

Dome-Shaped Papule With a Bloody Crust

Li-qiang Zheng, MD; Xiang-chun Han, MD; Cheng-xin Li, MD, PhD



A 3-year-old girl was referred to our clinic for a lesion on the face that had been present since birth and had enlarged slowly with slight itching. Physical examination revealed a 1.0×1.0-cm, sessile, flesh-colored, sharply demarcated, and dome-shaped papule with a bloody crust. It was firm and slightly painful to palpation. Dilated hair follicle-like orifices and thick central terminal hair were not found. Sebaceous material was not discharged. There was no notable family history or evidence of systemic disease. The lesion was surgically removed for cosmetic reasons and further histopathologic examination was performed.

What's the diagnosis?

- a. congenital folliculosebaceous cystic hamartoma
- b. molluscum contagiosum
- c. pilomatrixoma
- d. sebaceous trichofolliculoma
- e. Spitz nevus

Drs. Zheng and Li are from the Department of Dermatology, Chinese People's Liberation Army General Hospital, Beijing. Dr. Zheng also is from the 251st Hospital of Chinese PLA, Zhangjiakou City, China. Dr. Han is from the Department of Pathology, First Affiliated Hospital of Hebei North University, Zhangjiakou City.

The authors report no conflict of interest.

Correspondence: Li-qiang Zheng, MD, Department of Dermatology, Chinese People's Liberation Army General Hospital, 28 Fuxing Rd, Beijing 100853, China (zlqiang1976@163.com).

The Diagnosis: Congenital Folliculosebaceous Cystic Hamartoma

Folliculosebaceous cystic hamartoma (FSCH) is a rare skin condition that is either congenital or acquired. It presents as a slow-growing and flesh-colored papulonodular lesion¹ that mainly occurs on the head and neck. Involvement of the nipples, perineum, back, forearms, genital areas, and subcutaneous tissue also has been reported but usually indicates a larger lesion.^{1,2}

Histologically, FSCH is considered a hamartoma composed of both ectodermal and mesodermal elements.¹ Folliculosebaceous cystic hamartoma is a more complex lesion composed of infundibulocystic structures connected to maloriented folliculosebaceous units surrounded by whorls of highly vascularized fibrous stroma and adipocytes. Clefts between fibroepithelial units and surrounding stroma usually are present.¹

Epithelial components contribute to the adnexal and folliculosebaceous cystic proliferations, and mesenchymal elements include vascular tissue, adipose tissue, and fibroblast-rich stroma.^{1,2} Acquired lesions arising in adults have been described,¹⁻⁵ but the congenital presentation of FSCH in infancy is rare.

Histopathologically, some variations of FSCH are mainly composed of epithelial components while others are composed of nonepithelial components. Nonepithelial components include neural proliferation, muscle components, vascular proliferation, and mucin deposition.¹⁻⁴ In some cases, FSCH may coexist with other diseases, such as nevus lipomatosus cutaneous superficialis and neurofibromatosis type I.^{4,5}

In our case, histopathology showed several dermal infundibulocystic structures that were lined by stratified squamous epithelium and contained horny material (Figure 1). Numerous immature sebaceous lobules and rudimentary hair follicles emanated from some of the cyst walls. Mesenchymal changes around the fibroepithelial units included fibrillary bundles of collagen, clusters of adipocytes, and an increased number of small venules (Figure 2). In addition, the stroma adjacent to the malformed perifollicle contained some amount of mucin. Prominent clefts formed between fibroepithelial units and the surrounding altered stroma.

The differential diagnosis mainly includes sebaceous trichofolliculoma, molluscum contagiosum, dermoid cysts, pilomatrixoma, Spitz nevus, and nevus

lipomatosus superficialis. The differential diagnosis between FSCH and sebaceous trichofolliculoma is challenging. Both lesions show an infundibular cyst and surrounding sebaceous nodules. According to Plewig,⁶ trichofolliculoma has a wide spectrum ranging from low to high differentiation represented by trichofolliculoma, sebaceous trichofolliculoma, and FSCH, respectively. It is not difficult to distinguish FSCH from other diseases according to its peculiar histopathologic features.

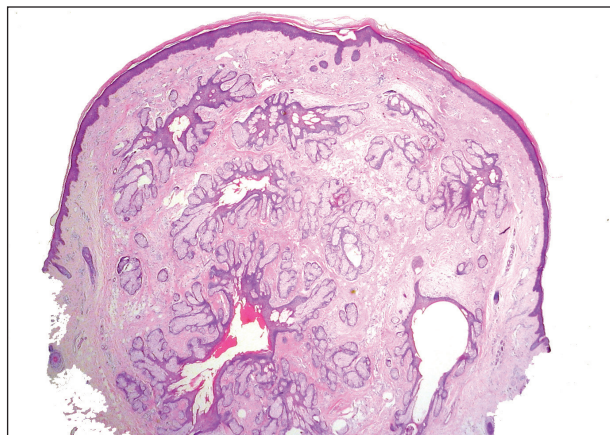


Figure 1. Several dermal infundibulocystic structures were lined by stratified squamous epithelium and contained horny material (H&E, original magnification $\times 40$).

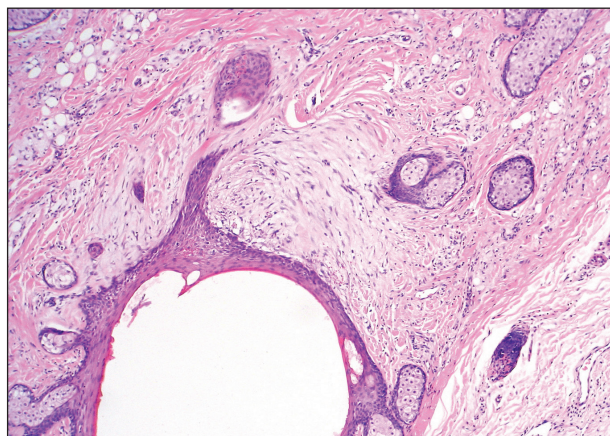


Figure 2. Ectopic focal adipocytes deposited in the dermis and among cystic structures. There were loose myxoid substance deposits prominently in perifollicle regions (H&E, original magnification $\times 200$).

The clinicopathologic features of our case were similar to those of reported FSCH cases, except for the following unique characteristics: congenital lesion, lack of terminal hair, and no sebaceous material extrusion. These features of hair and sebaceous material may be correlated with the patient's age and hormonal level.¹ Androgen may play a key role in sebaceous gland development at puberty, which leads to sebaceous gland hyperplasia and hypertrophy. Therefore, slight pressure from the lesions can make ivory-white sebaceous material discharge. Hence, the dermatologist and pediatrician must be poised and sensitive while making an initial diagnosis of FSCH.

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