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## Case Studies

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# Necrotizing Infundibular Crystalline Folliculitis: A Case Report of an Exceptional Lesion of Unknown Etiology

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## Key Words

Necrotizing infundibular crystalline folliculitis · *Malassezia* · Mucin · Birefringent crystals

## Abstract

Necrotizing infundibular crystalline folliculitis is a rare follicular lesion of which the etiology is not well understood. Here we describe the case of a 71-year-old male patient presenting with multiple hyperkeratotic lesions localized on the forehead. Histopathological analysis of one of the lesions revealed a follicular invagination containing cellular debris and keratin lamellae containing filamentous mucinous material and numerous crystals birefringent in polarized light microscopy.

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## Background

Necrotizing infundibular crystalline folliculitis is a relatively rare folliculocentric disorder of yet unknown origin. Several pathogenetic mechanisms have been proposed in the past, but the entity still remains unclear. Lucke et al. [1] first described its histopathological findings in 1999, where the transepidermal elimination of urate-like crystals was observed. The term ‘necrotizing infundibular crystalline folliculitis’ was first used in 2001 by Kossard et al. [2]. In 2011, Denisjuk et al. [3] proposed to rename the lesion to ‘necrotizing ostial crystalline folliculitis’ due to the fact that in their study the most important pathognomonic feature of urate-like crystals was their location in the ostia and rarely in the infundibular region of the hair follicles. Microscopically, a follicular invagination is observed, filled with cellular debris and keratin lamellae embedded in Alcian blue-positive filamentous mucinous material [4, 5] which resembles urate-like crystalline material [6].

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Here we describe the case of a 71-year-old male patient who presented with multiple hyperkeratotic lesions of the forehead (fig. 1), clinically diagnosed as folliculitis versus Favre-Racouchot syndrome.

## Methods

Biopsy material obtained from the patient was fixed in formalin, embedded in paraffin, cut at 5 µm and stained with hematoxylin-eosin, periodic acid-Schiff and Alcian blue according to standard procedures.

## Results

On histological examination, a follicular invagination containing some cellular debris and keratin lamellae was observed (fig. 2a, b). In the ostium of this follicular structure, filamentous mucinous material was detected (fig. 2c). The filamentous material contained numerous crystals birefringent in polarized light microscopy (fig. 2d). In the surrounding dermis, an inflammatory infiltrate composed of lymphocytes, macrophages, neutrophils and eosinophils was present. Numerous fungal elements consistent with *Malassezia* yeasts were seen (data not shown), but no bacteria were observed.

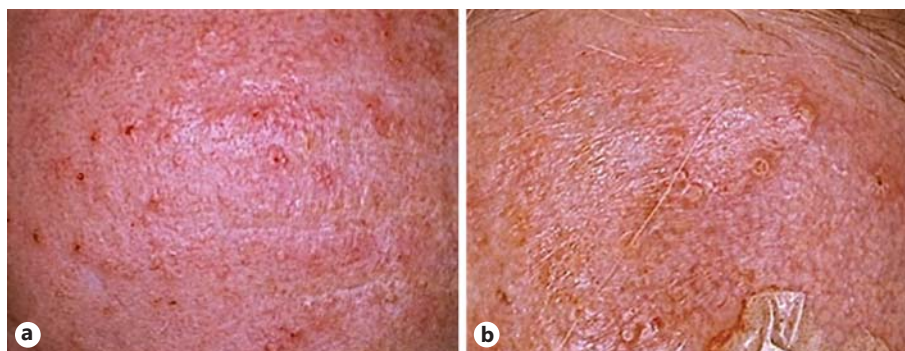
## Conclusion

The clinical and histological aspect of our case is quite typical for the so-called ‘necrotizing infundibular crystalline folliculitis’, a folliculocentric disease first reported in 2001 [2] on which only few data have been published. In a recently published study of Denisjuk et al. [3], 25 cases diagnosed in the past with necrotizing infundibular crystalline folliculitis (as solitary lesions or accompanied by skin neoplasms) or with ostiofolliculitis have been reviewed. Both sexes were equally affected; most of them in the 50th decade and presenting lesions at the same predilection sites as acne, but clinically different from it [3].

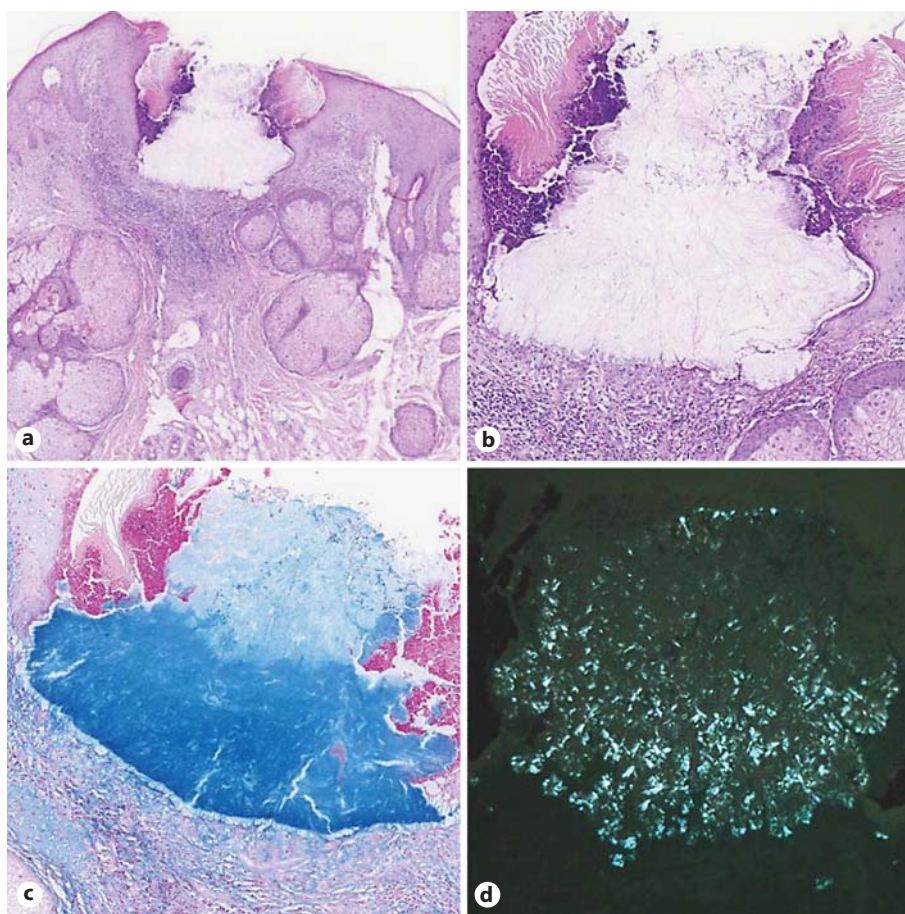
Etiology of this entity is not yet well understood. It remains unclear whether the presence of *Malassezia* plays a crucial role in the induction of crystalline structures [2] or mucin [7]. Another possible mechanism inducing the crystalline material seems to be a chemical or physical injury [2]. In our case, *Malassezia* yeasts were abundant in the biopsy described above, but they were absent in biopsies of clinically identical lesions of the same patient with a much less characteristic histological aspect. No bacteria have been observed in our case, and there were no anamnestic data including a possible chemical or physical injury. In the study of Denisjuk et al. [3], in 86% of all cases, yeasts and Gram-positive bacteria corresponding to *Propionibacterium acnes* have been identified.

Pathogenetically these lesions may represent a perforating disorder [5], but some cases have shown a primary folliculocentric necrosis [2] rather than a primary perforating process. Sebum accumulation as nutrient medium for the microorganisms has been discussed, accompanied by the accumulation of crystalline material due to the destruction of tonofilaments and the degradation of lipids, followed by a rupture of follicular epithelia and accumulation of inflammatory cells [3].

Histopathologically, the pathognomonic features are the birefringent urate-like crystals representing as Alcian blue-positive filamentous mucinous material. On electron microscopy, this crystalline material is composed of disrupted bundles of tonofilaments embedded in an



**Fig. 1.** Clinical appearance with hyperkeratotic papules on the forehead, clinically diagnosed as folliculitis versus Favre-Racouchot syndrome.



**Fig. 2.** Follicular invagination with cellular debris and keratin lamellae, surrounded by a dermal inflammatory infiltrate composed of lymphocytes, macrophages, neutrophils and eosinophils [hematoxylin-eosin; original magnification:  $\times 2$  (a) and  $\times 5$  (b)]; mucinous material within the follicular structure [Alcian blue; original magnification:  $\times 5$  (c)]; birefringent crystal deposits within the mucinous material [original magnification:  $\times 20$  (d)].

amorphous matrix [2]. It was proposed that it represents monosodium urate [1] produced in the skin as a result of increased cell proliferation following local physical or chemical injury [8]. A surrounding inflammatory infiltrate was observed in our case, but this feature has not been reported in all of the cases described in the literature. Clinical response to topical or systemic antimycotic treatment suggests a role of *Malassezia* yeasts in the pathogenesis of necrotizing infundibular crystalline folliculitis [3].

In conclusion, our case is one more example of this exceptional lesion of as yet unknown etiology. Analysis of more cases will shed light on the understanding of the exact pathogenesis of this peculiar skin disease.

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