ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

Retroperitoneal Liposarcoma Mimicking a Leiomyosarcoma - A Case Report

Ana Magdalena Bratu, Iulia Alecsandra Salcianu* and Constantin Zaharia

Department of Radiology and Medical Imaging, "Colțea" Clinical Hospital, Bucharest, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

***Corresponding Author**: Iulia Alecsandra Salcianu, Department of Radiology and Medical Imaging, "Colțea" Clinical Hospital, Bucharest, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania.

Received: May 11, 2020; **Published:** May 29, 2020

Abstract

Sarcomas are a large, diverse group of rare neoplasm that arise from mesenchymal cells at any site of the body and are divided into two groups: bone sarcomas and soft tissue sarcomas. Leiomyosarcoma, and liposarcoma are the most common types of retroperitoneal soft-tissue sarcomas.

We present the case of a female patient, aged 80 years, without notable pathological history, presented for asthenia and constipation with relatively early onset. An abdominal and pelvic CT was performed and identified giant tumor mass, 31 cm in largest diameter, with heterogeneous structure, relatively well defined, which occupies the left hemiabdomen and is related to the surrounding structures without infiltration. No images of tumor lymphadenopathy and bone lesions of oncological nature were seen in the examination field.

Keywords: *Leiomyosarcoma; Liposarcoma; CT*

Abbreviations

CT: Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

Sarcomas are a large, diverse group of rare neoplasm that arise from mesenchymal cells [1-3] at any site of the body [4], and are divided by the World Health Organization (WHO) into two broad groups: bone sarcomas and soft tissue sarcomas [2,5], each of which has multiple subtypes.

Soft tissue sarcomas constitute a heterogenous group of neoplasms of various histologies and account less than one percent of all malignancies [3,4,6].

A retroperitoneal tumor is defined as a tumor originating in the retroperitoneal space, with no connection with any of the normal organs, being relatively uncommon, accounting for less than 0.2% of all tumors [7].

Ten to twenty percent of sarcomas of soft tissue have localization in the retroperitoneum [4,6,8,9].

In the Surveillance, Epidemiology and End Results (SEER) database is reported the average annual incidence of retroperitoneal sarcomas of approximately 2.7 cases per million population [10].

Leiomyosarcoma and liposarcoma are the most common types of retroperitoneal soft-tissue sarcomas [3,4,11].

Leiomyosarcomas are rare malignant tumor tumors that may arise from any smooth muscle source [4], representing 5% to 10% of all soft tissue sarcoma [12], and 10% - 37% of all soft tissue sarcomas arising in the retroperitoneum [13-15].

On the other hand, liposarcoma constitute approximately 20% of soft tissue sarcomas [16] and account 41% of these tumors originating within the retroperitoneum [17,18].

Retroperitoneal leiomyosarcoma occurs most commonly in the fifth to seventh decade [4,8,19], with no known predilection for race or gender [4,9] and liposarcoma commonly occur in patients aged 40-60 years, and men and women are equally affected [6,20].

Although leiomyosarcoma commonly arise from the smooth muscle of the uterus and gastrointestinal tract, they may originate in the retroperitoneal space [7,19] and rarely in skeletal tissue [19].

Liposarcomas are located in order of frequency in the head, neck, trunk, mediastinum, upper and lower extremities, gastrointestinal tract, and retroperitoneum [20].

Retroperitoneal tumors typically have vague presenting symptoms [4], commonly non-specific [19] and these patients exhibited few symptoms for an average 3.5 months before presentation [4].

The typical scenario is the discovery of a large abdominal or pelvic mass in a patient who is imaged for abdominal fullness or pain [3].

Case Report

We present the case of a female patient, aged 80 years, known with a surgical history (ectopic pregnancy) without other notable pathological history, was presented for asthenia that appeared about 2 months ago, progressively worsened to the difficulty of maintaining orthostatism and constipation for about 2 weeks. Also, at admission it was discovered severe anemic syndrome, with hemoglobin level of 7.0 g/dl. The patient denied loss of appetite and weight loss, or hematemesis, melena, or hematochezia. She is hospitalized in the internal medicine clinic for specialized investigations.

At the clinical examination at hospitalization, an altered general condition was identified, mucocutaneous pallor, abdominal distension, without signs of peritoneal irritation, with left hemiabdomen enlarged in volume, where a voluminous tumor mass is palpated, with extension in epigastrium, mesogastrium, left hypochondrium and flank, left iliac fossa, of hard consistency, painless, immobile on deep planes and with breathing. As a result of blood tests, negative tumor markers and biologically inflammatory syndrome were identified.

An abdominal x-ray was performed that did not identify the presence of pneumoperitoneum or air-fluid levels, but it was observed that the left flank is completely occupied by an oval opacity with a maximum diameter of 19.5 cm.

Following the general surgery consult, a tumor was identified in the left flank that extends from below the costal rim to the pelvis, partially mobile, imprecisely delimited and it was decided to transfer the patient to the surgery department for management.

Due to the symptomatology and biological results, it was decided to perform upper and lower digestive endoscopy which revealed incomplete distension of the left colon, possibly by extrinsic compression. The rest of the digestive tract was normal.

After the endoscopies, abdominal and pelvic CT was performed and identified that the liver, the spleen, the pancreas, both adrenal glands, the uterus, and bladder were without CT changes with pathological significance, and some right renal cortical cysts up to 2.9 cm. Also, giant tumor mass, with heterogeneous structure, predominantly pseudo-cystic (Figure 1), with maximum diameters of 16.3/12.7/31 cm, with bulging contours, relatively well defined, with high enhancement nodules with peripheral disposition (Figure 2).

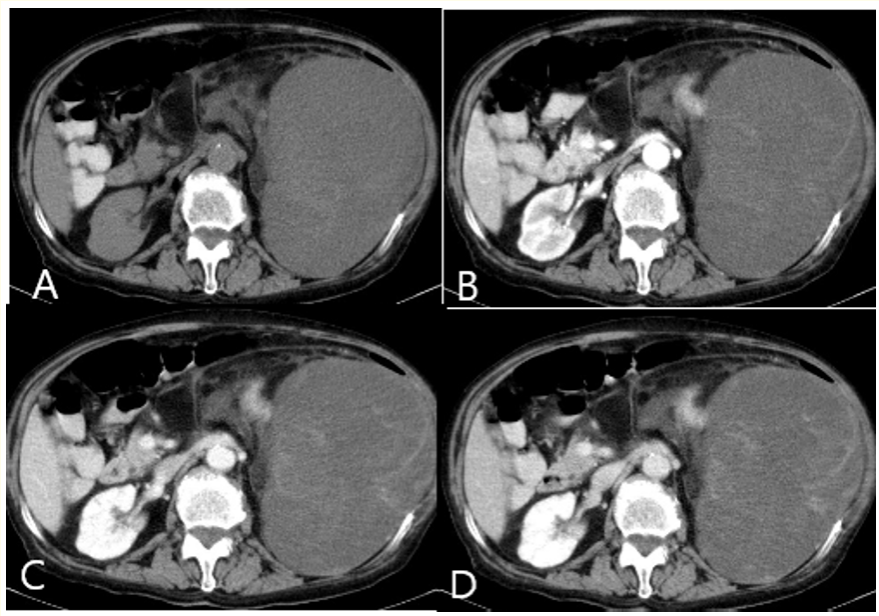

Figure 1: *Abdominal and pelvic CT - A. native examination, B. arterial phase, C. venous phase, D. delayed phase: giant tumor mass, with heterogeneous structure, predominantly pseudo-cystic, with bulging contours, relatively well-defined margins.*

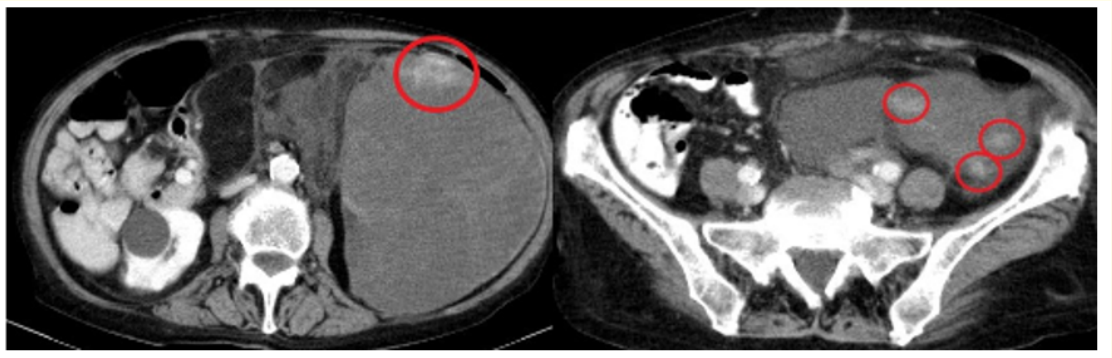

Figure 2: *Abdominal and pelvic CT-venous phase: high enhancement nodules with peripheral disposition (red circles)*

The lesion occupies the left hemiabdomen and is related to the tail of the pancreas, the posterior gastric surface, the splenic vessels, the postero-lateral surface of the left kidney, and the left pelvic ureter, the descending colon, the psoas muscle, the common and external iliac vessels on the left side, and intestinal loops. , on which it exerts a mass effect, but with a demarcation limit. The densification of mesenteric fat was also evident. No images of tumor lymphadenopathy and bone lesions of oncological nature were seen in the examination field.

The CT examination concluded that it was a retroperitoneal tumor with an imaging aspect of leiomyosarcoma, apparently well delimited, without locoregional invasion, having surgical indication.

The excision of the tumor "en bloc" is practiced with all its extensions, and the postoperative evolution is favorable, with good digestive tolerance. She is discharged in good general condition, afebrile, mobile abdomen with breathing, painless spontaneously and on palpation, without signs of peritoneal irritation, intestinal transit present, hemodynamically and respiratory balanced.

The result of the anatomopathological examination was liposarcoma, although the CT examination did not show any fat densities in the lesion, and the characteristics advocated for leiomyosarcoma.

Discussion and Conclusion

Retroperitoneal tumors are relatively rare, accounting for less than 0.2% of all tumors [7] and sarcoma with this localization is a rare tumor accounting for 1% to 2% of all solid malignancies [19].

Between 10 and 15% of soft tissue sarcomas originate within the retroperitoneal space, and the most common type among these is liposarcoma [18]. Liposarcoma is the most common sarcoma in adults and the most commonly occurring sarcoma in the retroperitoneum according to [21]. But overall, they are rare neoplasm, accounting for 0.2 - 0.3% of all malignancies [17].

On the other hand, leiomyosarcoma, represents a represents a percentage of 10% - 37% of all soft tissue sarcomas arising in the retroperitoneum [4].

Liposarcoma is the term for a malignant tumor that has fatty differentiation, and leiomyosarcomas are soft-tissue sarcomas with tumor cells resembling differentiated smooth muscle cells [3].

The retroperitoneum provides a widely expansible anatomic location for tumors arising there and these tumors often become very large before symptoms manifest [9].

Clinical symptoms of abdominal and pelvic sarcomas are most often related to tumor size. Because most liposarcomas occur in the retroperitoneum, they are clinically silent until they grow to large sizes [3], as in our case. The patient presented only asthenia with relatively recent onset, and recently installed constipation, and she denied loss of appetite, and weight loss, or hematemesis, melena, or hematochezia, although anemia was found.

In case of 71-year-old woman presented by Oh., *et al.* [6] it is noticed that the patient had complaint only of progressive abdominal distension, and had not experienced abdominal pain, nausea, vomiting, constipation, dyspepsia, or dyspnea. In another paper [7], are presented two cases of retroperitoneal sarcomas, one with colic invagination, and other with mild left flank pain, and a palpable tender mass in the left lower quadrant of the abdomen.

Todd., *et al.* [9] reported that the most common clinical picture of retroperitoneal sarcoma cases at presentation includes back pain and weight loss (37.5% of patients with either symptom), fatigue (25%), increased abdominal girth (12.5%), and fever or night sweats (12.5%) being also noted.

Other studies of retroperitoneal sarcomas show nonspecific symptoms such as abdominal size increase, intermittent abdominal discomfort, a palpable abdominal mass, and a mild weight loss [4], or vague lower abdominal fullness and pain, tingling sensation on her right lower extremity [19]. Other study reported abdominal pain and weight loss were the most common symptoms [4]. However, in one rapport [7] is shown that the first clinical sign could be massive intraabdominal bleeding due to the rupture of the tumor.

Case history, physical examination, and radio-imaging methods may be used for diagnose. Especially, CT is useful for visualizing location of tumor and association with surrounding organs as well as metastasis and adenopathies [19].

Occasionally, liposarcoma may be discovered when CT is performed for unrelated abdominal symptoms or for vague or nonspecific symptoms [3].

According to Lin., *et al.* [4] a quarter of patients had masses discovered incidentally during routine examination or abdominal surgery. Todd., *et al.* [9] stated that CT was the preferred method for evaluation of a known mass prior to exploratory laparotomy.

CT is the most widely used imaging method for the abdomen and pelvis because of its wide availability, ease of use, and capability to image the breadth of anatomy simultaneously for staging [3].

In case of retroperitoneal sarcomas, CT frequently identifies a large tumor mass [4], with variable weight and dimensions [6], and those over 20 kg are called "giant liposarcomas" and are extremely rare [20].

Oh., *et al.* [6] report a giant retroperitoneal liposarcoma weighing 25 kg, encasing the entire left kidney and adherent to adjacent structures, but which was successfully removed.

Lewis., *et al.* [22] presents that 77% of his 500 patients diagnosed retroperitoneal leiomyosarcoma had tumors larger than 10 cm and the other 23% had 5 to 10 cm sized mass. In our case the tumor measured a maximum diameter of 31cm. In a paper [3] is stated that liposarcomas may be more than 20 cm in largest diameter at the time of diagnosis. Pain, symptoms of mass effect, organ compression, or physical deformity of the abdominal cavity bring patients to clinical attention.

But, Khan., *et al.* [23] and Lin [4] stated that retroperitoneal leiomyosarcomas are also largely asymptomatic and therefore grow to a large size before presentation, namely 50% are more than 15 cm in diameter.

In terms of contour, retroperitoneal sarcomas are lobulated, relatively smooth tumors [3,6,7,19], as in our case.

The liposarcoma is identified on CT images as large predominantly fat-attenuation mass, with thin septa and small soft-tissue nodules [3].

On the other hand, Leiomyosarcomas tend to be large with cystic degeneration [7], heterogeneous masses [3], with central low attenuation, typically a nonfatty mass without calcification [4,7]. Enhancement of solid portions of the tumor admixed with non-enhancing areas of degeneration, hemorrhage, and necrosis [3,24]. Although CT demonstrate no specific imaging features for retroperitoneal leiomyosarcoma it is important to recognize that leiomyosarcoma does not contain fatty elements [3].

Arakawa., *et al.* [7] presented also a case of leiomyosarcomas as large abdominal mass with homogeneous cystic component with thickened wall, without calcification or invasion to the adjacent organs.

Enhancement of a retroperitoneal leiomyosarcoma is often more marked at the periphery of the tumor [3,24,25].

In our case, the tumor was relatively well-defined mass with heterogeneous structure, predominantly pseudo-cystic, with high enhancement nodules with peripheral disposition. No fat densities or calcification were identified. Corresponding to these CT features, it is pertinent to conclude that the tumor resembles to retroperitoneal leiomyosarcoma.

Commonly, retroperitoneal sarcomas show no enlargement of lymph nodes around the mass and metastasis [7,19], as in the case we discus in this paper.

Surgery is the main treatment factor in the outcome of retroperitoneal leiomyosarcoma [4,15,24-26] and is the only treatment that has been shown to improve survival, with any role for chemotherapy or radiation still controversial [27,28].

Surgically complete excision of tumor is the most crucial aspect in treatment for retroperitoneal leiomyosarcoma in early stage and successful complete removal is the significant prognostic factor to predict local recurrence and metastasis [29]. Complete surgical resection with at least 3 cm margins is the treatment of choice but is rarely feasible due to invasion of adjacent structures by the tumor [4,9].

Erzen., *et al.* [15] pointed out that some tumors might need to be resected "en bloc" with adjacent structures in order to obtain free margins.

Resection margin involvement for liposarcoma also affects prognosis. Cases with involved resection margins are characterized by a tendency towards local recurrence [30].

In case of retroperitoneal leiomyosarcoma, most of studies have shown local recurrence within 3 years after excision of primary mass and approximately 80% to 87% of all local recurrences become evident within 2 years [19].

For liposarcoma, retroperitoneal origin is a poor prognostic factor [6]. This is because the retroperitoneal space allows the tumor to grow to a large size before the appearance of clinical signs and symptoms [31]. Therefore, the tumor is often diagnosed at the advanced stage, as it happened in the case presented in this paper.

Identification of a retroperitoneal mass at CT is an occasional, but frequently challenging, diagnostic scenario for the radiologist [32]. However, because of the recent development of various radiological modalities, including ultrasonography and CT, earlier detection of the tumor has become possible [7]. MRI can be a supplementary tool to determine staging of cancer [19].

But, in the end, pathological examination is the only method to distinguish leiomyosarcoma from liposarcoma, or other retroperitoneal sarcomas.

Conflict of Interest

There is no conflict of interest.

Bibliography

1. Yang J., *et al*. "The role of mesenchymal stem/progenitor cells in sarcoma: update and dispute". *Stem Cell Investigation* 1 (2014): 18.

2. Tobias J. "Cancer and its Management". 7th edition. Chichester, West Sussex, UK: John Wiley and Sons, Ltd (2015): 446.

3. Lev AD., *et al*. "Soft-Tissue Sarcomas of the Abdomen and Pelvis: Radiologic Pathologic Features, Part 1- Common Sarcomas: From the Radiologic Pathology Archives". *Radio Graphics* 37 (2017): 462-483.

4. Lin HC., *et al*. "Retroperitoneal Leiomyosarcoma: A Case Report and Review of the Literature". *Journal of American Society of Colon and Rectal Surgeons* (2008).

5. Fletcher CDM., *et al*. "WHO classification of tumours of soft tissue and bone". 4th edition. Lyon, France: IARC (2013): 10-11.

6. Oh SD., *et al*. "A Giant Retroperitoneal Liposarcoma Encasing the Entire Left Kidney and Adherent to Adjacent Structures: A Case Report". *Case Reports in Oncology* 9 (2016) :368-372.

7. Arakawa A., *et al*. "Radiological findings of retroperitoneal leiomyoma and leiomyosarcoma: report of two cases". *Computerized Medical Imaging and Graphics* 17.2 (1993): 125-131.

8. Catton CN., *et al*. "Outcome and prognosis in retroperitoneal soft tissue sarcoma". *Journal International Journal of Radiation Oncology* 29 (1994):1005-1010.

9. Todd CS., *et al*. "Retroperitoneal leiomyosarcoma: eight cases and a literature review". *Gynecologic Oncology* 59 (1995): 333-337.

10. Dalal KM., *et al*. "Subtype specific prognostic nomogram for patients with primary liposarcoma of the retroperitoneum, extremity, or trunk". *Annales Surgery* 244 (2006): 381-391.

11. Mack TM. "Sarcomas and other malignancies of soft tissue, retroperitoneum, peritoneum, pleura, heart, mediastinum, and spleen". *Cancer* 75 (1995): 211-244.

12. Kransdorf MJ. "Malignant soft-tissue tumors in a large referral population: distribution of diagnoses by age, sex, and location". *AJR American Journal of Roentgenology* 164 (1995): 129-134.

13. Lawrence W. "Operative management of soft tissue sarcomas: Impact of anatomic site". *Seminars in Surgical Oncology* 10 (1994): 340-346.

14. Heslin JM., *et al*. "Prognostic factors associated with long-term survival for retroperitoneal sarcoma: Implications for management". *Journal of Clinical Oncology* 15 (1997): 2832-2839.

15. Erzen D., *et al*. "Retroperitoneal sarcoma: 25 years of experience with aggressive surgical treatment at the institute of oncology Ljubljana". *Journal of Surgical Oncology* 91 (2005): 1-9.

16. Ki EY., *et al*. "A huge retroperitoneal liposarcoma: case report". *European Journal of Gynecological Oncology* 33 (2012): 318-320.

17. Shahaji C., *et al*. "Giant retroperitoneal liposarcoma: a case report". *Case Reports in Oncological Medicine* 4 (2012): 1-2.

18. Caizzone A., *et al*. "Giant retroperitoneal liposarcoma: case report and review of the literature". *International Journal of Surgery Case Reports* 9 (2015): 23-26.

19. Cho DH., *et al*. "Unusual presentation of retroperitoneal leiomyosarcoma mimicking an adnexal tumor with highly elevated serum CA-19-9". *Obstetrics and Gynecology Science* 57 (2014): 77-81.

20. Leao P., *et al*. "Giant recurrent retroperitoneal liposarcoma initially presenting as inguinal hernia: review of literature". *International Journal of Surgery Case Reports* 3 (2012): 103-106.

21. Goldblum JR., *et al*. "Liposarcoma" In: Goldblum JR, Folpe AL, Weiss SW, eds. Enzinger and Weiss's soft tissue tumors. 6th edition. Philadelphia, Pa: Saunders/Elsevier (2013): 484-523.

22. Lewis JJ., *et al*. "Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution". *Annals of Surgery* 228 (1998): 355-365.

23. Khan IN., *et al*. "A Retroperitoneal Leiomyosarcoma Presenting as an Adrenal Incidentaloma in a Subject on Warfarin". *Case Reports in Endocrinology* (2015): 4.

24. Hartman DS., *et al*. "Leiomyosarcoma of the retroperitoneum and inferior vena cava: radiologic-pathologic correlation". *Radio Graphics* 12 (1992): 1203-1220.

25. Stoeckle E., *et al*. "Prognostic factors in retroperitoneal sarcoma: a multivariate analysis of a series of 165 patients of the French Cancer Center Federation Sarcoma Group". *Cancer* 92 (2001): 359-368.

26. Bautista N., *et al*. "Retroperitoneal soft tissue sarcomas: prognosis and treatment of primary and recurrent disease". *The American Journal of Surgery* 66 (2000): 832-836.

27. Merlo M., *et al*. "Leiomyosarcoma of the inferior vena cava: a clinicopathologic review and report of four cases". *Minerva Chirurgical* 63 (2008): 209-221.

28. Alexander A., *et al*. "Leiomyosarcoma of the inferior vena cava: radical surgery and vascular reconstruction". *World Journal of Surgical Oncology* 7 (2009): 56.

29. Hueman MT., *et al*. "Management of retroperitoneal sarcomas". *Surgical Clinics of North America* 88 (2008): 583-957.

30. Kirane A., *et al*. "The importance of surgical margins in retroperitoneal sarcoma". *Journal of Surgical Oncology* 113 (2016): 270-276.

31. McCallum OJ., *et al*. "Retroperitoneal liposarcoma weighing over one hundred pounds with review of the literature". *Gynecologic Oncology* 103 (2006): 1152-1154.

32. Emily M., *et al*. "Can CT Features Differentiate Between Inferior Vena Cava Leiomyosarcomas and Primary Retroperitoneal Masses?". *AJR* 200 (2013): 205-209.

Volume 7 Issue 6 June 2020