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Steatocystoma Multiplex of the Vulva: A Case Report

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1. Abstract

Steatocystoma multiplex is a rare and unique skin disorder, characterized by the development of multiple benign sebum-containing dermal cysts. These cysts are most commonly observed on the upper anterior portion of the trunk, upper arms, thighs, scalp but can occur anywhere. Only a few cases of the patients with steatocystoma multiplex strictly confined to the vulva had been reported. Herein, We report a case of steatocystoma multiplex of the vulva succefully treated by mini-incisions of the cyst followed by expression of cyst contents.

2. Introduction

Steatocystoma Multiplex (SM) is a rare clinical disorder implying hamartomatous malformation of the pilosebaceous duct junction. Even if it is often inherited in an autosomal dominant fashion, many poradic cases has been reported. SM is characterized clinically by the presence of multiple symptomatic cysts principally located on the trunk, scalp, axilla, inguinal region, and proximal extremities. Diagnosis is clinical and is confirmed by histopathology [1]. SM limited to the vulva is a rare clinical entity with only a few cases reported in literature.

3. Observation

A 24-year-old women who presented to our dermatology department with a history of numerous, painless papules over the genital area for the last 5 years. Physical examination showed a multiple, small, mobile, firm, white and yellow cystic papulonodules discretely distributed over the vulva (Figure 1). There was no similar familiar history of lesions or nail dystrophy. The laboratory findings were normal.A skin biopsy from a cyst showed stratified squamous epithelium without any granular layer or eosinophilic cuticle. Hence diagnosis of steatocystoma multiplex was made and classical excision of the lesions under local anesthesia was performed. The patient was followed up for 8 months with no recurrence. The DLQI score decreased from 18 to 5 points.

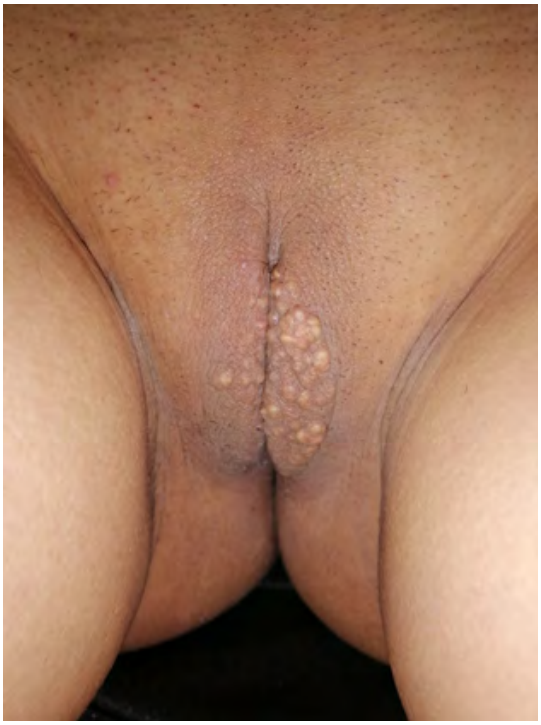

Figure 1: Multiple, small, mobile, firm, white and yellow cystic papulonodules discretely distributed over the vulva.

4. Discussion

SM is a benign autosomal dominant disorder associated with mutations in the keratin 17 gene. It was first described in 1873 by Jamieson and named by pringle in 1899 [2]. The lesions may appears shortly after birth but also late in life [3]. There is no predilection to gender [4].The etiology of this entity is still unclear but infection, trauma, and immunological disorders may play a role of this condition [4]. In our case, there were no associated findings. Clinically, SM is characterized by several, uniform, soft,yellowish or skin coloured cystic nodules and papules of 1 to 1,5 cm diameter, mostly on the chest, neck, axilla, scalp and proximal extremities [5]. In our case, it has occurred on the genital area. The clinical differential diagnosis of steatocystoma multiplex eruptive vellus hair cysts, neurofibromatosis, trichilemmal cysts, epidermal inclusion cysts and lipoma. The SM can be associated with pachyonychia congenital, hypertrophic lichen planus, hidradenitis suppurativa [6]. Diagnosis is based generally on clinical fundings but can be confirmed by histopathology. Most patients are asymptomatic and seek treatment for esthetic reasons. Treatment options include surgery, oral isotretinoin, CO2 Laser therapy, cryotherapy, needle aspiration. Mini incision with drainage and extraction is still the better choice [7].

5. Conclusion

SM limited to the vulva is a rare clinical entity and its incidence is still unknown. Treatment is usually required due to its psychological affecting of the well-being of the patients.

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