UCD: 617-089. 04
IRSTI: 76.29.49.
DOI: 10.24412/2790-1289-2022-4-8083

RETROPERITONEAL LEIOMYOSARCOMA FATAL SPONTANEOUS RUPTURE: LITERATURE REVIEW AND PRESENTATION OF EXTREMELY RARE CASE

[1, 2] Talgat Tajibayev, [2] Galimzhan Esenaliev, [2] Nurlybek Omarov, [2] Alikhan Abdreshit,
[1, 2] Dilyar Maripzhanov
[1] NEI «Kazakh-Russian Medical University», Kazakhstan, Almaty
[2] Talgar Cities Central Regional Hospital, Kazakhstan, Talgar

Summary

Retroperitoneal sarcoma (RPS) is a heterogeneous group of malignant tumors with an incidence of approximately 1 case per 100,000 population per year, which includes leiomyosarcoma. Leiomyosarcoma is a malignant neoplasm with smooth muscles differentiation. It is the second most common sarcoma affecting the retroperitoneal space.

In this article, we describe a case of fatal hematoma of the retroperitoneal space, caused by spontaneous rupture of leiomyosarcoma. A 65-year-old woman presented with severe pain in her left side and hypovolemic shock. After an additional examination of hemodynamic stabilization, she was operated on an emergency basis. The tumor was removed and the bleeding was stopped. The patient was transferred to the intensive care unit (ICU) after surgery, despite the ongoing intensive therapy, the condition remained extremely serious, against which the deterioration result from cardiac arrest and death is occurred.

In the world literature, spontaneous rupture of retroperitoneal leiomyosarcoma is described in isolated cases. Moreover, the main publications are aimed at managing patients with tumors of the retroperitoneal space only in a planned manner. Thus, this case could be of clinical interest among emergency medical practitioners as well as in the scientific community. Literature search for a review of the problem was carried out in the following scientific databases and search engines: PubMed, Web of Science, Scopus, Google Scholar, eLIBRARY.

Key words: *retroperitoneal leiomyosarcomas, spontaneous rupture of leiomyosarcoma, retroperitoneal sarcoma, rupture, hemoperitoneum, retroperitoneal hematoma.*

Introduction. Retroperitoneal sarcoma (RPS) represents a heterogeneous group of malignant tumours with an incidence of approximately one per 100 000 population per year [1]. Leiomyosarcoma is a malignant neoplasm that shows smooth muscle differentiation. It is the second most common sarcoma to affect the retroperitoneum. Retroperitoneal leiomyosarcomas can grow to large sizes before detection and may be an incidental finding at imaging. When symptomatic, retroperitoneal leiomyosarcoma may cause compressive symptoms, including pain. Retroperitoneal leiomyosarcoma most commonly manifests as a large soft-tissue mass, with areas of necrosis [2]. Winan J van Houdt et al described a new randomized trial, STRASS-2, to analyze the role of neoadjuvant chemotherapy for high-grade liposarcoma and leiomyosarcoma of the retroperitoneum [3].

Treatment of retroperitoneal sarcomas is complex and all patients should be treated in multidisciplinary sarcoma centers. Liposarcomas tend to recur locally, whereas distant recurrences are more often seen in leiomyosarcoma and other subtypes. Outcome improves when patients are treated in high volume sarcoma centers [4]. The following articles describe emergency cases of spontaneous rupture and surgical approaches of retroperitoneal sarcomas.

The article by Grasso M. et al. Revieweda case of spontaneous rupture of leiomyosarcoma in a 45-year-old woman, presenting with severe left flank pain and perirenal hemorrhage [5].The case of retroperitoneal leiomyosarcoma with extra- and intravascular invasion described a involvement of inferior vena cava by leiomyosarcoma, which may be locally resected in some cases but has a poor long-term survival rate[6].In the case of a 68-year-old man with the main complaint of left flank pain, was described diagnostic difficulty because of the absence of the characteristic diagnostic signs in this malignant tumor [7]. Aksoy Y. et al described the first report of spontaneous rupture of renal angiosarcoma and a cause of retroperitoneal hematomas [8]. The first description of tumor rupture with retroperitoneal hematoma was mentioned in a case of a 31-year-old man with complaining of acute violent pain of the right lower abdominal quadrant [9]. Spontaneous RPS rupture exposes the patient to peritoneal seeding and sarcomatosis and is associated with a poor prognosis [10]. Haemorrhagic shock caused by an acute bleeding from a retroperitoneal liposarcoma at first presentation is extremely rare, and there are very few published cases in the literature [11]. Sarcoma surgery is rarely performed on an urgent or emergent basis. The reportof a retroperitoneal leiomyosarcoma that presented with spontaneous rupture and hemoperitoneum, which required surgical treatment in an urgent manner, was earlier reported in the literature [12].

We describe a case of spontaneous rupture of leiomyosarcoma with extravascular involvement 65-year-old woman.

Case presentation. A 65-year-old woman was admitted on an emergency room with complaints of a sharp pain in

the left hypogastrium that appeared suddenly, swelling in the left hypogastrium, weakness, pallor of the skin. She has a history of coronary heart disease, arterial hypertension, underwent an appendectomy 30 years ago. The general condition of the patient upon admission is extremely severe, due to hypovolemic shock, hemodynamics was unstable. The patient has a hypersthenic constitution, overweight. Consciousness is stunning, according to the Glasgow coma scale 12-13 points, the position is forced on a gurney. The skin is pale, covered with cold and sticky sweat. Respiratory system: respiratory rate is 22 per minute, rhythmic. Auscultation of the lung: vesicular breath sounds are heard over lung fields, no adventitious sounds. Comparative percussion: clear pulmonary sound over the fields of both lungs. Cardiovascular system: muffled heart sounds heard during auscultation. There are no pathological murmurs. BP is 70/40 mmHg, heart rate is 130 beats per minute, a weak pulse.

Status localis: Tongue dry, coated by a white film. The abdomen is swollen, asy mmetric due to an oval protrusion in the left hypogastrium with a diameter of up to 40 cm, sharply painful on palpation, systolic bruit is not detected on auscultation. Symptoms of peritoneal irritation are positive. The liver at the edge of the costal arch, gallbladder, pancreas are not palpable. According to the laboratory tests, there were signs of severe post-hemorrhagic anemia.

The patient immediately was hospitalized in the intensive care unit. After stabilization of hemodynamics, the patient underwent an abdominal CT scan which revealed "Formation of the retroperitoneal space on the left is not excluded. Retroperitoneal hematoma" (Figure 1)

The patient was taken for emergency surgery. A vascular surgeon was called urgently and a laparotomy was performed. There was an extensive retroperitoneal hematoma in the retroperitoneal space (Figure 2). The revision revealed that the formation surrounds the left kidney and intimately adjoins the left renal artery and vein, but there are no signs of damage or bleeding of the main vessels. Bleeding was detected from the rupture of the formation, the removal of the formation of the retroperitoneal space was performed. Unstable hemodynamic persisted during the operation, which was supported by cardiotonic drugs; anuria was noted.

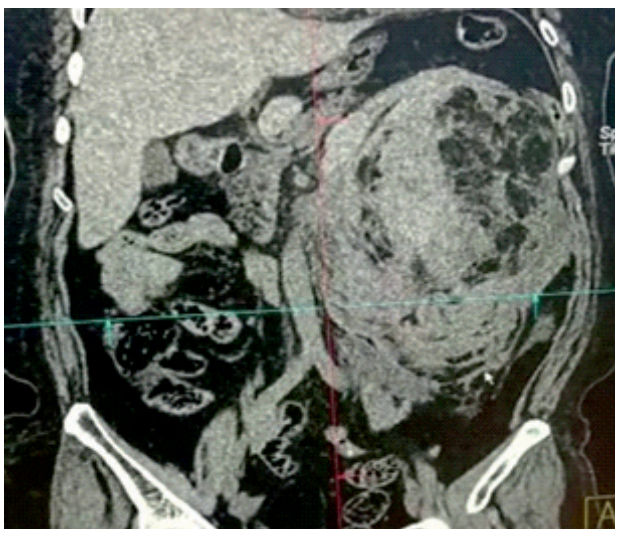

Figure 1. *CT scan: Retroperitoneal formation, 15x20 cm.*

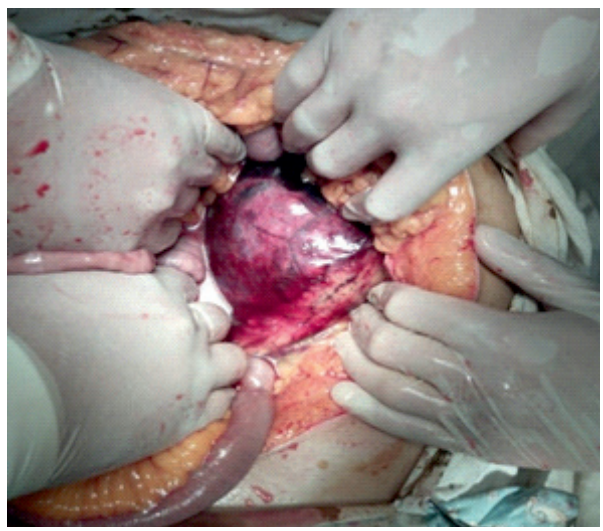

Figure 2. *Intraoperativeview.*

The patient was transferred to the ICU after surgery, despite the ongoing intensive therapy, the condition remained extremely serious, against which the deterioration result from cardiac arrest and death is occurred. Resuscitation measures were urgently started. Despite ongoing resuscitation, cardiac activity could not be restored. Biological death was declared.

Histological examination (staining with hematoxylin-eosin): Leiomyosarcoma. Secondary hemorrhages in tumor cells with areas of necrosis (Figure 3A, 3B).

Figure 1. *CT scan: Retroperitoneal formation, 15x20 cm.*

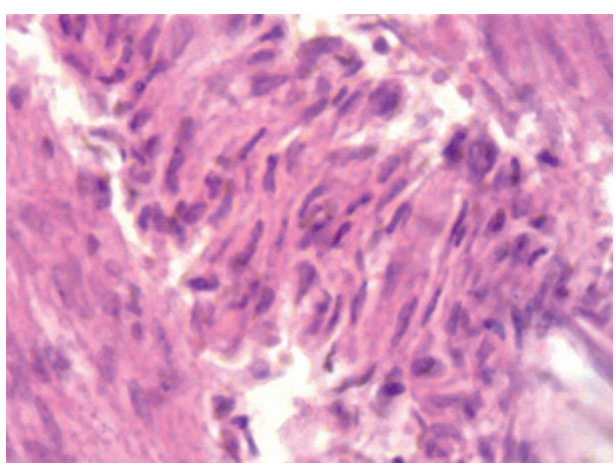

Figure 2. *Intraoperativeview.*

Conclusion. Retroperitoneal sarcomas are well known to present difficulties in their complete resection because of their inaccessible location and the absence of early symptoms, resulting in tumors of large size by the time the diagnosis is made. In this report, we describe a case of a retroperitoneal leiomyosarcoma that presented with spontaneous rupture and hemoperitoneum, which required surgical treatment in an urgent manner. Thus, this case could be of clinical interest among emergency medical practitioners as well as in the scientific community.

References:

1. L.M. Almond, F. Tirotta, H. Tattersall, J. Hodson, T. Cascella, M. Barisella, A. Marchianò, G. Greco, A. Desai, S.J. Ford, A. Gronchi, M. Fiore, C. Morosi, Diagnostic accuracy of percutaneous biopsy in retroperitoneal sarcoma, British Journal of Surgery, Volume 106, Issue 4, March 2019, Pages 395 – 403.

2. Marko J., Wolfman D.J. Retroperitoneal Leiomyosarcoma from the Radiologic Pathology Archives. Radiographics. 2018 Sep-Oct; 38 (5): 1403-1420. doi: 10.1148/rg.2018180006. PMID: 30207936; PMCID: PMC6166742.

3. van Houdt W.J., Raut C.P., Bonvalot S., Swallow C.J., Haas R., Gronchi A. New research strategies in retroperitoneal sarcoma. The case of TARPSWG, STRASS and RESAR: making progress through collaboration. Curr Opin Oncol. 2019 Jul; 31 (4): 310-316. doi: 10.1097/CCO.0000000000000535. PMID: 30893150.

4. van Houdt W.J., Zaidi S., Messiou C., Thway K., Strauss D.C., Jones R.L. Treatment of retroperitoneal sarcoma: current standards and new developments. Curr Opin Oncol. 2017 Jul; 29 (4): 260-267. doi: 10.1097/CCO.0000000000000377. PMID: 28509807.

5. Grasso M., Blanco S., Fortuna F., Crippa S., Di Bella C. Spontaneous rupture of renal leiomyosarcoma in a 45-year-old woman. Archivos Espanoles de Urologia. 2004 Oct; 57 (8): 870-872. PMID: 15560282.

6. Cantwell C.P., Stack J. Abdominal aortic invasion by leiomyosarcoma. Abdom Imaging 31, 120 – 122 (2006). https://doi.org/10.1007/s00261-005-0163-5.

7. Tamaki M., Terai A., Terachi T., et al. [A case of incidental renal leiomyosarcoma] Hinyokikakiyo. Acta Urologica Japonica. 1994 May; 40 (5): 415-418. PMID: 8023763.

8. Aksoy Y., Gürsan N., Ozbey I., Biçgi O., Keleş M. Spontaneous rupture of a renal angiosarcoma. Urol Int. 2002; 68 (1): 60-2. doi: 10.1159/000048419. PMID: 11803270.

9. Girszyn N., Mabro M., Kahn J.E., Roullet-Audy J.C., Sautet A., Bletry O. Synovialosarcomerétropéritonéalré vélé par un choc hémorragique [Primary retroperitoneal synovial sarcoma revealed by hemorrhagic shock]. Presse Med. 2006 Jul-Aug;35(7-8):1185-7. French. doi: 10.1016/s0755-4982 (06) 74778-9. PMID: 16840897.

10. Samà L., Tzanis D., Bouhadiba T., Bonvalot S. Emergency Retroperitoneal Sarcoma Surgery for Preoperative Rupture and Hemoperitoneum: A Case Report. Cureus. 2021 Mar 17; 13(3): e13936. doi: 10.7759/cureus.13936. PMID: 33868865; PMCID: PMC8050804.

11. Al Sheikh M., Simson N., Obi-Njoku O., Qteishat A. Acute haemorrhage from a retroperitoneal liposarcoma: a rare presentation. BMJ Case Rep. 2018 Feb 8;2018: bcr 2017222737. doi: 10.1136/bcr-2017-222737. PMID: 29437802; PMC43.

12. Samà L., Tzanis D., Bouhadiba T., et al. (March 17, 2021) Emergency Retroperitoneal Sarcoma Surgery for Preoperative Rupture and Hemoperitoneum: A Case Report. Cureus 13 (3): e13936. doi:10.7759/cureus.13936.

РЕТРОПЕРИТОНЕАЛДЫ КЕҢІСТІК ЛЕОМИОСАРКОМАСЫНЫҢ СОНТАНДЫ ЖАРЫЛЫС: ӘДЕБИЕТКЕ ШОЛУ ЖӘНЕ СИРЕК КЕЗДЕСЕТІН ЖАҒДАЙДЫ СИПАТТАУ

[*1, 2] Т.К. Таджибаев, [2] Г.К. Есеналиев, [2] Н.О. Омаров, [2] А.М. Абдрешит, [1, 2] Д.М. Марипжанов

[1] «Қазақстан - Ресей медициналық университеті» МЕББМ, Қазақстан, Алматы

[2] Талғар қаласының орталық аудандық ауруханасы, Қазақстан, Талғар

Түйінді

Ретроперитонеальді саркома (РПС) қатерлі ісіктердің гетерогенді тобы болып табылады, жылына 100 000 адамға шаққанда 1 жағдай жиілігімен, оған лейомиосаркома кіреді. Лейомиосаркома - тегіс бұлшықеттердің дифференциациясы бар қатерлі ісік болы саналады. Бұл ретроперитонеальды кеңістікке әсер ететін екінші ең таралған саркома.

Бұл мақалада біз ретроперитонеальды кеңістіктің өлімге әкелетін гематомасының жағдайын сипаттаймыз, оның себебі лейомиосаркома ісігінің спонтанды жарылуы болды. 65 жастағы әйел сол жақ бөлігіндегі дереу ауру сезіміне және гиповолемиялық шокқа шағымданды. Гемодинамиканың тұрақтануын қосымша тексеруден кейін шұғыл түрде ота жасалып, ісік алынып, қан тоқтатылды. Операциядан кейін науқас анестезиология және реанимация бөлімшесіне ауыстырылды, жүргізіліп жатқан интенсивті терапияға қарамастан, жағдайы өте ауыр болып, жүрегітоқталып және өлімге әкелді.

Әлемдік әдебиеттерде ретроперитонеальді лейомиосаркоманың өздігінен жарылуы оқшауланған жағдайларда сипатталған. Сонымен қатар, негізгі мақалалар ретроперитонеальды кеңістіктегі ісіктері бар науқастарды тек жоспарлы түрде басқаруға бағытталған. Осылайша, бұл жағдай жедел жәрдем дәрігерлері арасында да, ғылыми қоғамда да клиникалық қызығушылық тудыруы мүмкін. Мәселеге шолу жасау үшін әдебиеттерді іздеу келесі ғылыми деректер қорларында және іздеу жүйелерінде жүргізілді: PubMed, Web of Science, Scopus, Google Scholar, eLIBRARY

Кілт сөздер: *ретроперитонеальді лейомиосаркома, лейомиосаркоманың спонтанды жарылуы, ретроперитонеальді саркома, жарылу, гемоперитонеум, ретроперитонеальды гематома.*

ФАТАЛЬНЫЙ СПОНТАННЫЙ РАЗРЫВ ЛЕЙОМИОСАРКОМЫ ЗАБРЮШИННОГО ПРОСТРАНСТВА: ОБЗОР ЛИТЕРАТУРЫ И ОПИСАНИЕ РЕДКОГО СЛУЧАЯ

[*1, 2] Т.К. Таджибаев, [2] Г.К. Есеналиев, [2] Н.О. Омаров, [2] А.М. Абдрешит, [1, 2] Д.М. Марипжанов

[1] НУО «Казахстанско – Российский медицинский универститет», Казахстан, Алматы

[2] Талгарская центральня районная больница, Казахстан, Талгар

Аннотация

Забрюшинная саркома представляет собой гетерогенную группу злокачественных опухолей с частотой возникновения примерно 1 случай на 100 000 населения в год, в которую входит лейомиосаркома. Лейомиосаркома представляет собой злокачественное новообразование с дифференцировкой гладкой мускулатуры. Это вторая по распространенности саркома, поражающая забрюшинное пространство.

В данной статье нами описывается случай фатальной гематомы забрюшинного пространства, причиной которой явился спонтанный разрыв образования – лейомиосаркомы. Шестидесяти пятилетняя женщина поступила с сильными болями в левом боку и гиповолемическим шоком. После дообследования стабилизации гемодинамики оперирована в экстренном порядке, выполнено удаление образования и остановка кровотечения. После операции больная переведена в отделение ОАРиИТ, несмотря на проводимую интенсивную терапию, состояние оставалось крайне тяжелым, на фоне которого наступило ухудшение - остановка сердечной деятельности и смерть.

В мировой литературе спонтанный разрыв лейомиосаркомыза брюшинного пространства описывается в единичных случаях. Более того основные публикации направлены на ведение пациентов с опухолями забрюшинного пространства лишь в плановом порядке. Таким образом, данный случай мог бы представлять клинический интерес среди практикующих врачей экстренной службы, а также в научном обществе. Поиск литературы для обзора проблемы проводился в следующих научных базах данных и поисковых системах: PubMed, WebofScience, Scopus, GoogleScholar, eLIBRARY.

Ключевые слова: *забрюшинные лейомиосаркомы, спонтанный разрыв лейомиосаркомы, забрюшинная саркома, разрыв, гемоперитонеум, забрюшинная гематома.*

Конфликт интересов. *Все авторы заявляют об отсутствии потенциального конфликта интересов, требующего раскрытия в данной статье.*

Корреспондирующий автор. *Таджибаев Талгат Кыдыралиевич, НУО «Казахстанско-Российский медицинский университет», докторант 1 года обучения по специальности 8D10102 – «Медицина», Казахстан, г. Алматы. E-mail: dr.tajibayev@gmail.com; https://orcid.org/0000-0002-9007-063X.*

Вклад авторов. *Все авторы внесли равноценный вклад в разработку концепции, выполнение, обработку результатов и написание статьи. Заявляем, что данный материал ранее не публиковался и не находится на рассмотрении в других издательствах.*

Финансирование. *Отсутствует.*

Статья поступила: *15.11.2022.*

Принята к публикации: *25.11.2022.*

.....

Conflict of interest. *All authors declare that there is no potential conflict of interest requiring disclosure in this article.*

Corresponding author. *Tajibayev Talgat K., NEI «Kazakh-Russian Medical University», doctoral student of 1-year study in the specialty 8D10102 – «Medicine», Kazakhstan, Almaty. E-mail: dr.tajibayev@gmail.com; https://orcid.org/0000-0002-9007-063X.*

Contribution of the authors. *All authors have made an equal contribution to the development of the concept, implementation, processing of results and writing of the article. We declare that this material has not been published before and is not under consideration by other publishers.*

Financing. *Absent.*

Article submitted: *15.11.2022.*

Accepted for publication: *25.11.2022.*