9-22-2020

Crusted, ulcerated plaques on the scalp and face

Alison Tisack

Angela J. Jiang

Jesse Veenstra

CED
Clinical and Experimental Dermatology

CPD

Crusted, ulcerated plaques on the scalp and face

A. Tisack,[1] A. Jiang[2] and J. Veenstra[2]

[1]*Wayne State University School of Medicine, Detroit, MI, USA; and* [2]*Department of Dermatology, Henry Ford Health System, Detroit, MI, USA*

doi: 10.1111/ced.14441

Clinical findings

A 67-year-old black man presented with a progressively itchy, red and scaly rash that first began on the face and spread to involve the scalp. Prior treatment with ketoconazole 2% shampoo and hydrocortisone 2.5% cream had been ineffective. The patient had a history of hypertension and heart failure with preserved ejection fraction. Physical examination revealed extensive erythematous plaques with overlying thick, yellow to brown crust on the scalp and temporal face, extending to the nose, cheeks and conchal bowls in a seborrhoeic distribution (Fig. 1a,b). Affected areas of the scalp displayed prominent alopecia. A punch biopsy of the cheek and scalp was performed.

Histolopathological findings

Examination of the biopsy revealed a dense infiltrate of histiocytes and noncaseating granulomatous inflammation throughout the dermis (Fig. 2a,b). Periodic acid–Schiff and Ziehl–Neelsen stains were negative.

What is your diagnosis?

Correspondence: Dr Angela Jiang, MD, Department of Dermatology, Henry Ford Health System, 3031 West Grand Boulevard, Ste 800, Detroit, MI 48202, USA
E-mail: ajiang2@hfhs.org

Conflict of interest: the authors declare that they have no conflicts of interest.

Accepted for publication 4 August 2020

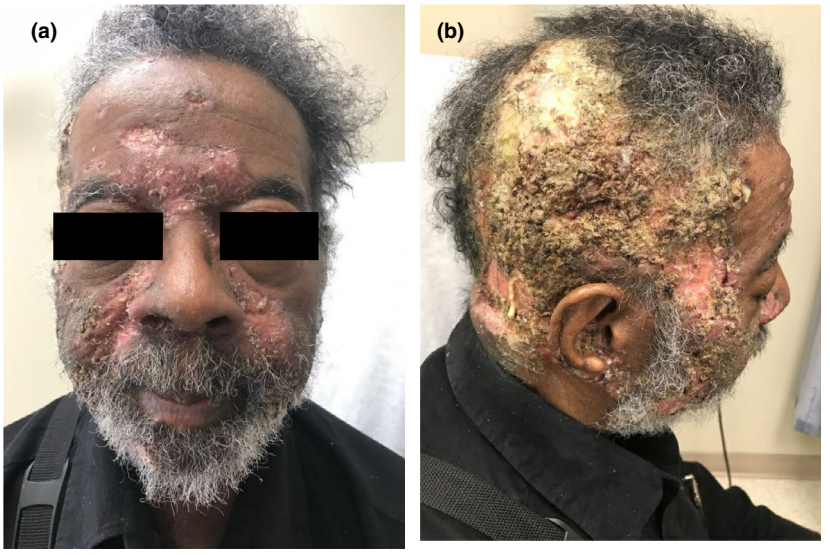

Figure 1 (a) Crusted plaques in a seborrhoeic distribution on the face with overlying yellow crusting; (b) extensive erythematous plaques with thick yellow to brown crusting on the scalp with associated alopecia.

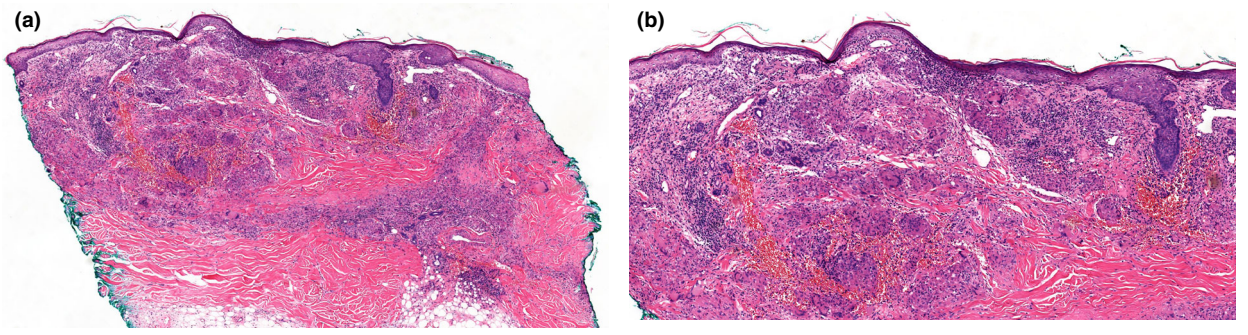

Figure 2 (a, b) Punch biopsy from scalp showing a dense infiltrate of histiocytes and noncaseating granulomatous inflammation throughout the dermis. Haematoxylin and eosin, original magnification (a) × 40; (b) × 100.

Diagnosis

Ulcerative sarcoidosis (US).

Discussion

This patient was diagnosed with US and admitted to the hospital shortly thereafter due to several new-onset syncopal episodes. Electrocardiography demonstrated right bundle branch block and cardiac magnetic resonance imaging showed severe asymmetrical hypertrophy of septal walls. These findings together raised the possibility of cardiac sarcoidosis. Further investigations demonstrated mediastinal and hilar adenopathy, subpleural and perifissural nodularity of the lungs, and posterior uveitis, confirming sarcoidosis involvement of the lungs and eyes. During his 7-day admission, the patient had worsening kidney function with suspicion for renal sarcoidosis, although renal biopsy had yet to be completed. Owing to concern about possible arrhythmia prompting his syncopal episodes and a risk for sudden cardiac death, the patient was started on tapered oral corticosteroid 100 mg and instructed to use a wearable cardioverter–defibrillator. For his cutaneous symptoms, triamcinolone 0.1% ointment was started. Both the cutaneous and systemic symptoms began to improve over the course of the following 2 weeks while the patient was on corticosteroids, to the point that he declined further use of the wearable cardioverter–defibrillator. He was subsequently lost to follow-up by any of the speciality clinics (dermatology, cardiology, ophthalmology and pulmonology).

Sarcoidosis is a multisystem, chronic granulomatous disorder of unknown aetiology. About 20–35% of patients with sarcoidosis develop cutaneous manifestations, and in approximately a third of such patients, the skin is the earliest affected organ.[1] Cutaneous sarcoidosis commonly appears as red–brown to violaceous papules, plaques or nodules along the eyelids, nasolabial folds, neck or upper trunk.[2] This presentation is common in patients with acute sarcoidosis, defined as a sudden onset of cutaneous findings along with hilar lymphadenopathy and acute uveitis. For > 60% of patients, lesions will resolve within 2–5 years.[2] However, US, which is defined by ulcerated, crusted and atrophic cutaneous plaques, is less likely to resolve without treatment.[3] This variant is seen in ~5% of patients with cutaneous sarcoidosis, with a predilection for black and female patients.

Alopecia secondary to sarcoidosis is rare, with most cases affecting the scalp.[4] Sarcoidosis-induced alopecia is most often cicatricial in nature, though nonscarring variants have also been described.[4] Of patients with sarcoidosis-induced alopecia, 75% have additional cutaneous involvement and 85% have concomitant systemic involvement.[4] Owing to the increased risk for systemic manifestations, patients with sarcoidosis-induced alopecia should undergo a thorough physical examination, investigating both cutaneous and systemic signs.

Systemic sarcoidosis commonly affects multiple organcs, including the skin, lung, heart and eye, but to varying degrees. Cardiac sarcoidosis is a rare and potentially life-threatening finding, seen in only 5% of patients with sarcoidosis, with high-grade atrioventricular (AV) block or tachyarrhythmias in a minority of these cases.[1,5] The initial cardiac finding may be sudden death, making cardiac evaluation early in the disease course critical. Pulmonary involvement is common, occurring in 90–95% of sarcoidosis cases, presenting with dyspnoea, chest tightness and wheezing.[2] Ocular involvement occurs in up to half of all cases, most commonly manifesting as anterior uveitis.[1] Common symptoms include blurry vision, eye redness and eye pain; however, patients without ocular symptoms should still be regularly evaluated by an ophthalmologist as ocular damage leading to permanent blindness may occur even in asymptomatic patients.

On histopathology, sarcoidosis classically presents with noncaseating granulomas with surrounding lamellar hyaline collagen.[1] These granulomas consist of macrophages and epithelioid cells surrounding lymphocytes. suggesting systemic reaction to an unknown antigen. On the scalp, fibrosis of hair follicles may be noted, depending on disease progression, similar to other forms of cicatricial alopecia.[5]

Learning points

- US presents as ulcerations within crusted, atrophic plaques.
- Alopecia secondary to sarcoidosis is rare, but affected patients are likely to have further systemic involvement and require more thorough investigations.
- Cardiac sarcoidosis is rare, but can present with high-grade AV block, tachyarrhythmias or even sudden cardiac death.
- The mainstay of sarcoidosis treatment is steroids, with systemic administration preferred over topical for treatment of sarcoidosis-induced alopecia.

References

1 Haimovic A, Sanchez M, Judson MA, Prystowsky S. Sarcoidosis: a comprehensive review and update for the dermatologist: part II. Extracutaneous disease. *J Am Acad Dermatol* 2012; **66**(): 719.e1–10, quiz 729–30.
2 Haimovic A, Sanchez M, Judson MA, Prystowsky S. Sarcoidosis: a comprehensive review and update for the dermatologist: part I. Cutaneous disease. *J Am Acad Dermatol* 2012; **66**(): 699.e1–18, quiz 717–18.
3 Yoo SS, Mimouni D, Nikolskaia OV *et al.* Clinicopathologic features of ulcerative-atrophic sarcoidosis. *Int J Dermatol* 2004; **43**: 108–12.
4 House NS, Welsh JP, English JC 3rd. Sarcoidosis-induced alopecia. *Dermatol Online J* 2012; **18**: 4.
5 Kandolin R, Lehtonen J, Airaksinen J *et al.* Cardiac sarcoidosis: epidemiology, characteristics, and outcome over 25 years in a nationwide study. *Circulation* 2015; **131**: 624–32.
6 Ungprasert P, Ryu JH, Matteson EL. Clinical manifestations, diagnosis, and treatment of sarcoidosis. *Mayo Clin Proc Innov Qual Outcomes* 2019; **3**: 358–75.

CPD questions

Learning objective

To demonstrate knowledge of the demographics of sarcoidosis and the systemic effects of the disease.

Question 1

You diagnose a patient with cutaneous sarcoidosis. Which other system is most likely to be involved?

(a) Cardiac.
(b) Ocular.
(c) Pulmonary.
(d) Renal.
(e) Hepatic.

Question 2

You diagnose a patient with cicatricial alopecia and biopsy the area. The results are consistent with sarcoidosis. Which treatment is most likely to improve the alopecia?

(a) Intralesional steroids.
(b) Topical steroids.
(c) Oral steroids.
(d) Topical calcineurin inhibitors.
(e) Methotrexate.

Instructions for answering questions

This learning activity is freely available online at http://www.wileyhealthlearning.com/ced

Users are encouraged to

- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures.
- Reflect on the article.
- Register or login online at http://www.wileyhealthlearning.com/ced and answer the CPD questions.
- Complete the required evaluation component of the activity.

Once the test is passed, you will receive a certificate and the learning activity can be added to your RCP CPD diary as a self-certified entry.

This activity will be available for CPD credit for 2 years following its publication date. At that time, it will be reviewed and potentially updated and extended for an additional period.