

# A Case of Vulvar Cavernous Lymphangioma

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## Abstract

*Cystic (cavernous) lymphangioma of the vulva is a benign tumor of lymphatic vessels with only 10 cases previously reported in the medical literature worldwide. The following is a case of bilateral vulvar cavernous lymphangiomas in a 23-year-old woman with bilateral soft tissue masses of the labia majora. Prior unsuccessful management included aspiration, incision, and drainage. Surgical marsupialization of both masses led to resolution and the subsequent histologic diagnosis of cavernous lymphangioma. Although rare, cavernous lymphangioma should be included in the differential diagnosis of soft tissue masses of the distal labia. Cavernous lymphangioma can mimic other more common vulvar soft tissue masses such as Bartholin's cyst.*

## Keywords

cavernous lymphangioma, cystic lymphangioma

## Introduction

Vulvar cavernous lymphangioma is a rare benign disorder with only 10 cases previously reported worldwide including a single case of bilateral occurrence.<sup>1</sup> This report is a case of a young woman with suspected recurrent and bilateral Bartholin's gland cysts who underwent marsupialization after multiple outpatient treatments. Although the gross appearance and the contents of the cysts were consistent with Bartholin's cysts, the histologic evaluation revealed cavernous lymphangioma. Lymphangiomas are uncommon soft tissue masses that can further be divided into 2 subtypes, capillary lymphangioma and cavernous lymphangioma (CL). Capillary lymphangioma, also known as lymphangioma circumscriptum, is the most common cutaneous form of lymphangioma and appear superficially, located in the upper dermis and epidermis. In contrast, cavernous lymphangioma is an ill-defined, poorly circumscribed subcutaneous swelling without epidermal changes because the dilated lymphatics are located within the deep reticular dermis and subcutaneous fat.<sup>2</sup> CL can be present at birth or develop in early childhood. CL is typically found in the neck or axilla of children. The mass is made up of extremely dilated lymphatic spaces lined by endothelial cells and separated by connective tissue stroma containing lymphoid aggregates.<sup>3</sup>

## Case

A 23-year-old woman, gravida zero was presented to the gynecology clinic for evaluation of recurrent bilateral masses of the labia majora. Two years prior, she had undergone incision and drainage of a presumed left-sided Bartholin's gland cyst. She presented again to gynecology clinic a year later with

complaint of bilateral labial swelling which was treated conservatively with sitz baths, ibuprofen, and acetaminophen given the small size of the masses. Several days later, she presented to the emergency department for worsening pain, and incision and drainage of the left labium majus were performed. A large amount of purulent drainage was noted, and a Word catheter was placed. A culture was obtained at that time which resulted as *Streptococcus agalactiae*. *Streptococcus agalactiae* typically is an asymptomatic colonizer of the vagina found in up to 36 % of women, and thus she was not treated with antimicrobial therapy.<sup>4</sup> Two weeks later she presented to her primary care physician who removed the Word catheter. A 4-centimeter right-sided labial mass was noted at that time which was aspirated with a needle. The fluid was sent for culture that was subsequently found to have no growth.

Approximately 7 months after her initial presentation the patient returned to her primary care physician with complaint of recurrent bilateral labial swelling. She was referred back to gynecology. The gynecologist noted bilateral, symmetrical appearing soft tissue masses of the distal labia majora that were each approximately 3 centimeters in diameter. The working diagnosis was bilateral Bartholin's gland cysts. Marsupialization was recommended and performed 10 days later. The left labium majus was scored vertically with a scalpel and the cyst was opened. Clear yellow mucinous fluid was drained and a portion of the cyst wall was sent for pathology. Marsupialization was then performed. The right labium was opened, drained and marsupialization was completed in a similar fashion. However, a sample was not sent for pathology as the gross findings were identical to the left side. The surgery was uncomplicated and the patient was discharged home on the day of surgery. She was then subsequently readmitted the same evening with bilateral labial hematomas. She was managed conservatively with observation, serial laboratory assessments and analgesics for pain control. The patient was discharged in stable condition after 4 days in the hospital. Her subsequent course was unremarkable with resolution of the bilateral labial masses without recurrence to date.

The histopathology of the left labial cyst sample subsequently showed CL (Figure 1). The histologic differential diagnosis of a cyst lined by flat cells included lymphangioma, hemangioma, and hydrocele (of the canal of Nuck). The diagnosis was confirmed by immunohistochemistry. The lymphatic endothelium marker D2-40 was positive (Figure 2), and blood vessel endothelium marker (FVIII-related antigen) and mesothelium marker (CK5/6) were negative.

Table 1. Summary of Case Reports on Vulvar Cavernous Lymphangioma			
Authors	Year	Site	Age at Treatment (yr.)
Krebs, et al, <sup>5</sup>	1984	Right labium majus	31
Brown, et al, <sup>6</sup>	1989	Right labium majus	23
Gupta, et al, <sup>7</sup>	1998	Right labium majus	17
Forsnes <sup>8</sup>	2002	Right labium majus	19
Bagga, et al, <sup>9</sup>	2004	Left labium majus	17
Noël, et al, <sup>10</sup>	2007	Left labium majus Left labium majus	28 19
Watanabe, et al, <sup>11</sup>	2010	Bilateral labia minora	15
Amaranathan, et al, <sup>12</sup>	2013	Right labium majus	27
Yoon, et al, <sup>1</sup>	2015	Left labium majus	19

## Discussion

Vulvar lymphangiomas are classified as either lymphangioma circumscriptum or cavernous lymphangioma (known as cystic lymphangioma) and are extremely rare. Ten cases of vulvar cavernous lymphangioma had been previously described worldwide including one case of bilateral involvement (Table 1). Of the 10 reported cases, they were all found in reproductive age women ranging from 15 to 31 years old. The cases were evenly divided between lesions of the left and right vulva and all but one occurred in the labia majora. All were treated with uncomplicated local excision. Cavernous lymphangiomas consist of dilated spaces that are lined by lymphatic endothelium. The tumors tend to have indistinct borders making complete resection difficult.<sup>2</sup> In our case, indistinct borders and incomplete resection may have contributed to her return to the hospital and subsequent readmission for bilateral hematomas several hours after the marsupialization. This patient's left cyst was biopsied and both cysts were marsupialized without difficulty or excessive bleeding. It was unexpected that she returned the evening of her surgery with bilateral hematomas. The patient had a prior incision and drainage of the left labial mass with purulent drainage noted. It appeared consistent with a recurrent Bartholin's abscess. Although cavernous lymphangiomas are rarer than Bartholin cyst, they are likely underdiagnosed as Bartholin's cyst. Wall biopsies are not often evaluated histologically. Vulvar cavernous lymphangioma should be a consideration whenever surgical management of a suspected recurrent Bartholin's cyst is undertaken. Although bilateral presentation was not confirmed with histologic biopsy the identical presentation suggest that this case may be the second of bilateral occurrence. Further the cases highlighted the need to expand the differential diagnosis of vulvar masses, especially with recurrence when conventional therapy does not achieve resolution.

## Conflict of Interest

None of the authors identify any conflict of interest.

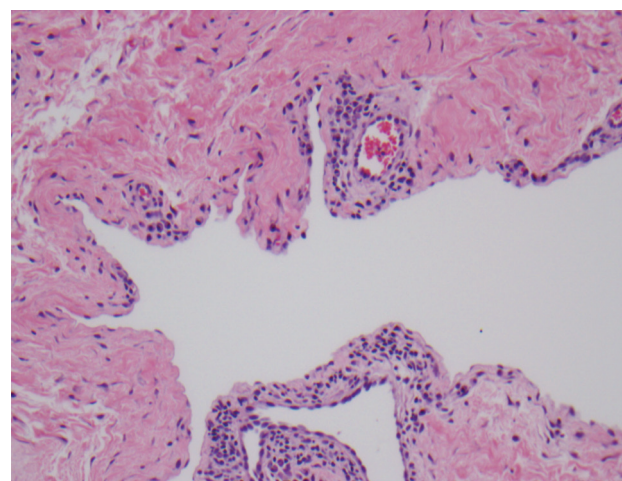


Figure 1. Cyst Lined by Flat Cells. Focal Adjacent Lymphoplasmacytic Inflammation. (Hematoxylin & Eosin Stain).

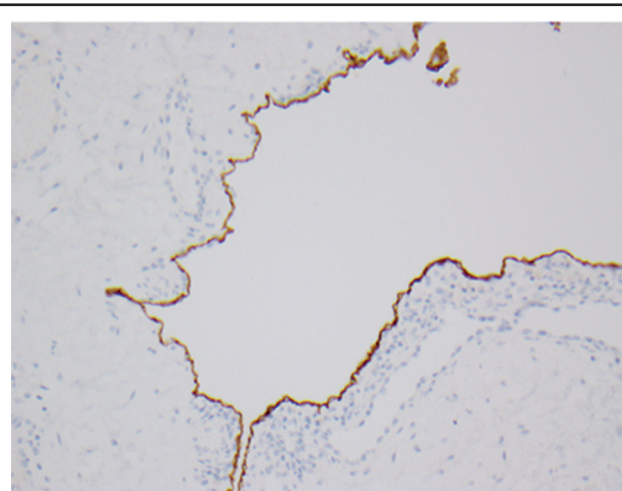


Figure 2. Cyst Lining Cells Reactive for D2-40, Consistent with Lymphatic Endothelium. The Cells are Negative for Vascular Endothelium Marker Factor VIII and Mesothelial Marker CK5/6 (Not Shown) (Immunohistochemistry Staining).

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