Cavernous Lymphangiomas Involving Bilateral Labia Minora

Takashi Watanabe, MD, Shigeki Matsubara, MD, Takehiko Yamaguchi, MD, and Yuji Yamanaka, MD

BACKGROUND: The vulva is an extremely rare site for lymphangioma, a benign tumor of the lymphatic vessels. A small number of case reports have described cavernous lymphangioma involving the unilateral labium majus of young women. We report a case of cavernous lymphangiomas involving the bilateral labia minora.

CASE: A 14-year-old girl presented with bilateral tumors of the labia minora, which were removed surgically and histologically diagnosed as cavernous lymphangioma. Positive nuclear staining for estrogen and progesterone receptors was demonstrated in the stromal fibroblasts immunohistochemically.

CONCLUSION: It is suggested that an increase in estrogen and progesterone levels may have led to, or at least affected, the development of these tumors in this patient. Excision followed by cosmetic repair may be the optimal treatment.

(Obstet Gynecol 2010;116:510–2)

Cavernous lymphangioma arising from the vulva of young women is a rare disorder; only a small number of cases have been reported in which tumors arose from the unilateral labium majus.1–6 We describe a patient with cavernous lymphangiomas involving the bilateral labia minora. The tumors were excised followed by successful reconstruction. Cavernous lymphangioma should be considered in tumors of the external genitalia of young women.

CASE

A 14-year-old girl was referred to us because of vulvar tumors. She had no history of trauma, surgery, or infection at the site. Her family history was unremarkable. She had achieved menarche at 12 years 3 months, had a regular cycle, and had not experienced intercourse. Just after achieving menarche at 12 years 3 months, had a regular cycle, and had not experienced intercourse. Just after

mass gradually enlarged and caused friction during walking and bicycle-riding, leading her to visit our hospital at 14 years, 11 months old. Physical examination revealed soft, banana-sized masses involving the bilateral labia minora (Fig. 1A–B). Magnetic resonance imaging and abdominal ultrasonography revealed no abnormal findings of the uterus, bilateral ovaries, or other pelvic organs. Serum levels of luteinizing hormone, follicular stimulating hormone, and estradiol were within normal ranges for her age: 1.2 milliunits/mL, 2.8 milliunits/mL, and 53.3 pg/mL, respectively.

Under general anesthesia, we first incised the lateral side of the right tumor and found that the tumor was not encapsulated and difficult to shell out. As shown in Figure 1A–B, the margin between the normal labia and tumors was not clear, so we resected the skin overlying the tumor surface together with the tumors. Thus, parts of the labia were transected. The incision lines were set for the convenience of cosmetic repair. Bilateral tumors measuring 8.0×3.0×3.0 cm (left side) and 9.5×3.5×3.5 cm (right side) were resected. The deeper part of the tumor, or the tumor floor, could be removed easily from the underlying tissue, and, thus, we believe that there was no remaining tumor tissue. Tumor vessel supply was derived mainly from the bottom of the tumor, but the blood supply was poor. As shown in Figure 1C, we removed redundant skin of the labia, and the skin incision site was sutured with monofilament threads. Normal-shaped labia were constructed. The labia majora were not touched during surgery, indicating that the labia majora were free from the tumors. Thus, we confirmed that the tumors originated from the bilateral labia minora. Total blood loss during the procedure was 30 mL. The postoperative course was unremarkable. Examination 6 months later showed normal-shaped labia and no tumor recurrence.

An ill-defined tumor in the dermis consisted of numerous dilated lymphatic vessels with a paucicellular fibrous background (Fig. 2). The endothelial cells were flattened and nuclei were bland. An immunohistochemical study using paraffin-embedded sections with the avidin–biotin complex method revealed a positive reaction for a marker for lymphatic vessel endothelial cells, D2-40 (monoclonal, dilution: 1:5; Nichirei Co, Tokyo, Japan) and also a marker for vascular endothelial cells, CD31 (monoclonal, dilution: 1:100; DAKO Cytomation A/S, Glostrup, Denmark), in the tumor endothelial cells. Also, positive nuclear staining for estrogen receptor (monoclonal, dilution: 1:100; DAKO Cytomation A/S) and progesterone receptor (monoclonal, dilution: 1:200; DAKO Cytomation A/S) was demonstrated in the stromal fibroblasts. The findings were consistent with those of cavernous lymphangioma.

COMMENT

Cavernous lymphangioma, although extremely rare, can involve the vulva. We performed a literature search using PubMed and Web of Knowledge with
unlimited dates using the index words “lymphangioma*” AND “vulva*” and “lymphangioma*” AND “labi*” and restricting the search to English language publications. We also hand-searched relevant articles from the reference lists of the retrieved articles. We retrieved seven reports describing patients with cavernous lymphangiomas arising from the vulva1–6 summarized in Table 1 together with the present case. As for the tumor location, ie, labium minus compared with majus, or bilateral compared with unilateral, although some reports presented photographs,1–3 others only described the location in the article.4–6 Even in the former cases, it is difficult to confirm the exact location from photographs; thus, we used the description used in each article for the location information, producing Table 1. All seven cases reported had tumors involving the unilateral labium majus.1–6 Thus, within our literature search, this is the first reported case of cavernous lymphangioma arising from the bilateral labia minora.

In six of the eight cases (75%), tumors were first noticed around or just after puberty, and the age at treatment was a median of 19 years (range, 15–31 years). Thus, data indicated that most cavernous lymphangiomas of the vulva arose around or soon after menarche and were successfully treated in the teens and 20s without recurrence. No reports described the clinical course during pregnancy. At present, we have found no features characteristic of “bilateral” and “labia minora” tumors compared with reported “unilateral” and “labium majus” ones. Although the significance of the unique tumor location of the present case is unclear, we can at least suggest that bilaterality and development from the labia minora may never preclude the possibility of cavernous lymphangioma.

Although this is a speculation, estrogen, progesterone, or both may have been associated with the

Fig. 1. Gross appearance of the vulva before (A–B) and after (C) surgery. A. Two banana-like tumors are observed in the external genitalia. The origin of these tumors is unclear from this direction. B. After pushing the tumors laterally, it became evident that the tumors originated from the labia—the left one from the left labium and the right one from the right labium. Whether the tumors originate from the labia majora or minora is indiscernible in this photograph. C. After surgery. The tumors were resected, and the bilateral labia minora were well-constructed.

Fig. 2. Microscopic tumor findings. Numerous dilated lymphatic vessels are seen throughout the dermis. The endothelial cells are flattened and bland. The intervening stroma is paucicellular and fibrous (hematoxylin-eosin, original magnification ×100).

occurrence of the tumor in the present patient for the following two reasons. First, the tumor arose after menarche, when both hormone levels rapidly increase. As cited, 75% of cases reported also showed the same course. Second, these tumors had estrogen and progesterone receptors. Pulmonary lymphangiomyomatosis, which was also reported to show estrogen and progesterone receptors, occurs exclusively in women, mostly in those of reproductive age, and exacerbates during pregnancy as a result of an increase in both hormone levels. A case of pulmonary lymphangiomyomatosis in a 14-year-old girl associated with bilateral pneumothorax was reported. Although these two diseases belong to different clinical entities, both have estrogen and progesterone receptors and show the abnormal proliferation of lesions. Thus, it is reasonable to assume that increases in estrogen and progesterone levels may have led to, or at least affected, the development of this tumor in the present patient. Clinical course observation during pregnancy of cavernous lymphangioma may be useful to test our supposition.

Examination 6 months later showed no recurrence in this patient. Excision followed by cosmetic repair led to a favorable result. However, the prognosis of lymphangioma of the bilateral labia minora is uncertain. Close follow-up is needed.

**REFERENCES**


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**Table 1. Clinical Features of Cavernous Lymphangioma of the Vulva**

<table>
<thead>
<tr>
<th>Study</th>
<th>Age at Onset (y)</th>
<th>Site</th>
<th>Age at Treatment (y)</th>
<th>Size (cm)</th>
<th>Recurrence, Follow-up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Krebs et al(^1)</td>
<td>30</td>
<td>Right labium majus</td>
<td>31</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Brown and Stenchever (^2)</td>
<td>SP</td>
<td>Right labium majus</td>
<td>23</td>
<td>4.0x1.5x3.2</td>
<td>ND</td>
</tr>
<tr>
<td>Gupta et al(^3)</td>
<td>ND</td>
<td>Right labium majus</td>
<td>17</td>
<td>10</td>
<td>ND</td>
</tr>
<tr>
<td>Forsnes(^4)</td>
<td>9</td>
<td>Right labium majus</td>
<td>19</td>
<td>8x4x2</td>
<td>(–), 2 y</td>
</tr>
<tr>
<td>Bagga et al(^5)</td>
<td>SP</td>
<td>Left labium majus</td>
<td>17</td>
<td>15</td>
<td>ND</td>
</tr>
<tr>
<td>Noël et al(^6)</td>
<td>27</td>
<td>Left labium majus</td>
<td>28</td>
<td>10</td>
<td>(–), 9 mo</td>
</tr>
<tr>
<td>Present case</td>
<td>13</td>
<td>Bilateral labia minora</td>
<td>15</td>
<td>8.0x3.0x3.0</td>
<td>(–), 6 mo</td>
</tr>
</tbody>
</table>

SP, since puberty; ND, not described. All were treated with local excision.