International Journal of Research in Dermatology
Kamble P et al. Int J Res Dermatol. 2025 Nov;11(6):533-536
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20253399

Case Report

An uncommon presentation of scalp sarcoidosis: window to systemic sarcoidosis

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Received: 09 July 2025
Revised: 07 August 2025
Accepted: 12 August 2025

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ABSTRACT

Sarcoidosis is a multisystem disease with obscure etiology, slight female preponderance and mainly affects lungs, lymph nodes, skin and eyes. Cutaneous involvement is polymorphic, seen in 25-30% of patients and represents tip of an iceberg of systemic involvement. Scalp sarcoidosis though common, it rarely presents as sole manifestation. Though histopathology and reticulin staining serve as confirmatory, full body examination, pulmonary function test and blood examination including serum angiotensin converting enzyme and calcium levels, ophthalmological examination and tuberculosis testing is necessary. We present such an unusual case of scalp sarcoidosis which found to be a window to a systemic disease.

Keywords: Scalp sarcoidosis, ACE levels, Naked granuloma

INTRODUCTION

Sarcoidosis is a multisystem granulomatous disease of unknown origin characterized by hyperactivity of the cellular immune system. It frequently involves the lungs, lymphatic system, and skin.[1]

Cutaneous lesions develop in 25-35% of patients with systemic sarcoidosis and are considered "great mimickers" due to their myriad presentations.[2] Scalp sarcoidosis although rare, presents with important associations that are relevant to diagnosis and treatment. We present a case of scalp sarcoidosis with uncommon presentation, found as a window to systemic disease.

CASE REPORT

A 46-year-old male presented with an asymptomatic, non-progressive, skin-colored raised lesion with central umbilication on the right parietal scalp for 2 years (Figure 1 A). His personal and family history was unremarkable except for long-lasting complaints of occasional breathlessness (Grade 2). On cutaneous examination, a solitary, firm, non-tender, greyish plaque of 1.5×1.5 cm with central umbilication and indurated border was present on the right parietal scalp (Figure 1 B). Deep dermal tenderness was negative. General examination revealed tender non-matted lymph nodes of around 2cm diameter over bilateral post auricular and cervical region. A dermoscopy of the lesion showed arborizing vessels, structureless whitish areas with fibrous streaks, a few broken hairs with follicular plugs, and whitish scaling. A differential of discoid lupus erythematosus and cutaneous sarcoidosis were considered.

Histopathology from the indurated border demonstrated, diffuse non-caseating granulomas composed of epithelioid histiocytes and multinucleated giant cells with scarce lymphocytes and inflammatory cells at the periphery (naked granuloma) extending up to mid-dermis (Figure 2 A-C). Reticulin staining demonstrated peripheral condensation as well as permeation of the granulomas by reticulin fibres (Figure 2 D-F). On

laboratory examination, erythrocyte sedimentation rate, C-reactive protein, serum calcium levels, and calcium concentration in 24-hour urine were within the normal limits.

Angiotensin-converting enzyme (ACE) levels were raised (71.83 U/L; normal,7-25). Mantoux test was negative. On chest roentgenograms and thoracic computed tomograms, bilateral hilar lymphadenopathy was detected. Spirometry done showed a mild restrictive pattern.

Fine needle aspiration cytology of the cervical lymph node revealed granulomatous lymphadenitis.

Based on clinical, radiological, and histopathological grounds, a diagnosis of sarcoidosis was made and patient was started on tab. hydroxychloroquine 200 mg twice daily with topical clobetasol propionate 0.05% cream LABD and intralesional inj. triamcinolone acetonide 10 mg/ml (3 sessions 1 month apart). After 3 months, scalp lesion showed improvement without alopecia (Figure 3).

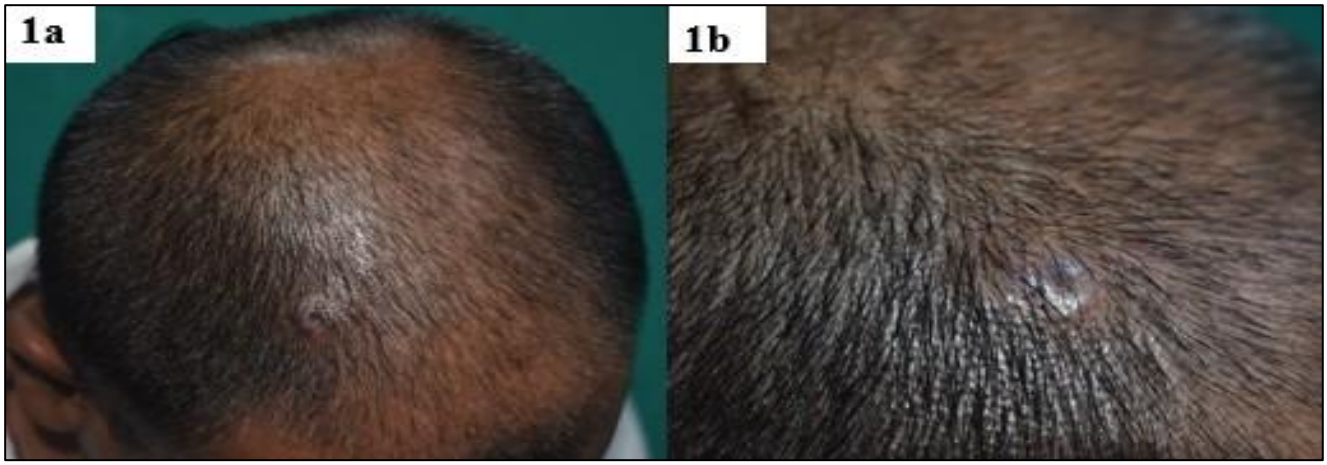

Figure 1 (A and B): Clinical photographs-A-46-years-old male presented with the asymptomatic lesion over scalp. A-A solitary lesion on right parietal scalp. B-An annular, firm, non-tender, greyish plaque of 1.5×1.5 cm with central umbilication and indurated border without alopecia.

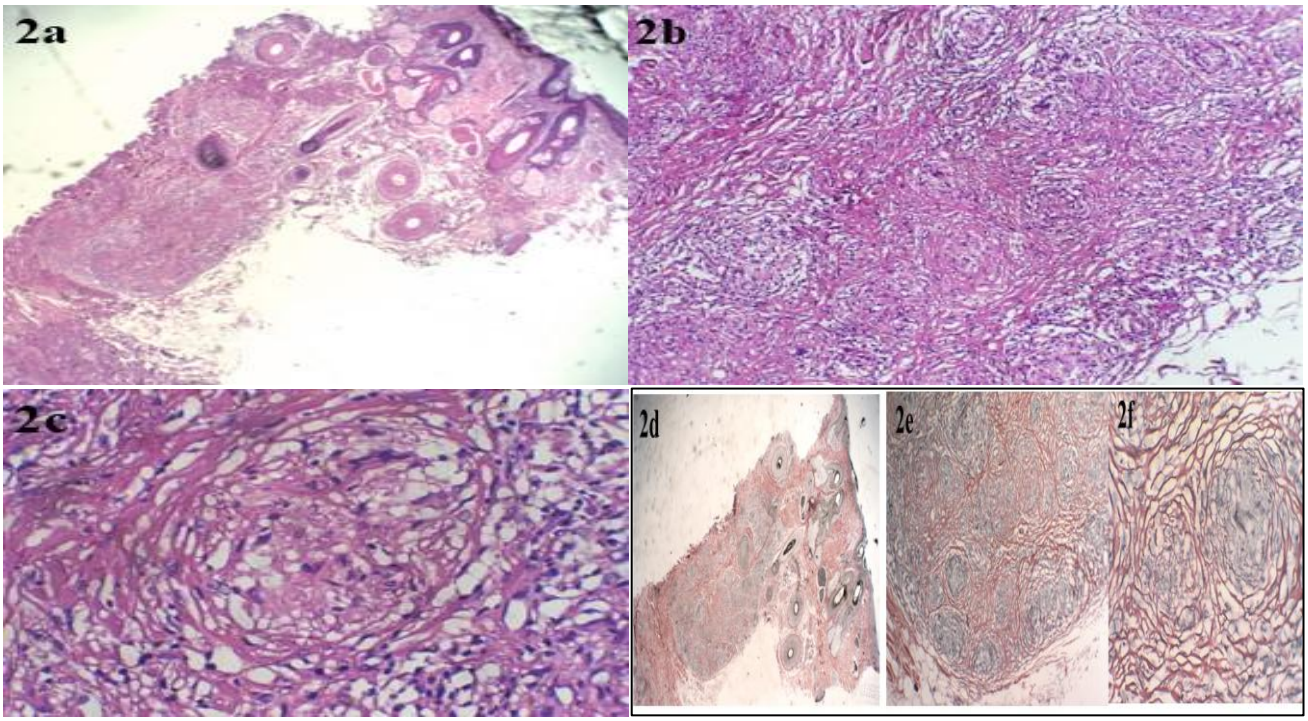

Figure 2 (A-F): Histopathology. A-Diffuse non caseating granulomas extending upto mid dermis, hematoxyline and eosin stain, 40×. B-Granulomas composed of epithelioid histiocytes and multinucleated giant cells, Hematoxyline and Eosin stain, 100×. C-Naked granuloma; scarce lymphocytes and inflammatory cells at the periphery, hematoxyline and eosin stain, 200×. D-F-Reticulin staining demonstrated peripheral condensation as well as permeation of the granulomas by reticulin fibers (40×,100× and 200×).

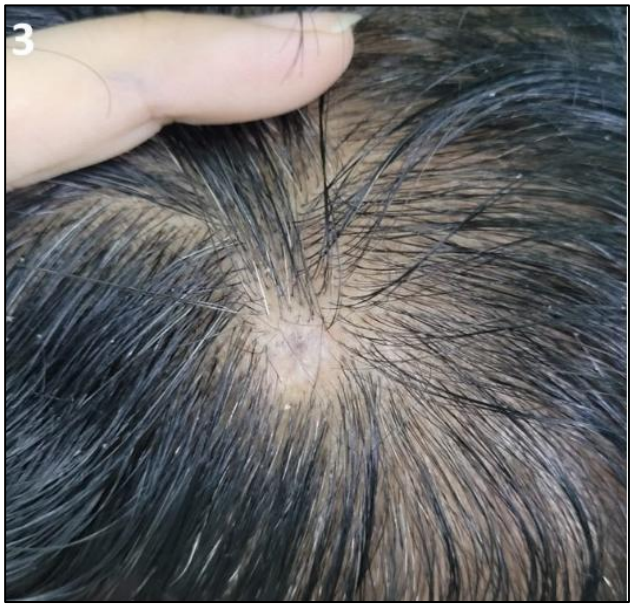

Figure 3: Post treatment photograph-improvement with injection triamcinolone acetonide and hydroxychloroquine.

DISCUSSION

Sarcoidosis is a disease characterized by a wide array of cutaneous and extracutaneous presentations. Hence, its diagnosis requires a compatible clinical picture, histologic demonstration of non-caseating granulomas, and exclusion of other diseases producing a similar clinico-histopathological picture.[3]

Raised serum ACE levels and serum calcium levels depict the activity of the disease and are due to the secretion of ACE by epitheloid cells of granuloma.[6,7]

Macrophages express 1-alpha-hydroxylase enzyme. This causes extrarenal synthesis of the active form of vitamin D in sarcoidal granuloma which leads to increase intestinal absorption of calcium and further increase in serum calcium levels.[4]

Cutaneous lesions of sarcoidosis are divided into specific and non-specific lesions.

Scalp involvement is considered as a specific lesion, but an uncommon one.

It may occur as a discoid lupus erythematosus, necrobiosis lipoidica, atrophic patch with erythema, and scaling, where histopathology plays an important confirmatory role.

Alopecia is common in sarcoidosis, which may be of scarring or non-scarring variety depending on the destruction of hair follicles by non-caseating granulomas. However, our patient did not have alopecia.

Scalp sarcoidosis is rarely the only cutaneous manifestation of sarcoidosis.

It is generally associated with cutaneous involvement at other sites in up to 21 of 23 cases. Our patient had no cutaneous involvement except from the scalp.

Unlike cutaneous sarcoidosis in general, which has systemic involvement in 25-35%, scalp sarcoidosis is associated with a high incidence of systemic sarcoidosis in 20 of 22 cases, as evidenced in our case.

Dermoscopy of the scalp sarcoidosis in previously described cases has shown perifollicular or follicular orange spots along with prominent telangiectasia.[5]

There is limited data for the treatment of scalp sarcoidosis which aims to suppress proinflammatory cytokines and chemokines responsible for granuloma formation. These options include the use of intralesional and oral corticosteroids and other immunosuppressive agents such as azathioprine, and antimalarials like hydroxychloroquine.[5]

Our case highlights the unique presentation of scalp sarcoidosis as a solitary umbilicated plaque without any other cutaneous lesions having high likelihood of systemic involvement. Though sarcoid is commonly associated with scarring as well as non-scarring alopecia, it may have normal hair. Negative tuberculin sensitivity is known feature of sarcoidosis and important in TB endemic country like us where there is high index of suspicion.[8,9] Hence a dermatologist should be aware of the myriad presentations of cutaneous sarcoidosis for early diagnosis and screening for systemic involvement and prompt treatment of this condition.[10]

CONCLUSION

This case highlights the significance of considering sarcoidosis in the differential diagnosis of unusual scalp lesions, even when hair is preserved and there are no other cutaneous manifestations. The presentation as a solitary umbilicated plaque is rare and may easily be overlooked or mistaken for other dermatological conditions. In tuberculosis-endemic countries, the presence of a negative tuberculin sensitivity test serves as an important supportive feature, aiding in distinguishing sarcoidosis from other granulomatous disorders. Given the high likelihood of systemic involvement in such cases, dermatologists should maintain a high index of suspicion and undertake thorough systemic evaluation. Early recognition and prompt management are essential not only for controlling cutaneous disease but also for preventing potential morbidity from unrecognized systemic sarcoidosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kamble P, Gole P, Choithani P, Mahajan S. An uncommon presentation of scalp sarcoidosis: window to systemic sarcoidosis. Int J Res Dermatol 2025;11:533-6.