Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.elsevier.com/locate/mjafi

Case Report

Lupus pernio

Lt Col Deepak Vashisht^{a,*}, Lt Col Prashant Sengupta^b,
Lt Col Naresh Bansal^c

^a Graded Specialist (Derm & Ven), 92 Base Hospital, C/o 56 APO

^b Classified Specialist (Pathology & Nephropathology), AH R&R, New Delhi, India

^c Graded Specialist (Medicine), Military Hospital, Jalandhar Cantt, Punjab, India

ARTICLE INFO

Article history:

Received 3 February 2012

Accepted 16 May 2012

Available online 14 September 2012

Keywords:

Sarcoidosis

Lupus pernio

Turkey ears

Introduction

Lupus pernio is a cutaneous manifestation of chronic multi-system sarcoidosis. Besnier's description of a case in 1889, reporting association between reddish-blue lesions of the face and nose with swellings of the fingers was probably the first description of sarcoidosis.¹

Sarcoidosis represents a granulomatous disease of obscure etiology. Although cutaneous manifestations of this disease were recognized as early as 1877, it was later demonstrated that sarcoidosis was a systemic disorder involving visceral tissues as well.²

Recognition of cutaneous lesions is important as they are easily accessible and provide a clue to the diagnosis.

Case report

58-year-old male patient presented with gradually increasing diffuse swelling of ear (Lt) of 20 years duration. It started as a solitary asymptomatic reddish nodule behind the ear. Gradually swelling increased to involve the entire ear and periauricular region. There was no h/o loss of weight, fever, cough, breathlessness, palpitations, chest pain, eye complaints, arthritis or arthralgia, or urinary symptoms. No h/o trauma, discharge from the lesion, loss of sensation or weakness of limbs. He had been earlier treated for lupus vulgaris and deep fungal infection with no relief of symptoms.

Dermatological examination revealed diffuse violaceous plaque involving the entire ear (Lt) and surrounding periauricular region. Ear was grossly enlarged and showed mild scaling (Fig. 1). Similar lesions were also present on (Lt) nape of neck and (Lt) parieto-temporal region of scalp. There was no hypesthesia or peripheral nerve thickening.

Investigations revealed normal hemogram with slightly raised ESR. Mantoux test showed 5 mm of induration. Liver function and renal function tests were normal. Chest X-ray showed no abnormality. Serum calcium and ACE levels were normal.

Skin biopsy taken from nape of neck and pre-auricular region showed scattered, discrete, non-caseating, epithelioid granulomas (Fig. 2). These granulomas were naked i.e. they lacked lymphocytes in the periphery. Granulomas also consisted of giant cells (Fig. 3); it spared the DEJ and adnexa. ZN stain showed no AFB organism (Fig. 4). Lupus pernio was

* Corresponding author. Tel.: +91 9878618878 (mobile).

E-mail addresses: its_deepak75@rediffmail.com, deepak3975@gmail.com (D. Vashisht).

0377-1237/\$ – see front matter © 2012, Armed Forces Medical Services (AFMS). All rights reserved.

<http://dx.doi.org/10.1016/j.mjafi.2012.05.004>

diagnosed based on typical clinical presentation resulting in Turkey ears and typical histopathology and absence of features of cutaneous tuberculosis.

Patient was treated with oral prednisolone, starting at 30 mg daily tapered over 3 months and pulse methotrexate 15 mg/weekly for 6 months along with topical cream clobetasol dipropionate 0.05% twice daily, to which he showed satisfactory response.

He responded favorably to the treatment. Size of the pinna has reduced to near normal size with preservation of its anatomical morphology (Fig. 5). He is on regular OPD follow up and continues to have progressive response.

Discussion

Sarcoidosis is a chronic granulomatous disorder. Skin lesions give the dermatologists advantage to diagnose this condition quite early.

It is a multi-systemic disorder and most often affected organs are: “lungs – 90%, liver – 60%, skin – 25%, eyes – 25%, upper respiratory tract – 20%, parotid glands – 6% of cases”.³ Incidence of systemic sarcoidosis varies from 6 to 10 patients per 100,000 of the population; females are more affected than males. The highest incidence of sarcoidosis is 64 persons per 100,000 population in Sweden.⁴

Sarcoidosis can have specific or non-specific cutaneous lesions. Non-specific lesions lack granulomas microscopically e.g. erythema nodosum (EN). Whereas specific lesions show

Fig. 1 – Grossly enlarged ear (Lt).

Fig. 2 – HE ×40 showing scattered discrete granuloma. No necrosis.

non-caseating epithelioid cell granulomas histologically, and includes papules, nodules, plaques, lupus pernio, and subcutaneous lesions. There may be massive enlargement of ear lobes (“turkey ears”) as seen in this case.⁵ Lupus pernio is a disfiguring skin lesion that is difficult to treat.

The etiology of sarcoidosis is unknown. About 50%–75% of patients with sarcoidosis have depression of T-lymphocyte function and increase in the B-lymphocyte activity (high levels of IgA & IgG).⁶

Diagnosis of sarcoidosis relies on the physical examination, chest X-ray; the Serological testing in the form of raised angiotensin converting enzyme, however it is raised in approximately 60% of the patients with active sarcoidosis.⁴ Biopsy of the clinically suspicious lesions is the most accurate reliable tool.⁷

Sarcoidosis with multiple systems involvement responds well to systemic corticosteroids or methotrexate. Lupus pernio has been reportedly treated successfully with hydroxychloroquine (antimalarial drug) and high potency topical steroid.¹ Carbon dioxide laser remodeling of lupus pernio has been reported in four patients with reasonable success.⁸

Fig. 3 – HE ×400 showing a giant cell.

Fig. 4 – ZN ×400 showing absence of AFB.

Fig. 5 – Pinna reduced to near normal size.

Newer treatment modalities include biologics like infliximab and adalimumab.⁹

In this case report we present a patient with progressively enlarging ear size for past 20 years. He had been treated in the past for lupus vulgaris and deep fungal infection with no relief of symptoms.

The present case is interesting in view of rare presentation of lupus pernio as elephantine ear and for the good response to tapering doses of oral corticosteroids along with pulse methotrexate.

Conflicts of interest

All authors have none to declare.

REFERENCES

1. Besnier E. Lupus pernio de la face: synovites fongueuses symétriques des extrémités supérieures. *Ann Dermatol Syphiligr.* 1889;10:333–336.
2. Kuznitsky E, Bittory A. Sarkoid mit Beteiligung innerer Organe. *Münch Med Wochensh;* 1915;1349–1359.
3. Teirstein AS, Padilla ML, De Palo LR, Schilero GJ. Sarcoidosis mythology. *Mt Sinai J Med.* 1996;63:335–341.
4. Lazarus AA. Sarcoidosis. *Otolaryngol Clin North Am.* 1982;15:621–633.
5. Burns T, Breathnach S, Cox N, Griffiths C. *Rook's Textbook of Dermatology.* Chapter 61. Sarcoidosis. 8th ed. Wiley-Blackwell; 2010:61.1–23.
6. Casuccio JR, Yanagisawa E. Diseases of obscure etiology: sarcoidosis, wegener's granuloma. *Otolaryngol Clin North Am.* 1981;14:331–345.
7. Miglets AW, Vaill JH, Kataria YP. Sarcoidosis of the head and neck. *Laryngoscope.* 1977;87:2038–2048.
8. O'Donoghue NB, Barlow RJ. Laser remolding of nodular nasal lupus pernio. *Clin Dermatol.* 2005;31:27–29.
9. Judson Marc A. Successful treatment of lupus pernio with adalimumab. *Arch Dermatol.* 2011;147:1332–1333.