Unmasking the Great Imitator: A Clinicopathological Challenge

Anubha Dev, Kittu Malhi, Debajyoti Chatterjee[1], Dipankar De

Departments of Dermatology, Venereology and Leprology, [1]Histopathology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

A 40-year-old female presented with mildly itchy reddish scaly lesions on her scalp and ear for 1 year. The lesions on scalp were associated with pain and pus discharge. She also reported mild shortness of breath. General and systemic examination were normal. On cutaneous examination, erythematous scaly plaques with central atrophy and whitish scales were present on the fronto-parietal area of the scalp extending onto the forehead [Figure 1a]. These lesions were associated with loss of hair follicles suggesting cicatricial alopecia. Similar lesions were present on the helices and the concha of the ears. On dermoscopic examination of the scalp lesions, central areas of atrophy were observed, along with arborizing telangiectasias, overlying whitish scales, and follicular plugging [Figure 1b]. Multiple smooth, erythematous to skin-colored papules of size 2–3 mm were also present on the nape of the neck [Figure 1c].

Two 3.5 mm punch biopsies were obtained from the erythematous plaque on the frontal scalp and a papule on the neck. On histopathological examination, pan-dermal naked granulomas, composed of epithelioid histiocytes and foreign body giant cells, with scant lymphocytic cuffing were observed in perivascular, periadnexal, and perineural locations [Figure 2]. Reticulin stain showed black reticulin-rich granulomas in the dermis. Ziehl-Neelsen stain and Fite-Faraco stain were negative.

Other investigations like complete blood count, serum calcium, inflammatory markers, angiotensin-converting enzyme levels, and chest X-ray were within normal limits. Anti-nuclear antibody was negative. High-resolution computed tomography scan of the chest, however, revealed mediastinal lymphadenopathy along with randomly distributed soft tissue nodules.

What is the diagnosis?

Sarcoidosis.

Discussion

Sarcoidosis is a multi-system granulomatous disease, affecting the lungs, lymph nodes, eyes, and skin.[1] Cutaneous involvement occurs in 20–35% of all sarcoidosis patients,[1] and these lesions can have a variety of morphologies ranging from papules and plaques to the rarer ichthyosiform and hypopigmented variants.[1]

Being one of the "great imitators," its various atypical presentations often pose a challenge. Sarcoidosis of the scalp is relatively uncommon and can present as both cicatricial and non-cicatricial alopecia, erythematous to atrophic plaques that extend into the forehead and crusting.[2] A very rare presentation of scalp sarcoidosis is lesions mimicking discoid lupus erythematosus (DLE), associated with scarring alopecia.[3] Such lesions may exhibit characteristic features of DLE at classical sites like central atrophy, peripheral hyperpigmentation, and follicular plugging, making a clinical differentiation from DLE very difficult.[4] The papular lesions that were observed on the neck in our patient are not generally observed in DLE. Such lesions, however, are commonly observed in sarcoidosis. Even though the scalp lesions in our patient resembled DLE, a classical histopathological picture of non-caseating naked granulomas[5] was able to help us clinch the diagnosis. Bronchoscopic biopsy was also done subsequently and revealed compact epithelioid cell granulomas just beneath the bronchial epithelium, without any necrosis or acid-fast bacilli. She was treated with oral prednisolone (40 mg), oral hydroxychloroquine (300 mg/day), and topical fluticasone, betamethasone and

How to cite this article: Dev A, Malhi K, Chatterjee D, De D. Unmasking the great imitator: A clinicopathological challenge. Indian Dermatol Online J 2024;15:564-5.

Received: 03-Jul-2023. **Revised:** 06-Dec-2023. **Accepted:** 24-Dec-2023. **Published:** 23-Apr-2024.

Address for correspondence:
Dr. Dipankar De, Department of Dermatology, Venereology and Leprology, Postgraduate Institute of Medical Education and Research, Chandigarh, India. E-mail: dr_dipankar_de@yahoo.in

Access this article online

Website: https://journals.lww.com/idoj

DOI: 10.4103/idoj.idoj_516_23

Quick Response Code:

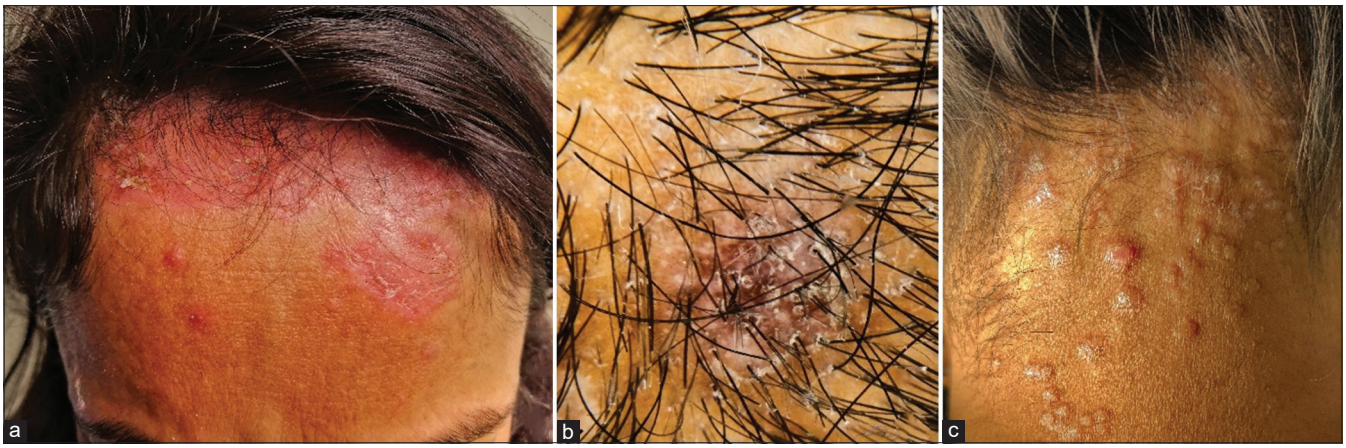

Figure 1: (a) Well-defined erythematous scaly plaques with central atrophy and whitish scales with overlying cicatricial alopecia on the fronto-parietal area of the scalp extending onto the forehead. (b) Dermoscopy of the lesions over the scalp shows areas of atrophy, follicular plugging, and perifollicular scaling. (c) Multiple erythematous to skin-colored papules of size 2–3 mm were observed over the nape of the neck

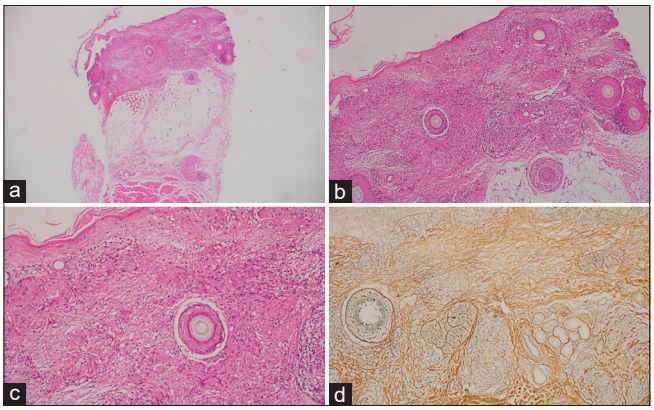

Figure 2: (a) Histopathological examination revealed orthokeratosis along with flattening of rete ridges. Well-formed granulomas are observed throughout the dermis (H&E, 4×). (b) On high-power examination, pan-dermal ill-formed naked granulomas are seen with scant lymphocytic cuffing in a periadnexal, perineural, and perivascular location (H&E, 10×). (c) The granulomas are composed of epitheloid histiocytes admixed with occasional foreign body giant cells (H&E, 40×). (d) Reticulin stain demonstrates black reticulin-rich granulomas (Reticulin stain, 40×)

tacrolimus. She reported improvement in erythema and scaling after five months of therapy and is currently being followed up regularly.

Other aspects that might help us to differentiate between the two entities are elevated serum calcium and ACE levels; pulmonary involvement in the form of hilar and mediastinal lymphadenopathy or parenchymal infiltration; and involvement of other systems like eyes, upper airways, heart, and central nervous system. Owing to the deep nature of the cutaneous infiltration, topical therapies alone are often inadequate in management.[3] This form of sarcoidosis is largely known to be treatment-resistant.[2] Improvement is seen with a combination of oral steroids, antimalarials, and other immunosuppressives.[3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Fernandez-Faith E, McDonnell J. Cutaneous sarcoidosis: Differential diagnosis. Clin Dermatol 2007;25:276-87.
2. Katta R, Nelson B, Chen D, Roenigk H. Sarcoidosis of the scalp: A case series and review of the literature. J Am Acad Dermatol 2000;42:690-2.
3. Henderson CL, Lafleur L, Sontheimer RD. Sarcoidal alopecia as a mimic of discoid lupus erythematosus. J Am Acad Dermatol 2008;59:143-5.
4. Chong WS, Tan HH, Tan SH. Cutaneous sarcoidosis in Asians: A report of 25 patients from Singapore. Clin Exp Dermatol 2005;30:120-4.
5. Bhattacharjee R, Chatterjee D, De D. A man with infiltrated plaques on the pretibial area. JAMA Dermatol 2018;154:955-9.