An atypical form of favus is undiagnosed for 50 years

Ru-zhi Zhang¹ and Wen-yuan Zhu²*

¹Department of Dermatology, Third Affiliated Hospital of Suzhou University, 185 Juqian Road, Changzhou, 213003, China
²Department of Dermatology, the First Affiliated Hospital of Nanjing Medical University, Nanjing 210029, China

Abstract
Tinea capitis favosa, also termed favus, is a chronic inflammatory dermatophyte infection of the scalp usually caused by Trichophyton schoenleinii. It can be acquired during childhood or adolescence and typically persists into adulthood. The reported duration is wide, ranging from 10 days to 59 years. In addition to typical scutular favus on the scalp, several atypical manifestations of favus have been described, such as pityoides, psoriasiform and impetiginous forms. We describe a 55-year-old woman who had crusts and scars on her scalp for 50 years. She did not receive a correct diagnosis or treatment until presentation to our clinic. She was diagnosed with atypical favus based on the clinical manifestations and a mycological examination, which detected Trichophyton schoenleinii.

Keywords: Favus; Tinea Favosa; Scalp; Differential diagnosis

Corresponding author: Wenyuan Zhu, Department of Dermatology, The First Affiliated Hospital of Nanjing Medical University, China; E-mail: zhuwenyuan@yahoo.com

Favus, also termed tinea favosa, is a chronic inflammatory dermatophytic infection of the scalp, usually caused by Trichophyton schoenleinii [1]. It is one of three primary patterns of hair infection (ectothrix, endothrix, favus) characterized by the presence of scutula. Of individuals infected with favus, 95% have overt clinical symptoms, e.g., scutula, pale hairs, atrophy and scarring [2]. The classic lesion is the scutula, a concave, cup-shaped yellow crust, typically centrally pierced by a hair. The scalp is characterized by an unpleasant ‘cheesy’ or ‘mousy’ odor [3]. In addition to the typical scutular form of favus, atypical tinea favosa makes up about 5% of the cases [2], including pityoides, psoriasiform and impetiginous forms [4]. Favus pityoides mimics dandruff or seborrheic dermatitis. Numerous small-to-large scales are present, and removal of those scales uncovers reddish, moist and scarring areas of skin. Favus psoriasiformis is a psoriasis-imitating favus. Instead of yellowish scutula, patients present with whitish scales mimicking the typical lesions of psoriasis. Favus impetiginous is characterized by yellowish (honey-colored) crusts imitating impetigo located on the scalp. Sabouraud noted another form of favus which is characterized by plaques of cicatricial alopecia bordered by folliculitis. This form does not correspond to a special clinical form but to a stage of the disease, a final result in its evolution, usually requiring many years to develop, which Sabouraud called “old, alopecia, and cicatricle favus.” Atypical tinea favosa may not be readily recognized and may persist for years before being correctly diagnosed. Herein, we report one case of atypical favus that persisted for 50 years before being correctly diagnosed.

Clinical Data

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Case Report
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A 55-year-old woman complained of crusty lesions and scars on her scalp for 50 years. She was born and spent most of her life in a rural village of the Anhui province in southeast China, where favus was endemic at that time. At the beginning, there were crusted plaques on her scalp associated with itching at the age of 5 years. The crusts increased in size over time and presented as yellowish cup-shaped lesions with an unpleasant mousy odor. During her childhood, she received various forms of external treatment at infrequent intervals. The lesions were replaced by smooth scars without hairs after the crusts fell off. Her condition improved after puberty.

Examination showed that there was a single plaque of partial alopecia involving almost the entire scalp except for a peripheral border made up of normal hairs that were free of infection. Close examination revealed seborrhic scales and follicular pustules associated with extensive atrophic scarring alopecia. The lesions were often centrally pierced by a single thin hair [Figure 1a].

**Figure 1:** Seborrhic scales and follicular pustules associated with extensive atrophic scarring alopecia on the patient’s scalp (a). Healing was obtained at the follow-up of 6 months (b).

The affected hairs could be easily epilated, and had a dull, lustreless appearance with adherent glassy follicular epithelial sheaths and peripilar yellowish-white scales at the base. Wood’s light examination revealed a green fluorescence of the infected hairs. Direct microscopic examination of infected hairs in 10% potassium hydroxide revealed patterns of endothrix consisting of many spores without hyphae and air spaces in the hair shafts [Figure 2].

**Figure 2:** Under microscopy, the infected hairs in 30% potassium hydroxide revealed the patterns of endothrix consisting of many spores without hyphae and air spaces in the hair shafts.

A fungal culture of the infected hairs and scales on Sabouraud glucose agar grew *Trichophyton schoenleinii*. The skin on her body and limbs was normal, and the nails were healthy. No other dermatological abnormality was observed. The patient was otherwise healthy with no history of immune suppression. No family members had a similar condition. Based on her medical history, clinical manifestations and the mycological examination, the patient was diagnosed with the atypical form of favus. She was treated orally with terbinafine 250 mg once a day for 6 weeks. Her scalp was shampooed daily, and all recent emerging hairs associated with minute pustules or with inflammatory reactions around the follicles were epilated with forceps followed by application of a liquid fungicide to the entire scalp. The patient has had an excellent response, and healing was observed at the 6 month follow-up visit [Figure 1b]. Meanwhile, subsequent microscopic examinations and fungal cultures of hairs were negative.
Figure: 1 B

Discussion

Favus is seen often in geographic regions where there is a great poverty and thus poor hygiene, malnutrition and little access to health care. A worldwide survey in the 2000s of the causative agents of tinea capitis revealed that *T. Schoenleinii* was still the predominant species in China (31.5%), Nigeria (28.1%) and Iran (21.5%) [2]. Improvements in living conditions and hygiene in China have been associated with the almost complete disappearance of favus, and favus is now rare in China. Thus, many dermatologists, especially the younger generation, often miss the diagnosis of this disease [5].

During initial infection, the fungal spores appear to enter through the unbroken cutaneous surface, and to germinate mostly in and around the hair follicles and sometimes in the hair shafts. The infection rarely resolves spontaneously, although the crusts, inflammation and debris may gradually subside over time. The course probably depends on host resistance and/or on a change in fungal virulence. According to the severity of the disease, three main stages have been described [2]. The first stage shows only erythema of the scalp, primarily around hair follicles. The second stage displays the formation of scutula with the beginning of hair loss. The third stage is the most severe stage which involves large areas of the scalp, with extensive hair loss, atrophy and scarring. According to Sabouraud, *Achorion schoenleini* is the only cause of the disease when it involves the scalp, no matter what its clinical form [4].

Although the initial infection is probably a childhood event in nearly all cases, it shows little if any tendency to clear spontaneously at puberty, particularly in women. Our patient may have displayed typical favus at the beginning of the disease, but did not receive an appropriate diagnosis or treatment. The specific type of hair invasion (endothrix favosa) contributes to the chronic course of favus, which may persist into adulthood [6]. Endothrix infections are more likely to result in outbreaks among family members or intimate friends [7]. Our patient is an astonishing example of a case of scalp favus with no material spread over a period of 50 years. Untreated or uncompleted favus is typically slowly progressive, eventually leading to alopecia of the scalp with atrophic scarring. The clinical presentation becomes atypical [8]. Our patient had been misdiagnosed with impetigo, however, oral antibiotics were ineffective. This case reminds us that when a patient’s scalp has the clinical appearance of seborrhea sicca, seborrheic dermatitis, psoriasis, tinea amiantacea, impetigo-like lesions or pseudopelade, their scalp should be thoroughly searched for miliary scutula, and in addition, filtered ultraviolet radiation should be employed because of the possibility of the presence of fluorescent hairs. It is very important to make a careful examination to avoid delaying the diagnosis and treatment.

The treatment outcome will depend to some extent on the stage at which the disease is arrested.

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