

## Childhood Asymmetry Labium Majus Enlargement

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Childhood asymmetry labium majus enlargement (CALME) is a distinctive clinicopathologic entity of pre- and early puberty first described in 2005. It is defined as an expansion of normal soft tissues of the vulva. Although CALME is not a rare lesion, it has been called lipoma, fibroma, hamartoma, and fibrous hyperplasia. CALME is not a true neoplasm and is a physiologic growth in response to hormone. It may tend to resolve spontaneously and recur after surgical resection. We report four cases of CALME with a review of the literature. To the best of the knowledge, this is the first Korean report.

**Key Words:** Child; Vulva; Hypertrophy; Enlargement; Puberty

Childhood asymmetry labium majus enlargement (CALME) is a distinctive clinicopathologic entity of pre- and early puberty first described by Vargas *et al.*<sup>1</sup> in 2005. Although CALME is not a rare lesion, it has been diagnosed for lipoma, fibroma, hamartoma, and fibrous hyperplasia according to the proportion of the histologic components. CALME is a site- and age-specific non-neoplastic physiologic growth response to hormone. Recognition of this distinctive entity is important to avoid surgical excision.

### CASE REPORT

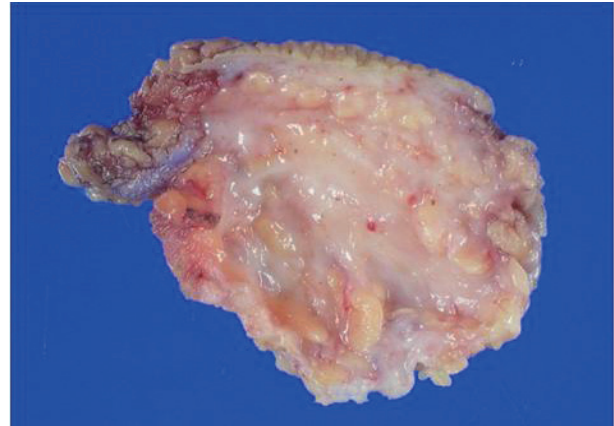
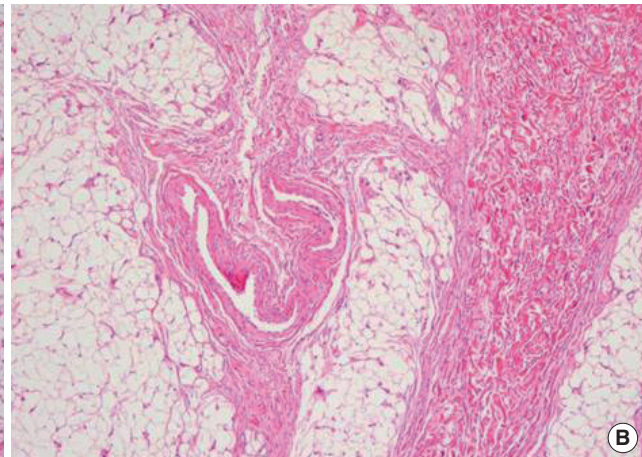
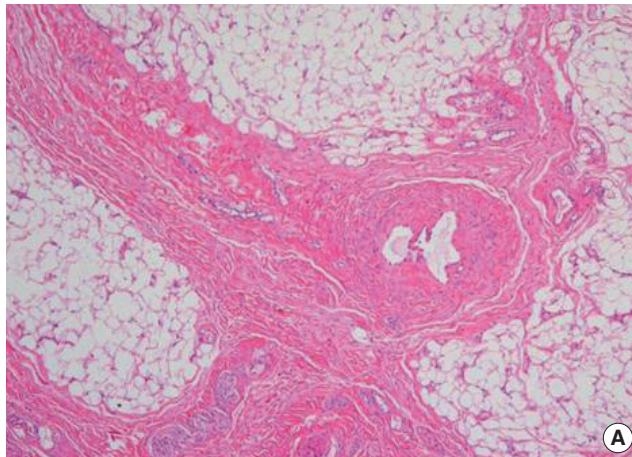
Clinical and pathologic features were reexamined for all patients under 13-year-old-age with vulva lesion from January 1, 2003 to January 1, 2009. Among the cases, three CALME cases previously diagnosed as lipoma or fibrolipoma in 2003, 2004, and 2007 were identified. One additional lesion was diagnosed as CALME in July, 2009. The clinical features of the four patients are summarized in Table 1. The age at presentation ranged from 5 to 8 years (median, 7 years). All the patients were female and were admitted complaining of bulging on labium majus or vulva enlargement (Fig. 1). Duration was uncertain. In all cases, preoperative ultrasonography showed a fat containing mass with an indistinct margin. Cases 2 and 3 were diagnosed lipoma on ultrasonography. Excision was performed. Gros-

sly, the specimens measured 3 to 5 cm in size (median, 4.4 cm) and were poorly margined, with borders that were difficult to discern. Sections obtained from cases 1 and 4 showed fibro-fatty tissue without hemorrhage and necrosis (Fig. 2). Consistency was soft to rubbery. Case 2 was grayish white homogeneous fibrotic tissue. Case 3 was composed of predominantly fat tissue with indistinct margin. On microscopic examination, the lesions except cases 2 and 3 consisted of the normal constituents of normal vulva soft tissue, including mature adipose tissue, fibroblast, collagen, blood vessels, and nerves. The fibrous tissue surrounded fat lobules, nerves and blood vessels (Fig. 3). There was no nuclear atypia, mitosis, and bi/multinucleation in adipocytes and fibroblasts. Case 2 showed increased fibrous tissue, composed of bland spindle-shaped cells in a collagenous to edematous stroma, containing variable sized vessels, fat lobules, and peripheral nerve bundles. Case 3 was predominantly composed of fat tissue and showed similar histologic features with fibrolipoma. The fibroblasts in all cases stained for estrogen receptor (ER) (Fig. 4). Fibroblasts were stained faintly for CD34 in cases 1, 2, and 4. Follow-up information was available for two patients. Case 1 showed another ill-defined heterogeneous soft tissue echo in right vulva on ultrasonography 14 months later. It was not excised. There was no recurrence in the left vulva. Case 4 had no recurrence after excision for 12 months.

**Table 1.** Clinicopathologic features of cases

| Case | Age (yr) | Sex | Clinical presentation     | Size (cm) | Treatment | Excision year |
|------|----------|-----|---------------------------|-----------|-----------|---------------|
| 1    | 7        | F   | Left vulva enlargement    | 5         | Excision  | 2004          |
| 2    | 8        | F   | Right vulva enlargement   | 3         | Excision  | 2003          |
| 3    | 5        | F   | Left labium major bulging | 4.5       | Excision  | 2007          |
| 4    | 8        | F   | Right vulva enlargement   | 5         | Excision  | 2009          |

F, female.

**Fig. 1.** An ill-defined bulge in the left labium majus (case 3).**Fig. 2.** The excised tissue composed of ill-defined fibro-fatty tissue without hemorrhage and necrosis (case 4).**Fig. 3.** On microscopic examination, the lesion consists of the constituents of normal vulva soft tissue, including mature adipose tissue, fibroblasts, collagen, blood vessels, and nerves. (A) Case 4. (B) Case 1.

## DISCUSSION

CALME is a distinctive clinicopathologic entity of pre and early puberty first described by Vargas *et al.*<sup>1</sup> in 2005. CALME encompasses prepubertal vulval fibroma, fibrolipoma, hamartoma, and fibrous hyperplasia that have been previously reported according to the proportions of the constituent.<sup>2-6</sup> Iwasa and Fletcher<sup>7</sup> designated it as prepubertal vulval fibroma because it

showed the distinct features in the site and age. Altchek *et al.*<sup>4</sup> called it prepubertal unilateral fibrous hyperplasia. However, Vargas *et al.*<sup>1</sup> proposed the designation CALME because it did not form a distinct mass and resembled normal vulva soft tissue, and could resolve spontaneously.

The most common clinical presentation of CALME is a painless, fluctuating, non-tender bulging on either side of labia majus.<sup>8</sup> Although the majority of the cases are unilateral, bilateral

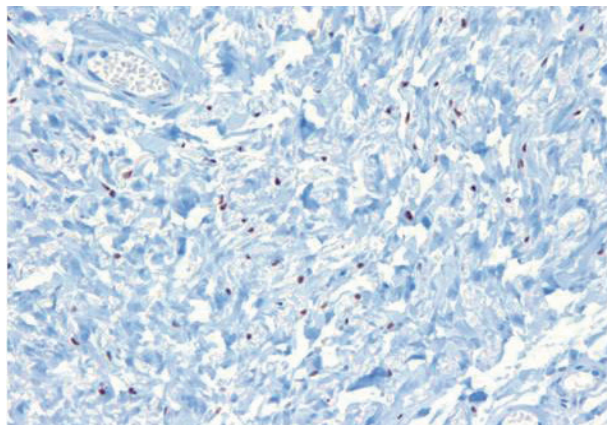


Fig. 4. Spindle shaped fibroblasts stained for estrogen receptor.

presentation is also noted. Oh *et al.*<sup>6</sup> described that vulva lipoma in children tend to occur on right antero-lateral side. All our cases were unilateral, two right and two left. An asymmetric presentation of CALME is still questionable although it occurs by hormonal response. The clinical onset of swelling usually occurs 7 years of age or younger. Physical examination showed ill-defined enlargement of the labium majus.

Radiologically, CALME reveals no abnormal findings except an enlargement of labial soft tissue.<sup>8</sup> Microscopically, CALME consists of normal constituents of vulva soft tissue, including fibroblasts, collagen, adipose tissue, blood vessels, and nerves. Vargas *et al.*<sup>1</sup> reviewed 14 cases of surgically resected CALME, all cases showed predominance of fibrous tissue. The fibroblasts were plump, round, or occasionally spindled with small dark slender nuclei, and were positive for estrogen and progesterone receptors.<sup>1</sup> In our cases, fibroblasts stained positive for ER in all cases and stained faintly for CD34 in three of the four cases.

Vargas *et al.*<sup>1</sup> described a recurrence rate of CALME of about 50% after resection. Soyer *et al.*<sup>8</sup> reported a case of conservatively treated CALME. They suggested that surgical intervention and biopsy should be reserved for the patients who have an un-

clear diagnosis and suspicion of malignancies, or who wish to achieve better cosmetic appearance, because CALME is a physiologic response to puberty and may resolve spontaneously and recur after total excision.<sup>2</sup> Altchek *et al.*<sup>4</sup> suggested that surgeons should not attempt a complete removal because simple excision is sufficient for the diagnosis and cosmetic effect.

In conclusion, CALME is a site- and age-specific non-neoplastic physiologic response to hormone. Recognition of this distinctive entity is important to avoid surgical excision.

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