



Case Report

Genital Folliculosebaceous Cystic Hamartoma: A Case Report and Concise Review of the Literature

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Abstract: Folliculosebaceous cystic hamartoma (FSCH) is an uncommon hamartoma that usually presents on the central face area of adults as an asymptomatic, solitary dome-shaped or pedunculated papule. We report a case of a 35-year-old female who presented with six-months history of skin lesions on her labia majora. Histological findings included cystically dilated hair follicles with branching epithelial strands and interconnecting sebaceous gland consistent with the diagnosis of FSCH. The genital variant of FSCH was first described in 1998 and since then only six cases have been reported in the literature. We aim to increase awareness of this rare presentation due to the significant psychological implications and the risk of misdiagnosis.

Keywords: folliculosebaceous cystic hamartoma; sebaceous trichofolliculoma; genital hamartoma

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1. Introduction

Folliculosebaceous cystic hamartoma (FSCH) is a hamartoma composed of follicular, sebaceous and mesenchymal elements. It usually presents as an asymptomatic, solitary dome-shaped or pedunculated papule or nodule on the head in adult patients, with predilection for the central face and nose [1]. Here, we report a case of the rare genital variant of FSCH.

2. Detailed Case Description

A 35-year-old female presented with multiple asymptomatic papules and nodules on the labia majora. The lesions were of 6 months duration and gradually increased in number and size. They were rounded, skin-colored, pedunculated, firm, had a smooth surface and ranged from 0.5–2 cm in diameter. The patient was concerned about having viral warts. Her husband did not have history of similar lesions. The clinical differential diagnosis included condyloma lata, condyloma accuminata, fibrolipomas, and adnexal tumors. Excision of 3 lesions were performed and histological examination revealed multiple cystically dilated and distorted hair follicles surrounded by thin anastomosing epithelial strands. Sebaceous glands were seen communicating with the cyst through small ductules. The surrounding stroma was composed of highly vascular, dense collagenous tissue and was separated from the surrounding adjacent uninvolved dermis by prominent clefting. A granulomatous inflammation was also noted (Figures 1–3). The features were found to be consistent with the diagnosis of genital folliculosebaceous cystic hamartoma.

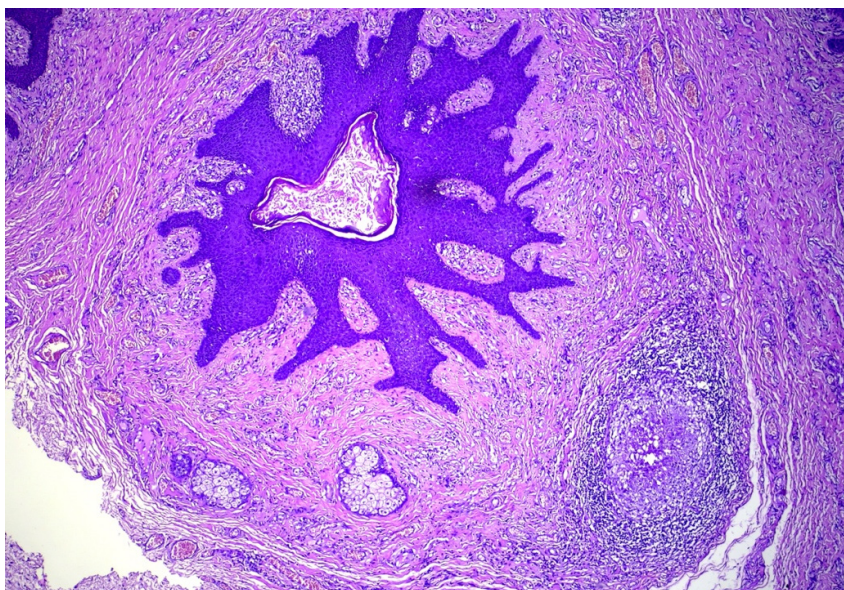


Figure 1. Cystically dilated hair follicle in the background of fibro-collagenous stroma. A secondary foreign body giant cell granulomatous reaction is also present. Hematoxylin and eosin, (H&E) $\times 100$.

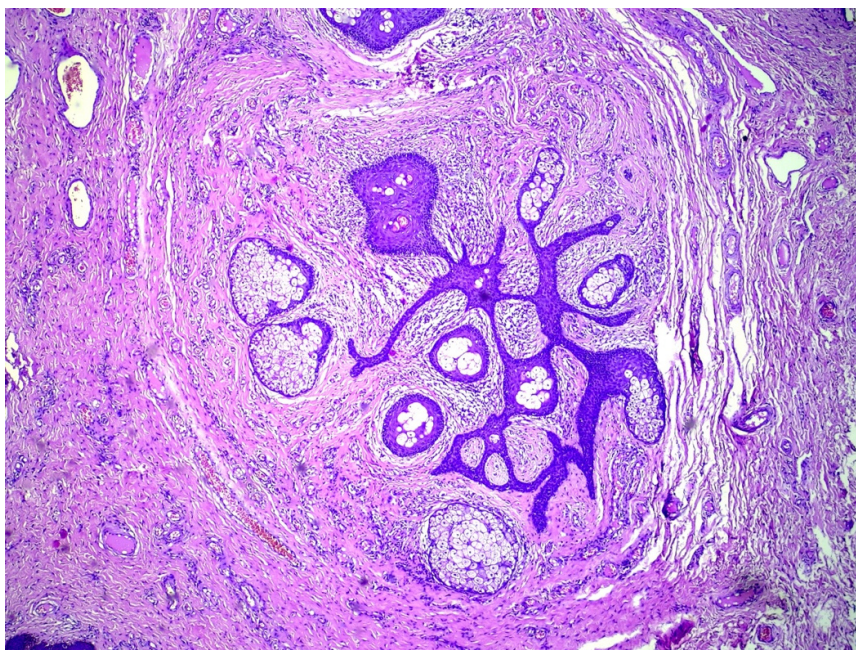


Figure 2. Anastomosing epithelial strands with multiple radiating sebaceous glands. Artefactual retraction can be seen between the mesenchymal component and surrounding uninvolved dermis. (H&E) $\times 100$.

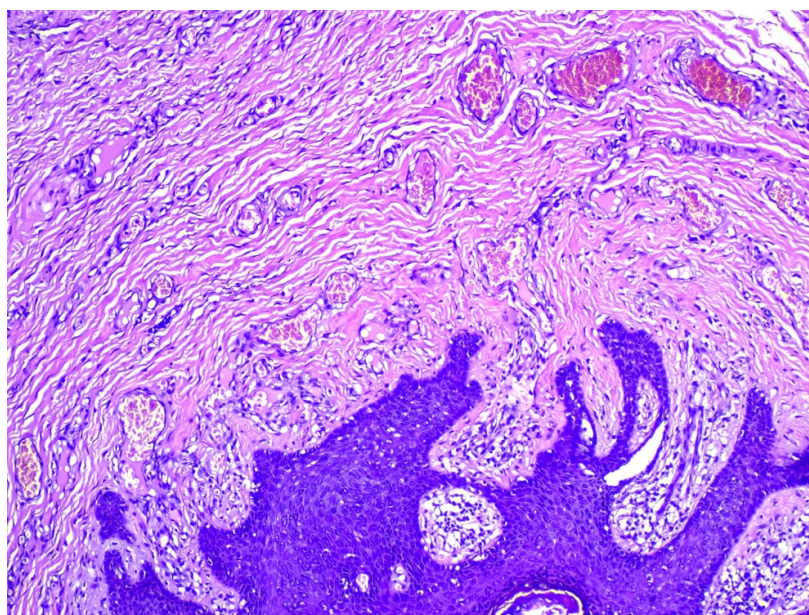


Figure 3. Stroma is collagenous and associated with numerous small sized blood vessels. (H&E) $\times 200$.

3. Discussion

FSCH was first described as a distinct entity by Kimura et al. in a series of five cases in 1991 [2]. The main diagnostic criteria on histological examination include presence of an epithelial component in the form of an infundibular cystic structure with radiating connected sebaceous lobules, and a mesenchymal element composed of collagenous stroma, adipocytes, and small blood vessels [3].

Bolognia and Longley described the genital variant of FSCH in 1998 [4]. The authors argued that a case of scrotal sebaceous trichofolliculoma (STF) published in 1991 by Nomura and Hata [5] in fact represented FSCH. Further six cases were reported afterwards in the literature [6–11]. Genital FSCH mostly manifests as single nodule or multinodular lesion on the labia majora in young to middle-aged females. Clinical characteristics from reported cases are summarized in (Table 1).

Table 1. Clinical features of genital FSCH cases reported in the literature.

Author	Age (Years)	Sex	Size	Site	Duration	Clinical Description	Notes
Bolognia and Longley [4]	34	F	4 cm	Left labia majora	Not available	Skin-coloured multinodular plaque	
Hamada et al. [6]	40	F	5 cm	Pubis	13 years	Three skin-coloured aggregated nodules	
Brucher et al. [7]	74	M	23 \times 16 cm	Whole scrotum	Since birth	Giant multinodular skin-coloured to yellowish lesion	
Park et al. [8]	28	F	Not available	Labia majora	Not available	Not available	Full article is not available in English
Wu et al. [9]	28	F	6 \times 2 \times 1.5 cm (left) and 5.5 \times 2.5 \times 2 cm (right)	Bilateral labia majora	More than 10 years	Skin-coloured polypoid and pedunculated hypertrophy	Associated with fibrous dysplasia syndrome
Alegría-Land et al. [10]	45	F	2 cm	Middle of right labia majora	Long-term (patient uncertain)	Skin-coloured nodule	
Khan et al. [11]	29	F	5 \times 3 cm	Right labia majora	3 months	Giant papulonodular lesion	

FSCH does not have distinguishing features clinically and can be easily mistaken for neurofibroma, lipoma, intradermal nevus, and sebaceous hyperplasia [3]. Viral warts

would be an important differential diagnosis in the context of genital lesions due to the potential misdiagnosis and the psychological distress it can cause to the patients. However, histological examination would be sufficient to differentiate FSCH [3].

The main histological differential diagnosis tends to be STF, which presents as a depressed lesion clinically. It tends to show hair shafts within the dilated hair follicle, displays branching finger-like smaller follicles, and a more frequent connection to the epidermis. It also lacks the characteristic stroma of FSCH [12]. Due to the overlapping features reported in some lesions, it is hypothesized that FSCH may represent a late stage in development of STF. Both share similar pathogenesis background in the form of CK15-positive hair follicle stem cells activation [13,14].

Treatment in most cases has been surgical excision [11]. Successful combination of CO₂ laser and acitretin has been described in one patient with a giant lesion [7].

4. Conclusions

To the best of our knowledge, we report the 8th case of genital FSCH. Awareness of this rare diagnosis is important due to the associated psychological distress, cosmetic and sexual issues, and anxiety of an HPV diagnosis.

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References

1. Haw, S.; Lee, M.-H. A Case of Giant Folliculosebaceous Cystic Hamartoma. *Ann. Dermatol.* **2009**, *21*, 63–65. <https://doi.org/10.5021/ad.2009.21.1.63>.
2. Kimura, T.; Miyazawa, H.; Aoyagi, T.; Ackerman, A.B. Folliculosebaceous cystic hamartoma. A distinctive malformation of the skin. *Am. J. Dermatopathol.* **1991**, *13*, 213–220.
3. Noh, S.; Kwon, J.E.; Gil Lee, K.; Roh, M.R. Folliculosebaceous Cystic Hamartoma in a Patient with Neurofibromatosis Type I. *Ann. Dermatol.* **2011**, *23*, S185–S187. <https://doi.org/10.5021/ad.2011.23.s2.s185>.
4. Bologna, J.; Longley, B. Genital Variant of Folliculosebaceous Cystic Hamartoma. *Dermatology* **1998**, *197*, 258–260. <https://doi.org/10.1159/000018008>.
5. Nomura, M.; Hata, S. Sebaceous Trichofolliculoma on Scrotum and Penis. *Dermatology* **1990**, *181*, 68–70. <https://doi.org/10.1159/000247867>.
6. Hamada, M.; Kiryu, H.; Satoh, E.; Moroi, Y.; Urabe, K.; Furue, M. A case of genital folliculosebaceous cystic hamartoma with an unique aggregated manifestation. *J. Dermatol.* **2006**, *33*, 191–195. <https://doi.org/10.1111/j.1346-8138.2006.00044.x>.
7. Brücher, J.-J.; Franke, I.; Ulrich, J.; Gollnick, H.; Leverkus, M. Giant genital variant of folliculosebaceous cystic hamartoma: Successful management by CO₂laser and acitretin therapy. *Br. J. Dermatol.* **2007**, *157*, 833–835. <https://doi.org/10.1111/j.1365-2133.2007.08121.x>.
8. Park, H.; Joo, M.; Lee, Y. A Case of Folliculosebaceous Cystic Hamartoma on the labia majora. *Korean J. Dermatol.* **2007**, *45*, 221–223.
9. Wu, H.-S.; Cheng, H.-T.; Jung, S.-M.; Chang, S.C.-N. Distinctive Genital Folliculosebaceous Cystic Hamartomas in a Patient With Fibrous Dysplasia. *J. Craniofacial. Surg.* **2012**, *23*, e132–e134. <https://doi.org/10.1097/scs.0b013e31824cdafc>.
10. Alegria-Landa, V.; Prieto-Torres, L.; Requena, L.; Velasco, M.J. Genital folliculo-sebaceous cystic hamartoma: A claim of the stroma as a clue in the diagnosis of proliferations with follicular differentiation. *J. Cutan. Pathol.* **2017**, *44*, 504–508. <https://doi.org/10.1111/cup.12920>.
11. Khan, A.; Gowda, C.; Vasudevan, G.; Rodrigues, G. Giant folliculosebaceous cystic hamartoma of the vulva: A rare occurrence. *Int. J. Gynecol. Obstet.* **2020**, *150*, 423–424. <https://doi.org/10.1002/ijgo.13286>.
12. Plewig, G. Sebaceous Trichofolliculoma. *J. Cutan. Pathol.* **1980**, *7*, 394–403. <https://doi.org/10.1111/j.1600-0560.1980.tb01213.x>.

13. Shin, E.-J.; Jeong, K.-H.; Shin, M.-K. A Case of Late Stage Sebaceous Trichofolliculoma Showing Overlapping Features with Folliculosebaceous Cystic Hamartoma. *Ann. Dermatol.* **2016**, *28*, 511–512. <https://doi.org/10.5021/ad.2016.28.4.511>.
14. Misago, N.; Kimura, T.; Toda, S.; Mori, T.; Narisawa, Y. A Revaluation of Folliculosebaceous Cystic Hamartoma: The Histopathological and Immunohistochemical Features. *Am. J. Dermatopathol.* **2010**, *32*, 154–161. <https://doi.org/10.1097/dad.0b013e3181a9de8c>.