers^(5,7,9,10,13). These meaningful results are secondary from our intention to perform CGR with contiguous organ resection in all patients. With the modern technology of multislice CT scan, preoperative evaluation for resectability of large RSTS is highly accurate resulting in uniformly successful aggressive surgical resection with low complications. Our favorable outcomes were obtained in spite of a relatively larger size of tumor than those previously reported (range 10-48 cm, median 27 cm in our study vs. previously reported median of 11 cm, 16 cm, 18 cm, 11 cm, 14 cm, 15 cm, and 17 cm, by Ballo et al, Gronchi et al, Bonvalot et al, Nishimura et al, Cho et al, Alvarenga et al, and Mussi et al, respectively)^(8-12,18,28).

Management of RSTS requires a multi-disciplinary approach with cooperative efforts of surgeons, radiologists, anesthesiologists, pathologists, radiotherapists, and oncologists. Postoperative adjuvant radiation and chemotherapy have been commonly practiced at King Chulalongkorn Memorial Hospital for years. The first case of preoperative radiotherapy at King Chulalongkorn Memorial Hospital was in September 2011. Although the effectiveness of this adjunctive therapy to control local recurrence is still inconclusive, they provide the only acceptable alternative of treatment for this highly lethal disease.

Our preliminary results of treatment of patients with RSTS may have some shortcomings. The relatively low number of patients and a short period of follow-up are the two major disadvantages. However, the outcomes of our study in term of surgical morbidity and mortality, local recurrence rate, and 5-year overall

survival have inspired us to continue collecting the data for long-term investigations. It would take some time to make more solid conclusions of this entity since the disease is extremely rare.

In conclusion, the authors present patients with RSTS who underwent surgical resection at King Chulalongkorn Memorial Hospital during the 9.4-year period. All except two had CGR with en-bloc resection of the contiguous organs. Surgical techniques are challenging owing to the enormous size of the tumor in most instances. The rarity of the disease is an important factor causing slow learning period of the low-volume institution resulting in poor outcomes. The outcomes of this study are promising with the local recurrence rate and 5-year overall survival comparable to the cancer institutions in the other Asian or Western countries. However, further study with a larger number of patients and longer period of follow-up are required for more meaningful interpretation of outcomes.

Potential conflicts of interest

None.

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ประสบการณ์ของการผ่าตัดรักษาเรโทรเพอริโตเนียล ซอฟท์ทิสซู ชาร์โคมา ที่โรงพยาบาลมหาวิทยาลัยแห่งหนึ่ง
ในประเทศไทย

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ภูมิหลัง: เรโทรเพอริโตเนียล ซอฟท์ทิสซู ชาร์โคมา เป็นมะเร็งที่พบได้น้อยมาก และมีลักษณะเฉพาะตัวคือโตขึ้นอย่างช้าๆ ในบริเวณหลังเยื่อช่องท้องทางด้านหลัง โดยไม่มีอาการในระยะแรก ผู้ป่วยมักมาพบแพทย์เมื่อก่อนมีขนาดใหญ่มาก โดยก้อนมะเร็งจะอยู่ติดหรือถูกห่อหุ้มล้อมโดยหลอดเลือดใหญ่และอวัยวะต่างๆ ภายในช่องท้อง ทำให้การผ่าตัดรักษาให้ได้ผลดีทำได้ยากมากและมีความเสี่ยงสูง วัตถุประสงค์ของการศึกษานี้เพื่อแสดงให้เห็นถึงประสบการณ์ของการผ่าตัดรักษามะเร็งชนิดนี้ที่โรงพยาบาลจุฬาลงกรณ์ กรุงเทพมหานคร และศึกษาถึงผลการรักษา ตลอดจนบรรยายถึงเทคนิคการผ่าตัดที่ค่อนข้างจะซับซ้อนของมะเร็งชนิดนี้

วัสดุและวิธีการ: เป็นการศึกษาย้อนหลังของผู้ป่วยมะเร็งชนิดเรโทรเพอริโตเนียล ซอฟท์ทิสซู ชาร์โคมา ที่ได้รับการผ่าตัดรักษาที่โรงพยาบาลจุฬาลงกรณ์ ตั้งแต่เดือนมิถุนายน พ.ศ. 2546 ถึง เดือนพฤศจิกายน พ.ศ. 2554 ผู้ป่วยทุกรายได้รับการติดตามหลังการผ่าตัดจนเสียชีวิตหรือมาตรวจที่ห้องตรวจผู้ป่วยนอกจนถึงเดือนตุลาคม พ.ศ. 2555 ข้อมูลที่เก็บรวบรวมได้แก่ ข้อมูลทั่วไปของตัวผู้ป่วยและก้อนมะเร็ง, รายละเอียดของการผ่าตัด, ภาวะแทรกซ้อนหลังผ่าตัด, ข้อมูลการรักษาโดยการฉายแสงและ/หรือการให้ยาเคมีบำบัดทั้งก่อนและหลังการผ่าตัด, การเกิดซ้ำของก้อนมะเร็ง, การรักษามะเร็งที่เกิดซ้ำ และอัตราการมีชีวิตรอดเมื่อเวลาผ่านไป 5 ปี นอกจากนี้คณะผู้นิพนธ์ยังได้วิเคราะห์ถึงปัจจัยที่อาจมีความเกี่ยวข้องกับการเกิดซ้ำของมะเร็งภายหลังการผ่าตัดด้วย

ผลการศึกษา: ในช่วงระยะเวลา 9 ปี 5 เดือน ที่ศึกษา มีผู้ป่วยเข้ารับการผ่าตัดจำนวน 18 ราย ผู้ป่วย 14 ราย (ร้อยละ 77.8) เป็นหญิง และ 4 ราย (ร้อยละ 22.2) เป็นชาย ผู้ป่วยมีอายุระหว่าง 44 ถึง 80 ปี (ค่ามัธยฐาน 53.5 ปี) ระยะเวลาที่มีอาการอยู่ระหว่าง 1 สัปดาห์ ถึง 24 เดือน (ค่ามัธยฐาน 3.5 เดือน) ขนาดของก้อนมะเร็งอยู่ระหว่าง 10 ถึง 48 เซนติเมตร (ค่ามัธยฐาน 27 เซนติเมตร) โดยวัดตรงส่วนที่กว้างที่สุดเมื่อผ่าตัดเอาก้อนมะเร็งออกมา ผู้ป่วยทุกรายได้รับการตรวจภาพถ่ายรังสีคอมพิวเตอร์ของช่องท้องก่อนผ่าตัด ผู้ป่วย 1 ราย ได้รับการเจาะตัดชิ้นเนื้อมาตรวจก่อนผ่าตัด ผู้ป่วย 1 ราย ได้รับการฉายรังสีรักษาก่อนการผ่าตัด ผู้ป่วย 16 ราย (ร้อยละ 88.9) ได้รับการผ่าตัดชนิดเอาก้อนมะเร็งออกหมด ผู้ป่วย 2 ราย (ร้อยละ 11.1) ได้รับการผ่าตัดแบบบรรเทาอาการโดยมีก้อนมะเร็งหลงเหลืออยู่ ผู้ป่วยที่ได้รับการผ่าตัดชนิดเอาก้อนมะเร็งออกหมดได้รับการตัดอวัยวะในช่องท้องที่อยู่ติดกับก้อนมะเร็งทุกราย โดยมีตั้งแต่ตัดออกเพียง 1 อวัยวะ จนถึงหลายอวัยวะพร้อมๆ กัน (ผู้ป่วยร้อยละ 87.5 ได้รับการตัดไตออก 1 ข้าง, ร้อยละ 50 ได้รับการตัดลำไส้ใหญ่ด้านซ้ายหรือด้านขวาออก, ร้อยละ 43.7 ได้รับการตัดต่อมหมวกไตออก 1 ข้าง) ระยะเวลาผ่าตัดอยู่ระหว่าง 120 ถึง 360 นาที (ค่ามัธยฐาน 330 นาที) ผู้ป่วยได้รับเลือดระหว่างผ่าตัดตั้งแต่ 0 ถึง 12 ยูนิต (ค่ามัธยฐาน 2.5 ยูนิต) ผู้ป่วย 3 ราย มีภาวะเลือดออกภายหลังการผ่าตัดและต้องรับการผ่าตัดซ้ำ ผู้ป่วย 1 ราย มีภาวะน้ำย่อยตับอ่อนรั่วชนิดไม่รุนแรงภายหลังการผ่าตัด ซึ่งหายเองโดยการรักษาแบบประคับประคอง ไม่มีผู้ป่วยเสียชีวิตจากการผ่าตัดในรายงานนี้ ผลการตรวจทางพยาธิวิทยาพบว่าผู้ป่วย 15 ราย (ร้อยละ 83.3) เป็นมะเร็งชนิดไลโปซาร์โคมา ผู้ป่วยที่เหลือ 3 ราย เป็นมะเร็งชนิดเอทิปิปกอลไลโปมาตัสทิวเมอร์, มาลิกันเนนท์ ไฟบรอส อีสทิโอซายโคมา, และสปีนเดิลเซลล์ทิวเมอร์ชนิดไม่แบ่งประเภทแน่นอนอย่างละ 1 ราย ผลการติดตามผู้ป่วยหลังผ่าตัดพบว่าผู้ป่วย 2 ราย ที่ได้รับการผ่าตัดแบบบรรเทาอาการเสียชีวิตเมื่อ 6 และ 16 เดือนหลังผ่าตัด ในผู้ป่วยที่ได้รับการผ่าตัดชนิดเอาก้อนมะเร็งออกหมดมีการเกิดซ้ำของมะเร็ง 5 ราย (ร้อยละ 31.3) และเสียชีวิต 1 ราย เมื่อ 60 เดือนหลังผ่าตัด ผู้ป่วยที่ได้รับการผ่าตัดชนิดเอาก้อนมะเร็งออกหมดมีค่ามัธยฐานของเวลาที่ติดตามผู้ป่วย 39.5 เดือน (พิสัย 12-114 เดือน) อัตราการรอดชีวิตเมื่อเวลาผ่านไป 5 ปี ของผู้ป่วยทั้งรายงานอยู่ที่ร้อยละ 73.5 (ช่วงความเชื่อมั่นร้อยละ 95: ร้อยละ 44.3-88.4) อัตราการรอดชีวิตเมื่อเวลาผ่านไป 5 ปีของผู้ป่วยที่ได้รับการผ่าตัดชนิดเอาก้อนมะเร็งออกหมดอยู่ที่ร้อยละ

83.3 (ช่วงความเชื่อมั่นร้อยละ 95: ร้อยละ 53.5-98.5) จากการวิเคราะห์ถึงปัจจัยที่อาจมีความเกี่ยวข้องกับการเกิดซ้ำของมะเร็ง
ภายหลังการผ่าตัดชนิดเอกาอ่อนมะเร็งออกหมด ไม่พบความแตกต่างอย่างมีนัยสำคัญทางสถิติของขนาดของก้อนมะเร็ง, ลักษณะ
ทางพยาธิวิทยา, การมีมะเร็งหลงเหลือที่ขอบเนื้อเยื่อที่ตัดออกมา, และลักษณะการผ่าตัด (ครั้งแรกหรือผ่าตัดซ้ำ) ในผู้ป่วยที่มีและ
ไม่มีมะเร็งเกิดซ้ำ

สรุป: รายงานนี้ถึงแม้จะมีผู้ป่วยจำนวนไม่มาก แต่ก็มีความสำคัญที่ได้แสดงให้เห็นว่า การรักษามะเร็งชนิดเรทโทรเพอริโตเนียล
ซอพท์ทิสซู ชาร์โคมา ให้ได้ผลดี สามารถทำได้โดยการผ่าตัดเอามะเร็งออกให้หมด คณะผู้นิพนธ์ได้รายงานถึงการผ่าตัดที่ซับซ้อนที่
สามารถทำได้อย่างปลอดภัย ถึงแม้ก้อนมะเร็งจะมีขนาดใหญ่ การตรวจภาพถ่ายรังสีคอมพิวเตอร์ของช่องท้องก่อนผ่าตัดมีความสำคัญ
ในการวินิจฉัยและวางแผนการรักษา การเจาะตัดชิ้นเนื้อก่อนผ่าตัดเพื่อการวินิจฉัยไม่มีความจำเป็นถ้าการตรวจภาพรังสีคอมพิวเตอร์
ของช่องท้องให้การวินิจฉัยได้แน่นอนแล้ว ความตั้งใจในการผ่าตัดรักษานอกเอกาอ่อนมะเร็งออกให้หมดมีความสำคัญและควรพิจารณา
ทำทุกครั้งที่ผ่าตัดผู้ป่วยเหล่านี้เพื่อให้ผลการรักษาที่ดีที่สุด กล่าวคืออัตราการเกิดซ้ำของมะเร็งน้อยที่สุดและผู้ป่วยมีอายุยืนยาวที่สุด
ประสิทธิภาพของทีมงานศัลยกรรมมีความสำคัญที่จะทำให้การผ่าตัดที่ยากและมีความเสี่ยงสูงนี้สำเร็จลุล่วงด้วยดี
