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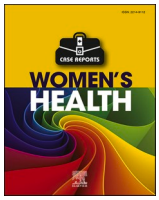

Retroperitoneal lymphangioma in a pregnant patient: A case report and literature review

Isabela Sánchez Mayorca [a,*], Mónica Viviana Ramírez Cifuentes [a], Diana Marcela Hoyos Guerrero [a], Roberto Gallo Roa [b], Santiago Vieira Serna [c], Rafael Leonardo Aragón Mendoza [d]

[a] Obstetrics and Gynecology resident physician, Hospital Universitario de La Samaritana, Bogotá, Colombia
[b] Obstetrician and gynecologist physician, Obstetrics and Gynecology Department, Hospital Universitario De La Samaritana, Bogotá, Colombia
[c] Obstetrician and gynecologist physician, gynecological oncology, Clínica Universitaria Colombia, Instituto Nacional De Cancerología, Bogotá, Colombia
[d] Obstetrician and gynecologist physician, maternal fetal medicine, Obstetrics and Gynecology Department, Hospital Universitario De La Samaritana, Bogotá, Colombia

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ABSTRACT

This case report describes the management of a woman diagnosed with a retroperitoneal cystic tumor during pregnancy. The 29-year-old patient presented at 29 weeks of pregnancy with abdominal pain. A retroperitoneal tumor measuring 224 × 156 × 235 mm was identified on ultrasound and magnetic resonance imaging. The patient underwent cesarean section delivery of a healthy neonate at 37 weeks. Uncomplicated laparoscopic surgery was performed during the postpartum period, resulting in a histologic diagnosis of a retroperitoneal lymphangioma. A review of articles published between 2003 and 2023 on the diagnosis, management, and prognosis of gestational lymphangiomas was conducted using the PubMed, SCOPUS and SpringerLink databases. Ten articles, including case reports of lymphangiomas diagnosed during pregnancy, were identified. The most frequent location was the gastrointestinal tract, with no cases reported in the retroperitoneal area. A good perinatal outcome was reported in the majority of cases. Lymphangiomas are rare benign tumors that are even more uncommon during pregnancy. Watchful waiting can offer a good obstetric and perinatal prognosis.

1. Introduction

Lymphangioma is a congenital benign tumor characterized by the benign focal proliferation of histologically well-differentiated lymphatic tissue, primarily consisting of dilated lymphatic channels lined by endothelium rich in connective tissue and smooth muscle cells, lymphocytes, fibroblasts, and collagen bundles [1,2]. There are three main subtypes, capillary, cavernous, and cystic, although they may all coexist within a single tumor [3]. They can occur in any part of the body, but most commonly in extra-abdominal locations such as the cervical, axillary, and inguinal regions. Intra-abdominal lymphangiomas are less common [2]. The diagnostic approach typically involves imaging studies such as ultrasound or computed tomography, with diagnosis confirmed by histopathology following surgical resection, which is considered the definitive treatment for this condition [1].

The presence of lymphangiomas during pregnancy is rare, with only a few cases reported in the literature. There appear to be no prior descriptions of this tumor in the retroperitoneal location during pregnancy [4]. These tumors pose a diagnostic and therapeutic challenge during pregnancy, as treatment options must consider both the mother and the fetus. Furthermore, there is limited evidence regarding fetal prognosis in these patients [5].

We report the case of a patient at 29 weeks of gestation with a retroperitoneal cystic tumor who underwent clinical observation prior to delayed surgical resection and histopathological confirmation of retroperitoneal lymphangioma. We discuss the maternal and fetal outcomes and review the literature on the diagnosis, treatment, and prognosis of this condition during pregnancy.

2. Case Presentation

A 29-year-old patient, at 29 weeks of her second pregnancy, presented to the emergency department with complaints of a first episode of pain occurring one month prior. She described the pain as a pressure

* Corresponding author at: carrera 8 # 0-29, 110411, Departamento de Ginecología y Obstetricia Hospital Universitario De La Samaritana, Bogotá, Colombia.
E-mail addresses: isanchez_94@hotmail.com (I. Sánchez Mayorca), monramirez@uniboyaca.edu.co (M.V. Ramírez Cifuentes).

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sensation in the left hypochondrium, rated 6/10 in intensity, with no accompanying symptoms. An incidental finding of an abdominal cystic tumor measuring 185 × 141 × 100 mm, with a volume of 2202 cm^3, was noted on obstetric ultrasound performed at 22 weeks. Regarding obstetric history, there was a 5-year interval since the previous pregnancy, which resulted in a term cesarean delivery due to fetal cephalopelvic disproportion.

On physical examination, the patient was alert, with a blood pressure of 112/68 mmHg, heart rate of 99 beats per minute, respiratory rate of 18 breaths per minute, and temperature of 36.5 degrees Celsius. Abdominal examination revealed tenderness on palpation over the left hypochondrium and flank, with no signs of peritoneal irritation. Singleton pregnancy with fetus in longitudinal cephalic position and fetal heart rate of 130 beats per minute were observed, with a uterine height of 30 cm. The gynecological exam was unremarkable. The patient was admitted with a diagnosis of abdominal pain secondary to a cystic tumor, and analgesic management with hyoscine and paracetamol was initiated, along with additional workup.

Obstetric ultrasound confirmed a fetus of appropriate weight for gestational age. Total abdominal ultrasound revealed an anechoic image in the left hemiabdomen, possibly retroperitoneal, with regular contours, measuring 224 × 156 × 235 mm in size and a volume of 4332 cm^3, showing no acoustic shadow or uptake on color Doppler (Fig. 1). Non-contrast magnetic resonance imaging (MRI) showed a large, unilocular, well-defined retroperitoneal cystic mass measuring approximately 230 × 140 × 210 mm, displacing the left kidney anteriorly, medially, and inferiorly, and the stomach to the right of the midline. A suspected diagnosis of retroperitoneal lymphangioma was reported (Fig. 2). Tumor markers revealed an elevated alpha-fetoprotein (AFP) level of 150.04 ng/mL, a negative carcinoembryonic antigen (CEA) level of 1.38 ng/mL, a negative CA 125 level of 10.50 U/mL, and a negative carbohydrate antigen 19–9 (CA 19–9) level of 8.56 U/mL.

A multidisciplinary meeting was held involving Maternal and Fetal Medicine, Gynecology, General Surgery, Urology, Neonatology, Social Work, and Psychology services. Due to suspicion of a benign retroperitoneal cystic tumor, no percutaneous biopsies were performed. Symptomatic management was initiated, with clinical follow-up based on serial fetal testing and planned term delivery. Definitive surgical resection was scheduled postpartum.

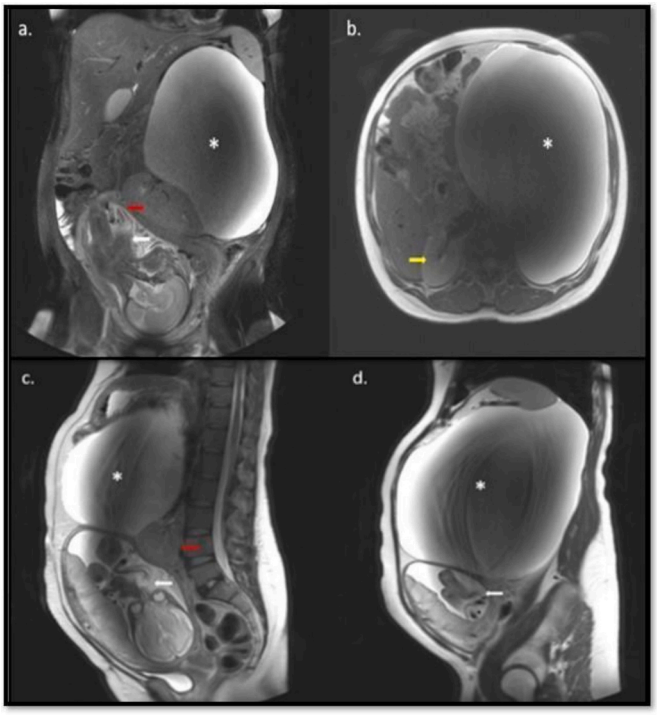

Fig. 2. Magnetic resonance imaging of the retroperitoneal lymphangioma. T2 sequence: large retroperitoneal unilocular cystic mass with hyperintense content. a. Coronal section; b. Axial section; c. Mid-sagittal section; d. Left parasagittal section. *Retroperitoneal lymphangioma. Red arrow: left kidney with anterior, medial and inferior displacement. Yellow arrow: right kidney. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

The patient underwent cesarean section at 37 weeks, delivering a healthy neonate weighing 2965 g and measuring 48 cm, with APGAR scores of 7-7-8 at 1, 5, and 10 min, respectively. Following an

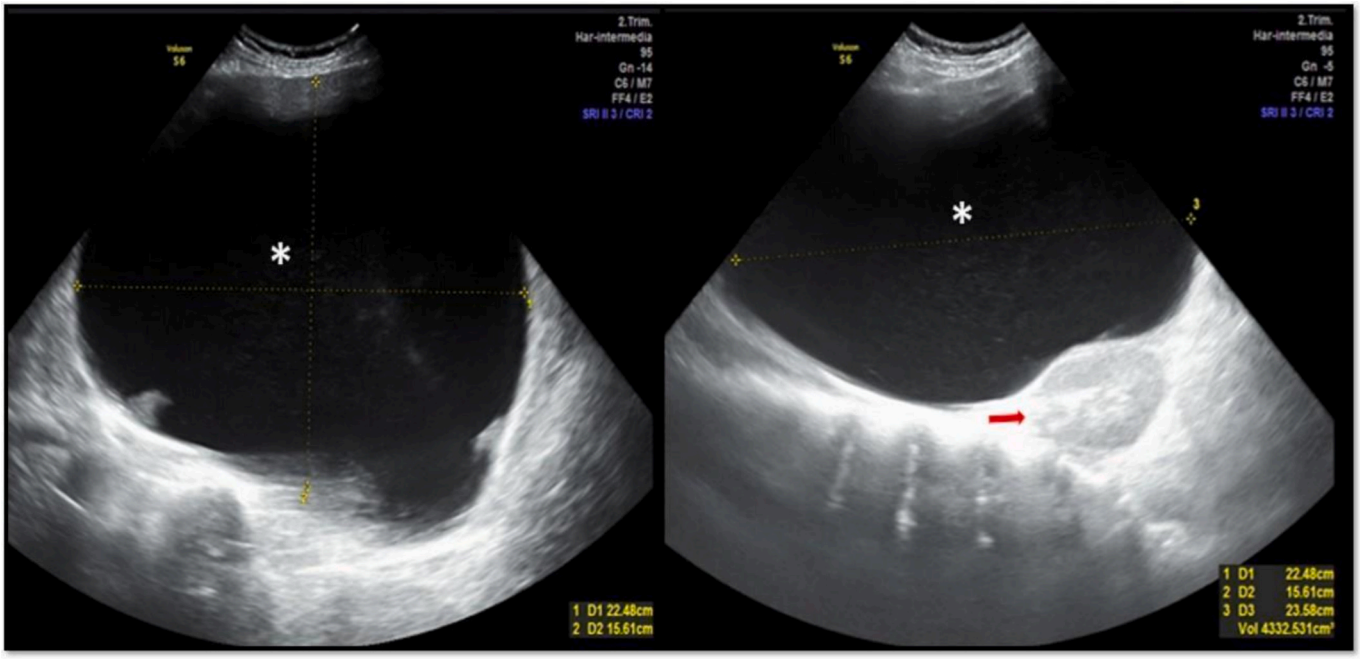

Fig. 1. Ultrasound image of the retroperitoneal lymphangioma: anechoic retroperitoneal tumor of regular contours, 224 × 156 × 235 mm in size, with no uptake on Doppler ultrasound. Arrow: left kidney.

uncomplicated surgical procedure, the patient's postoperative recovery progressed well, and she was discharged home after two days.

Four months postpartum, the patient presented with persistent abdominal pain and underwent laparoscopic appendectomy and resection of the left retroperitoneal cyst. Intraoperatively, a large, thin-walled lymphangioma measuring 30 × 30 cm and containing 6000 cc of yellowish fluid was found, originating from the left inframesocolic space. The mass caused displacement of surrounding structures, including cephalad displacement of the stomach and spleen, right displacement of the small bowel loops, and anterior and inferior displacement of the kidney. The patient recovered well postoperatively and was discharged on the third day. Histopathological analysis confirmed the presence of a retroperitoneal cyst with an endothelial component positive for CD34, CD31, and D240, and negative for calretinin, with AML and desmin positive vascular walls, providing a definitive diagnosis of lymphangioma. Additionally, acute appendicitis with transmural liquefaction necrosis and acute periappendicitis were noted in the histopathology report.

3. Discussion

A search was conducted in the PubMed, SCOPUS and SpringerLink databases, taking into account articles published between January 2003 and December 2023, using the Mesh terms "Lymphangioma," "Cystic Lymphangioma," "Abdominal Cystic Lymphangioma," and "Pregnancy." Articles published in English and Spanish were included, while cases where lymphangioma was diagnosed and treated before pregnancy or diagnosed during the postpartum period were excluded. Various variables were assessed, including tumor location, trimester of diagnosis, symptoms, diagnostic methods used during pregnancy, route of delivery, prognosis during pregnancy, and treatment modalities.

The search identified 15 articles concerning pregnant women with lymphangiomas [1–4,6–16]. Five articles were excluded from the analysis: four due to postpartum diagnosis [3,6–8] and one due to pregestational diagnosis [9]. This resulted in a total of 10 articles published in English or Spanish that discussed lymphangiomas diagnosed during pregnancy, including topics such as tumor location, trimester of diagnosis, symptoms, diagnostic methods, route of delivery, prognosis during pregnancy, and treatment modalities (Table 1).

Lymphangiomas diagnosed during pregnancy were documented in both abdominal [2,4,10,14,15,16] and extra-abdominal [1,11–13] locations, most commonly in the gastrointestinal tract (Table 2). Notably, none of the reported gestational lymphangiomas were located retroperitoneally. Diagnosis occurred in the second trimester in five cases

Table 2
Lymphangioma locations during pregnancy.

Abdominal	Extra abdominal
Mesentery [15,16]	Nasopharynx [12]
Lesser omentum [10]	Lung [1]
Pancreas [14]	Axilla [13]
Uterus [2]	Breast [11]
Ovaries, bilateral [4]	

Table 1
Reported cases of gestational lymphangiomas.

Author	Location, size	Week at the time of diagnosis	Symptom	Diagnostic method	Delivery, weeks of gestation	Complications during pregnancy	Treatment
Dhillon, 2014 [1]	Lung, NR	Second trimester, week not reported	Hemoptysis	CT scan	NR	Hemoptysis	Lobectomy in the postpartum period
Boumans, 2012 [14]	Pancreas, 18.5 × 16.8 cm	37 weeks	Abdominal pain/ Hemorrhagic shock	Intraoperative CT scan	Cesarean section at 37 weeks, NB death	Splenic artery rupture with hypovolemic shock and unfavorable fetal status, with perinatal death	Urgent laparomoty resection of the pancreatic tumor in the immediate postpartum period
Shore, 2014 [2]	Uterus, 8.6 × 6.5 × 2.9 cm	15 weeks	Asymptomatic	Ultrasound/MRI	Term vaginal deliver, healthy NB	No complications	Hysterectomy in postpartum month 8
Ozdemir, 2017 [10]	Lesser omentum, 148 × 47 mm	26 weeks	Abdominal pain	Ultrasound/MRI/ prenatal biopsy	Cesarean section at 39 weeks, healthy NB	No complications	Surgical tumor resection through laparotomy during the same cesarean procedure
Galili, 2019 [12]	Nasopharynx, 4.5 × 1.5 × 0.8 cm	32 weeks	Dysphagia and dyspnea	MRI	Cesarean section at 41 weeks	No complications	Tumor resection during pregnancy at 32 weeks
Choudhary, 2021 [4]	Ovaries	38 weeks	Asymptomatic / Ascites	Intraoperative / ovarian biopsy	Cesarean section at 38 weeks	Tension ascites during the postpartum period	Bilateral oophorectomy on postpartum week 3
Quack, 2005 [13]	Axilla, 4 cm	26 weeks	Pain and inflammation	Ultrasound/MRI	Vacuum vaginal delivery at 39 weeks	No complications	Serial drainage during pregnancy followed by total resection two weeks into the postpartum period due to overinfection
Torashima, 2004 [15]	Mesentery, 15 × 10 cm	12 weeks	Abdominal pain	MRI	Vaginal at 40 weeks	Clinical acute abdomen at 25 weeks	Resection of the tumor and of 30 cm of the ileum through laparotomy at 25 weeks
Konstantinidis, 2005 [16]	Mesentery, NR	15 weeks	Abdominal pain	Intraoperative	NR	Clinical acute abdomen at 15 weeks	Resection of the tumor and bowel portion through laparotomy at 15 weeks
De Guerké, 2005 [11]	Breast, 3 cm	25 weeks	Mass sensation/ bulkiness	Mammography/ Cystography/ Ultrasound/Prenatal biopsy	NR	No complications	Spontaneous resolution two weeks into the postpartum period

MRI: magnetic resonance imaging. NB: newborn. NR: not reported.

[1,10,11,13,16], in the third trimester in three cases [4,12,14], and in the first trimester in only two cases [2,15]. De novo diagnosis of lymphangioma during pregnancy could be attributed to symptoms associated with larger lymphangiomas during gestation [10,12,13]. In our case, tumor growth was progressive, reaching over 18 cm in diameter during the second trimester, increasing to 25 cm in the third trimester, and reaching 30 cm by the time of surgical resection.

Symptoms varied depending on the anatomical location and size of the lymphangiomas. Abdominal pain was the most common presentation in abdominal lymphangiomas [10,13,14,15,16], while mass effect was predominant in extra-abdominal lymphangiomas [11,12]. Two patients were asymptomatic, with uterine and ovarian lymphangiomas found incidentally [2,4].

Multiple imaging methods were employed, including ultrasound and MRI, with MRI being preferred for studying retroperitoneal tumors in pregnancy due to its improved ability to characterize lesions and assess surgical resection options. Although percutaneous biopsy is standard in retroperitoneal tumors, it carries a risk of capsule rupture and associated complications such as tension ascites, as reported in one case [4].

Definitive diagnosis typically requires histopathology after tumor resection [4,10,11,13]. The diagnostic yield of tumor markers in this type of lesion remains unknown, with tumor markers requested in only one case, yielding normal results [10]. In our case, only alpha-fetoprotein was elevated, though this marker is nonspecific for lymphangioma and can be elevated in pregnancy [17].

Treatment should be individualized based on symptoms and tumor location. Expectant management during pregnancy, with close clinical and imaging assessment and fetal wellbeing tests, is generally recommended due to the benign nature of the tumor. Surgical resection, the preferred treatment, is typically deferred until after childbirth [5]. However, surgical management during pregnancy may be necessary in cases of mass effect, rupture of adjacent structures, or uncontrolled abdominal pain. Our literature review found three cases where surgery was performed during pregnancy, including one due to airway compression [12] and two due to acute abdomen [15,16]. Additionally, one case involved surgical treatment during cesarean section [10], while surgical resection during the postpartum period was reported in five cases [1,2,4,13,14]. Notably, one case of breast lymphangioma resolved spontaneously postpartum [11].

Prenatal complications included hemoptysis [1], splenic artery rupture with hemodynamic shock [14], and the need for multiple drainage procedures due to discomfort and pain related to the mass effect [13]. Urgent laparotomy for tumor and related bowel resection was required in two cases [15,16]. There were no cases of maternal mortality, and no recurrences were reported after definitive treatment. Cesarean section and vaginal delivery were both reported, with a total of nine healthy newborns. Adequate obstetric outcomes were achieved in the majority of cases, with no reported complications or unfavorable outcomes, except for one case of perinatal mortality due to hemorrhagic shock secondary to splenic artery rupture from pancreatic lymphangioma [14]. Although a specific delivery route based on lymphangioma diagnosis was not documented in reported cases, given the potential mass effect of abdominal tumors, attention to those below fetal presentation is crucial, as they may cause dystocia requiring cesarean section [5].

4. Conclusion

Lymphangiomas are exceptionally rare benign tumors, and their occurrence during pregnancy is even more uncommon. They can manifest in various locations, with the abdomen being the most prevalent site. The diagnostic approach during pregnancy relies on non-invasive imaging studies, while treatment typically involves surgical resection, with definitive diagnosis based on histopathological examination of the specimen. During pregnancy, management is personalized based on patient symptoms and tumor location.

Due to their benign nature, these tumors can be monitored with vigilant observation until definitive treatment after childbirth. However, it is crucial to remain vigilant for potential tumor growth and complications such as acute abdomen, bleeding, and compromise of adjacent structures, which may necessitate immediate intervention, thereby increasing maternal morbidity. Therefore, regular clinical follow-up and fetal wellbeing assessments are imperative. Further research is necessary to enhance our understanding of this condition and to refine therapeutic approaches to lymphangiomas in the context of pregnancy.

Contributors

Isabela Sánchez Mayorca contributed to patient care, conception of the case report, acquiring and interpreting the data, undertaking the literature review and revising the article critically for important intellectual content.

Monica Viviana Ramírez Cifuentes contributed to acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Diana Marcela Hoyos Guerrero contributed to patient care, acquiring and interpreting the data from de clinical history, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Roberto Gallo Roa contributed to acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Santiago Vieira-Serna contributed to patient care, acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Rafael Leonardo Aragón-Mendoza contributed to patient care, conception of the case report, acquiring and interpreting the data, undertaking the literature review and revising the article critically for important intellectual content.

All authors approved the final submitted manuscript.

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Patient consent

Obtained.

Ethical approval

Ethical considerations were upheld throughout the study. The publication of the case was authorized by the institution's research and ethics committee. Measures were taken to ensure the confidentiality of information and patient anonymity. Photographs included in the report were taken by the authors and used with permission.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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