Steatocystoma Multiplex Limited to the Vulva: Report of a Very Rare Case Successfully Treated by a Simple Surgical Method

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Authors’ contributions

This work was carried out in collaboration between all authors. Author SPK provided the case, performed the surgery and wrote the draft of the manuscript. Authors ES, MA and MG designed the pathological figures, managed literature searches and contributed to the writing. All authors read and approved the final manuscript.

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ABSTRACT

Aim: Steatocystoma multiplex (SM) is a rare malformation characterized by multiple cutaneous cystic lesions containing oily substance. SM has autosomal dominant mode of inheritance, though there are a significant number of sporadic cases reported as well. Characteristically, the lesions manifest around the time of puberty and are found on the sternal areas, face, trunk and scrotum. SM limited to the vulva is a very rare condition as there are only a five cases in the literature.

Case: A 32-year-old woman presented with a 1 year-history of nodules, gradually increasing in size and number, on the labia majora. Similar lesions were not present on other parts of her body and family members. On physical examination, yellowish papules and nodules ranging in size from 2-15mm without punctum were present on the labia majora. Biopsy performed at the time revealed
histological changes with steatocystoma multiplex. After a mini incision, evacuation of the cyst followed by the removal of the cyst wall was performed without complication. The lesions healed rapidly without scarring. The patient was followed by yearly controls. Recurrence was not observed until the fourth year of surgery. Smaller lesions were observed on previously untreated areas of vulva which were removed again successfully.

**Discussion:** SM treatment is difficult. In addition to medical treatments including antibiotic and anti-inflammatory drugs and isotretinoin, there are different surgical treatments including total excision and grafting, cryotherapy and CO2 laser with limited success.

**Conclusion:** This is a very rare case of SM limited to the vulva, simply and successfully treated by a simple surgical method, emphasizing the importance of cyst wall removal to achieve long time remission.

**Keywords:** Steatocystoma multiplex; vulva; treatment; surgery.

1. **INTRODUCTION**

Steatocystoma multiplex [SM] is a rare hamartomatous malformation characterized by multiple cutaneous cystic lesions varying in size, containing oily substance. SM has a predominantly autosomal dominant mode of inheritance and has been linked to defects in the keratin 17 gene [1]. In addition to familial cases, sporadic cases have been described. Characteristically, the lesions manifest around the time of puberty and are distributed in areas where high numbers of sebaceous glands are found, most commonly the chest, arms, axillae, and neck. There are only five cases of SM limited to the vulva reported in the literature [1,2]. We present another case of SM limited to vulva treated successfully by a simple surgical removal.

2. **CASE**

The patient was a 32-year-old woman who had a 1-year history of nodules, gradually increasing in size and number, on the labia majora. Similar lesions were not present on other parts of her body and family members. On physical examination yellowish, soft, movable papules and nodules, ranging in size from 2-15 mm, without punctum and inflammation were present on the labia majora (Fig. 1a).

No other cutaneous abnormalities were present. After a written informed consent was taken from the patient biopsy performed at the time showed sebaceous gland lobule in the cyst wall (Fig. 1b). SM was diagnosed. We used a simple surgical method for the treatment. We performed a careful anesthetic injection by avoiding the cyst wall rupture. After a mini incision using no. 15 scalpel blade or sharp cautery tip, evacuation of the cyst followed by the removal of the cyst wall by forceps were performed without complication (Fig. 2a).

Topical and systemic antibiotics were commenced and the wound was left to heal secondarily. The lesions healed rapidly without scar (Fig. 2b). She was followed by yearly controls (Fig. 2c). 4 year after the surgery, we noticed new lesions in small sizes and less amounts which had been slowly growing for 3 months (Fig. 3a).

Fig. 1a. Steatocystoma multiplex lesions on external part of the labia majora, [b] Sebaceous gland on the cyst wall [Arrows] H&E x200
We performed a similar surgery with success (Figs. 3b-d). When compared with previous lesions, new lesions were observed to be located on previously untreated areas of vulva (Fig. 4).

3. DISCUSSION

SM has been reported on the face and neck, scrotum, penis, palms and soles, acral sites, scalp, retroauricular and gluteal region before [1-7]. Generally, cysts appear during adolescence and early adulthood and there is no sex predilection [8]. SM have been associated with pachyonichia congenita type 2 (PC-2) (Jackson-Lawyer syndrome) and some other conditions like ichthyosis, acrokeratosis verruciformis, hypohidrosis, hidradenitis suppurativa [2,8]. The familial SM is associated
with a mutation in the keratin 17 gene identical to the mutations found in patients with PC-2 which is an autosomal dominant syndrome characterized by hypertrophic nail dystrophy, hair and teeth abnormalities as well as pilosebaceous cysts [8]. Our patient had no family history, features of PC-2 or any cutaneous or systemic abnormalities. The differential diagnoses of the disease are eruptive vellus hair cysts (EVHCs), epidermal inclusion cyst, milia, trichilemmal cysts and follicular infundibulum tumours. The histopathological features leads the diagnoses. EVHCs are dermal cysts containing vellus hair shafts in the lesion. The epidermoid cysts are lined with stratified squamous epithelium with granular layer and laminated keratin inside the lumen. The milium is mainly located in the superficial dermis with a granular layer. The follicular infundibulum tumours include dermal tumour islands in connection with epidermis. The characteristic histopathological finding of SM is the presence of sebaceous lobules close to the cystic wall, which is lined by stratified squamous epithelium without a granular layer [8]. The sebum containing cysts generally have normal overlying epidermis with no central punctum. The histopathological findings of our case showed the characteristics for SM. Although any part of the body might be affected, there are only five reports dealing with sebaceous cystic lesions of the vulva [1,3,9,10]. The previous five reports are outlined in Table 1.

According to the reported five cases vulvar SM seems to manifest in elderly. However, our patient was 32 years old at the time of her first visit. Among reported patients, none were treated satisfactorily because, isotretinoin therapy, as reportedly, resulted in worsening of the lesions and only one patient had underwent surgery with limited success and others remained untreated [1,2].

SM is usually asymptomatic unless became infected, however patients are affected psychologically. Indeed, our patient had embarrassment and demanded treatment. Although it has benign nature, SM treatment is difficult. In addition to medical treatments including antibiotic and anti-inflammatory drugs and isotretinoin, there are different surgical treatments including total excision and grafting, cryotherapy and CO2 laser with limited success [1-7]. As total excision has risk of scarring, suggested simple surgical methods are summarized in Table 2.

Drainage and extraction following incision has been tried for SM located on body and face [4-7,11-15]. We used a simple and safe method to treat the lesions. No recurrence was observed until the fourth year of surgery, which was a satisfactory result for the patient. Interestingly it was noticeable that new lesions were located especially on previously untreated areas.

### Table 1. Reported cases of vulvar steatocystoma multiplex

<table>
<thead>
<tr>
<th>Report</th>
<th>Year</th>
<th>Patient age</th>
<th>Disease duration</th>
<th>Histopathology</th>
<th>F/S</th>
<th>Treatment</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lewis B, [9]</td>
<td>1948</td>
<td>37</td>
<td>Unknown</td>
<td>Non characteristic</td>
<td>Familial</td>
<td>Excision</td>
<td>Unknown</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Sebaceous gland on the cyst wall</td>
<td>Sporadic</td>
<td>Some lesions were excised</td>
<td>Unknown</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Sebaceous gland on the cyst wall</td>
<td>Sporadic</td>
<td>No treatment</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

### Table 2. Suggested simple surgical options for steatocystoma multiplex

| Keefe M, et al. [13] | 1992 | 4 mm-1 cm incision with no.15 scalpel blades and evacuation of the cyst, followed by removal of the cyst wall with forceps |
| Adams B, et al. [14] | 1999 | Incision with no. 11 blade and cyst wall removal by small artery forceps |
| Kaya T, et al. [7]   | 2001 | Small puncture with sharp tipped cautery and squeezing out the content followed by grasping the protruded sac and pulling it out |
| Madan V, et al. [15] | 2009 | Perforation with Co2 laser and extirpation of the cyst wall using Volkmann’s spoon |
After the SM surgery, oral antibiotics are reported to be generally unnecessary. However, we preferred to commence systemic empiric antibacterial treatment that generally used for skin and soft tissue, because our patient’s lesions were on vulvar area, which was susceptible to infection.

4. CONCLUSION

Our treatment revisited a simple surgical technique emphasizing the importance of cyst wall removal that resulted in long time remission without scarring and complication.

CONSENT

All authors declare that 'written informed consent' was obtained from the patient for publication of this paper and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES


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