Annals of Clinical and Medical Case Reports

Case Report ISSN 2639-8109 | Volume 9

Isolated Retroperitoneal Lynphangioleiomyomatosis: Robot-Assisted Surgical Treatment

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Received: 28 May 2022
Accepted: 07 Jun 2022
Published: 13 Jun 2022
J Short Name: ACMCR

Keywords:

Lymphangioleiomyomatosis; Retroperitoneal lymphangioleiomyomatosis; Robot-assisted laparoscopy

Citation:

Fernandes LA, Isolated Retroperitoneal Lynphangioleiomyomatosis: Robot-Assisted Surgical Treatment. Ann Clin Med Case Rep. 2022; V9(7): 1-3

1. Abstract

Lymphangioleiomyomatosis (LAM) is a rare disorder and its etiology is still unknown. There is an abnormal proliferation of smooth muscle cells, it is more frequent in young women, accompanied with pulmonar impairment in most of times. Its retroperitoneal apresentation is rare and rarely mentioned in literature. In this paper, we will describe the case of a 44-year-old female patient diagnosed with isolated retroperitoneal lymphangioleiomyomatosis and treated with a robot-assisted laparoscopic approach.

1.1. Objectives: To review the literature briefly and correlate it to the patient's condition, discussing pathological and therapeutic aspects.

1.2. Methods: Case report of a patient accompanied by members of the author's team. Information was obtained by reviewing the medical records about the case.

2. Case Report

44-year-old female patient without comorbidities. She presented to the service with the chief complaint of diffuse and nonspecific abdominal pain of undetermined origin.

Total abdominal US was requested, which showed the presence of a large mass of retroperitoneal origin on the left. For a better evaluation of the finding, a CT scan of the total abdomen was requested, which revealed a retroperitoneal tumor on the left, measuring 8 cm in the longest axis and the presence of enlarged coalescents lymph nodes in the interaortocaval and para-aortic space. The patient was also submitted to a chest CT, which showed no changes.

An MRI was then performed for better surgical planning, which showed a solid-cystic lesion of the retroperitoneum measuring 8.6 x 5.3 x 4.7 cm, with partially defined contours, multiloculated, predominantly cystical with a solid component of 3 cm. There was an apparent continuity between the lesion and the retroperitoneal lymphatic branches, branches that were slightly ectatic. The lesion had an intimate medium distal contact with the main renal vessels on the left, with no signs of their infiltration. Laterally, it maintained contact with the renal hilum and lower third of the left kidney without any signs of infiltration by this method. The diagnostic method concluded that the findings were indeterminate but allow the inclusion of isolated retroperitoneal lymphangioleimyomatosis as a differential diagnosis.

In a retrospective analysis of the service's image files, an MRI of the lumbar spine performed in July 2012 showed the presence of periaortic lymphadenopathy, with the largest ganglion measuring 1.2 cm, suggesting a benign insidious disease (Figure 1).

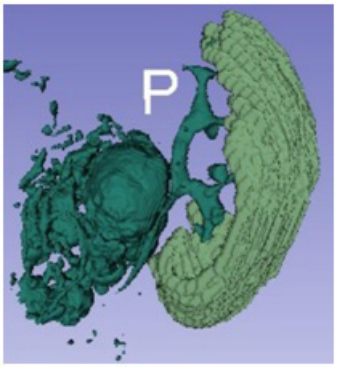

Figure 1:

The finding brought as the main diagnostic suspicion Lymphangioleiomyomatosis and the differential diagnosis of lymphoma could not be pushed away, thus, different therapeutic decisions were considered. In a discussion between a multidisciplinary team including a urology, oncologic surgery and oncologist team together with the patient and her family, two possibilities were exposed: Preoperative biopsy of the lesion, in which if the diagnosis was confirmed, it would proceed to guided treatment or opt for complete resection of the lesion with freezing biopsy performed in the same procedure, if the freezing biopsy suggested intraoperative lymphoma, the surgery would be interrupted.

Due to the low probability of lymphoma, with absence of suggestive symptoms such as night sweats, absence of lymphadenopathy in other regions and absence of fever, the surgical procedure was chosen.

The patient underwent a robot-assisted laparoscopic approach, using the Da Vinci Si – Intuitive Stratner platform, in lateral docking, using the 4 robotic arms and a 12 mm laparoscopic auxiliary.

After robotic docking, the main mass was easily visualized and a fragment of it was sent for a freezing biopsy, which revealed a 9.2 cm Fusocellular Neoplasm in its longest axis. Due to the absence of lymphoma suggestive findings, the proposal for resection of the mass and retroperitoneal lymph nodes was continued. During the surgery, it was possible to perform dissection of the main tumor mass, preserving the renal hilum and ureter, which enabled the preservation of the entire renal unit. The resection of the intra-aortocaval and paraortic lymph nodes was performed with the patient in lateral docking with no noteworthy technical difficulties. For the ligature of larger size lymphatic vessels, it was used polymer clips. There was a point of more adherence of the lymph nodes to the medial part of the inferior vena cava, which resulted in a 2 mm small lasceration, that was promptly sutured with a X point using Prolene 3.0.

The procedure lasted 120 minutes, with an estimated blood loss of 200 ml. We chose not to send the patient to a closed unit, which allowed hospital discharge within 24 hours after surgery (Figure 2).

The final histopathological analysis together with immunohistochemistry confirmed the diagnosis of isolated retroperitoneal lymphangioleiomyomatosis.

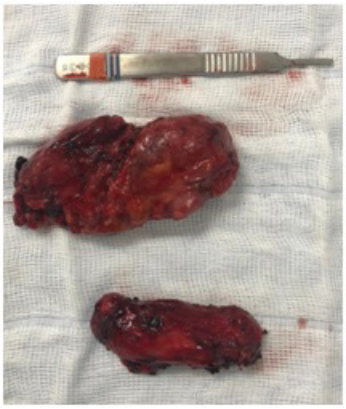

Figure 2:

3. Discussion

LAM is a rare disease that affects 1;1.000.000 people [1-3]. It is a multisystemic pathology that affects lungs, kidneys, retroperitoneal lymph nodes, liver, uterus and pancreas. The pulmonary form is the most common, characterized by the abnormal proliferation of smooth muscle cells, with destruction of the lung parenchyma [4]. Currently considered a low-grade neoplasm with variable metastatic potential [5]. It can also affect smooth muscle cells of lymphatic vessels [6, 7].

The pathophysiology of the disease is still unknown, however some theories relate the disease to be stimulated by some hormones, where some receptors sensitive to progesterone and estrogens found in LAM cells have been documented in the literature [8], associating the intimate relationship of impairment with greater frequency in women with reproductive age, however, it is not a finding found in all patients [9].

Literature reports of retroperitoneal LAM without pulmonary involvement are scarce due to its important rarity. In an analysis of 188 cases of LAM, only 3 patients had an isolated extrapulmonary form (2%) [10].

4. Conclusion

Lymphangioleiomyomatosis is a rare disease, especially in cases of isolated retroperitoneal presentation, as reported. To decide the best course of action, factors such as the size and location of the mass are considered. In this case, a robot-assisted laparoscopy was performed with good results, with the possibility of preserving adjacent structures, low blood loss and discharge after 24 hours.

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