



16-Year-Old Male with Pink Scaly Patches with Papules and Hair Loss

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Abstract

Alopecia mucinosa is a rare condition associated with follicular papules created by mucin deposition leading to hair loss. There appears to be no predilection for gender or age. Most commonly it presents as an idiopathic condition. However, it has been found to be associated with malignancies and inflammatory conditions, including mycosis fungoides and systemic lupus erythematosus. A biopsy will show mucin within the hair follicle and a lymphocytic infiltrate, which can aid in the diagnosis of associated conditions. Although spontaneous remission may occur, several treatment options are available that target the inflammatory nature of this condition. Treatment strategies vary depending upon disease severity and include corticosteroids for limited active disease, hydroxychloroquine, and antibiotics for moderate disease, and cyclosporine for severe disease have shown variable success rates. Janus kinase (JAK) inhibitors are being studied for lymphocytic cicatricial alopecias. Long-term follow-up is essential to evaluate for the development of secondary conditions.

Keywords

Alopecia mucinosa · Cicatricial alopecia · Follicular mucinosis · Corticosteroids · Photodynamic therapy

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A 16-year-old male presented with a pink, scaly patch on the scalp and forehead. The lesion on the scalp lacked hair. The lesions had been present for around 8 months. He received a course of oral terbinafine for 2 months with no improvement. He denied hair loss elsewhere on the body.

On physical examination, there were two pink scaly patches observed with follicular papules, one on the posterior scalp with hair loss and one on the left forehead. Eyebrows and eyelashes were of normal density. The fingernails were normal. A skin biopsy showed mucinous degeneration of the hair follicles and perifollicular lymphocytic inflammation with no atypical cytological features. The overlying epidermis showed no significant abnormalities.

Based on the clinical case description, what is the most likely diagnosis?

1. Alopecia mucinosa
2. Seborrheic dermatitis
3. Alopecia areata
4. Discoid lupus erythematosus

Diagnosis

Alopecia Mucinosa.

Discussion

Alopecia mucinosa is a rare type of lymphocytic primary cicatricial alopecia that presents with follicularly based papules and scaly plaques with subsequent hair loss. Alopecia mucinosa can affect patients at any age and involve any part of the body [1]. It tends to affect the head and neck, especially the scalp and eyebrows [1]. Clinically, alopecia mucinosa presents as follicular papules with or without erythematous scaly plaques and associated loss of hair [2]. There can be multiple well-defined, indurated plaques with patulous follicular ostia, seen best with dermoscopy [3]. The papules are grouped and can be associated with pruritus [4]. Less commonly, alopecia mucinosa may present morphologically similar to follicular cysts or keratosis, alopecia areata-like patches, dermatitis, urticaria, or acneiform lesions [5].

The terms alopecia mucinosa and follicular mucinosis are used synonymously and interchangeably in the literature. Follicular mucinosis refers to the nonspecific deposition of mucopolysaccharides such as mucin within the hair follicle [6]. Follicular mucinosis is a pattern of epithelium that can present in many disease states and alopecia mucinosa is an inflammatory disease that can present with follicular mucinosis [7].

Alopecia mucinosa can be separated into three classifications based on the presentation and course. First, there is a primary, acute form of alopecia mucinosa that is an idiopathic and spontaneous or remitting type seen in children and young adults

[8]. Second, there is a primary, chronic relapsing type seen in older adults [8]. Third, there is secondary alopecia mucinosa associated with malignancies and inflammatory conditions [8]. The most common malignancy associated with secondary alopecia mucinosa is cutaneous T-cell lymphoma, specifically mycosis fungoides [8]. Other associated malignancies that have been documented include Hodgkin's lymphoma, chronic lymphocytic leukemia, acute myelogenous leukemia, chronic myelogenous leukemia, and leukemia cutis [8]. The malignancy may be diagnosed before, along with, or after the diagnosis of alopecia mucinosa [6, 8]. Secondary alopecia mucinosa is also associated with several inflammatory conditions such as lupus erythematosus, sarcoidosis, alopecia areata, and lichen simplex chronicus [8].

Histologically, alopecia mucinosa is characterized by mucinous degeneration of the outer root sheath until the entire pilosebaceous unit is replaced by pools of mucin [9]. Mucin accumulation can cause swelling of the follicular epithelial cells and loss of intracellular bridges [8]. The mucin is made of hyaluronic acid, secreted by follicular keratinocytes [10]. The mucin can be stained by colloidal iron or alcian blue [8]. A lymphocytic infiltrate can be found in the follicular, perifollicular, and perivascular areas and diffusely in the dermis [8, 9].

Although alopecia mucinosa can frequently be diagnosed based on clinical features alone, a biopsy is recommended for confirmation. Alopecia mucinosa is often associated with cutaneous T-cell lymphoma, especially in elderly individuals, and further diagnostic testing of biopsies such as staining, and immunohistochemistry should be performed to rule out malignancy [4]. Long-term follow-up and serial biopsies are recommended since malignancy can coincide with or follow alopecia mucinosa [4, 6].

Treatment

There is no standard therapy available for alopecia mucinosa [4]. The key to any scarring alopecia is early control of the underlying disease to prevent permanent hair loss and reduction of symptoms. Treatment approaches are aimed at suppressing inflammatory processes and early treatment halts disease progression and allows for the possibility of scalp hair regrowth [11]. Treatments are grouped into three tiers based on increasing severity [11]. Tier 1 treatments are for patients with limited active disease. Treatment options include topical high-potency corticosteroids, intralesional steroids, or topical nonsteroidal anti-inflammatory creams such as tacrolimus or pimecrolimus [11]. Tier 2 treatments are for patients with moderate disease. Treatment options include hydroxychloroquine, low-dose oral antibiotics, or acitretin [11]. Tier 3 treatments are for patients with severe disease and include cyclosporine, prednisone, or mycophenolate mofetil [11]. Topical or intralesional corticosteroids, as well as phototherapy and superficial radiotherapy, can be used [2, 12]. A recent case study demonstrated the use of tacalcitol before and during photodynamic therapy for follicular mucinosis to remove follicular hyperkeratosis, reduce inflammation, and enhance the penetration of 5-aminolevulinic acid into the skin for effective photodynamic therapy [13]. Other options include dapsone, isotretinoin,

and minocycline [2, 8, 9]. Janus kinase (JAK) inhibitors such as the pan-JAK inhibitor, tofacitinib, are a recent novel treatment option for lymphocytic cicatricial alopecias [11, 14]. These treatments have variable success rates and placebo-controlled trials have yet to be conducted. Spontaneous remission may occur after months or years [4, 6]. Alopecia may or may not be permanent depending on the damage to follicular stem cells and the degree of mucin deposition in the pilosebaceous units [15].

Key Points

- Alopecia mucinosa is a rare type of lymphocytic primary cicatricial alopecia that can be associated with mycosis fungoides or other malignancies and inflammatory conditions.
- Alopecia mucinosa presents as follicular papules most commonly on the scalp and eyebrows that can lead to scarring alopecia.
- Treatment of alopecia mucinosa includes intralesional or topical corticosteroids, antibiotics, immunosuppressive medications, phototherapy, and superficial radiotherapy with variable success rates.

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