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Original Research Paper

Dermatology

LUPUS VULGARIS-THE UNSHACKLING OF FENRIR- A CASE SERIES

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ABSTRACT **Background: -** Lupus Vulgaris is the most common form of cutaneous tuberculosis and accounts for 75% of the cases of cutaneous tuberculosis and 1-2% of all tuberculosis cases. The paucibacillary nature of the disease in some instances, makes it extremely difficult to diagnose, even when histopathology and amplification techniques like Polymerase chain reaction (PCR) are employed. Dermatologists must also be aware of the protean clinical manifestations of this condition as its often misdiagnosed when the index of suspicion is low. **Case Series: -** We report a series of five cases of lupus vulgaris among which three have atypical presentation which mimicked Hansen's disease, Discoid lupus erythematosus, Seborrheic dermatitis. Each case was diagnosed with the aid of clinical, laboratory reports and histopathological examination correlation. **Discussion And Conclusion: -** Lupus Vulgaris is arguably a great mimic, as it can present in a myriad of forms. Strong clinical suspicion, histopathology and response to anti-tubercular therapy may be needed for diagnosis especially when culture and polymerase chain reaction assays are not available or non-contributory. The association of lupus vulgaris and its various presentations with systemic tuberculosis make it essential to have a clear understanding of this condition and early diagnosis and management is the key to prevent the disfiguring and destructive nature of this disease.

KEYWORDS : Lupus Vulgaris, Anti-Tubercular Treatment, Mantoux Test, CBNAAT, Skin Biopsy.

BACKGROUND: -

Lupus Vulgaris is the most common form of cutaneous tuberculosis and accounts for 75% of the cases of cutaneous tuberculosis and 1-2% of all tuberculosis cases. It was first described by Erasmus Wilson in 1865, who compared the clinical appearance of the lesions to the ravages of a wolf. [1]

The incidence of cutaneous tuberculosis in India, has fallen from 2% to 0.1%. Yet, the literature is replete with reports of unusual manifestations of this disease many of which are close mimics of other common dermatoses in the tropics. [2]

The paucibacillary nature of the disease in some instances, makes it extremely difficult to diagnose, even when histopathology and amplification techniques like Polymerase chain reaction (PCR) are employed. [3] Dermatologists must be aware of the protean clinical manifestations of this condition as its often misdiagnosed when the index of suspicion is low. It is important to diagnose Lupus vulgaris early as it can be disfiguring and destructive.

Here, we report a series of five cases of lupus vulgaris among which three have atypical presentation.

Case 1: -

A 43-year-old female presented with raised, erythematous skin lesion on the left buttock and posterior aspect of left thigh (Figure.1) for 1 year which was insidious in onset and gradually progressive with a tendency for scarring. She had no history of fever, loss of appetite, cough, tingling or loss of sensation.

On clinical examination, a well to ill-defined erythematous plaque measuring 15x12 cm showing multiple areas of atrophy was seen. Preliminary work up revealed anaemia, hypoproteinaemia, and elevated Erythrocyte Sedimentation Rate (ESR).

Considering possibility of borderline tuberculoid leprosy, deep fungal infections, and lupus vulgaris, skin biopsy was done which revealed dermis showing collection of epithelioid cells, multinucleated giant cells, plenty of lymphocytes and plasma cells (Figure.2,3).

Mantoux test was positive. Cartridge- based nucleic acid amplification test (CBNAAT) of the lesion was negative. The patient was started on Anti-Tubercular Treatment (ATT) after which the patient showed good response. (Figure 4)

Figure.1:-

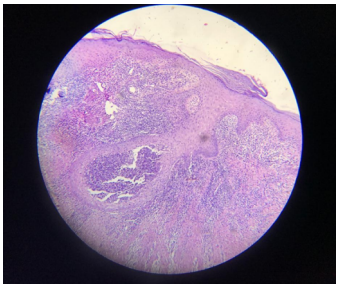

Figure.2:- Dermis showing collection of epithelioid cells,

multinucleated giant cells, plenty of lymphocytes and plasma cells.

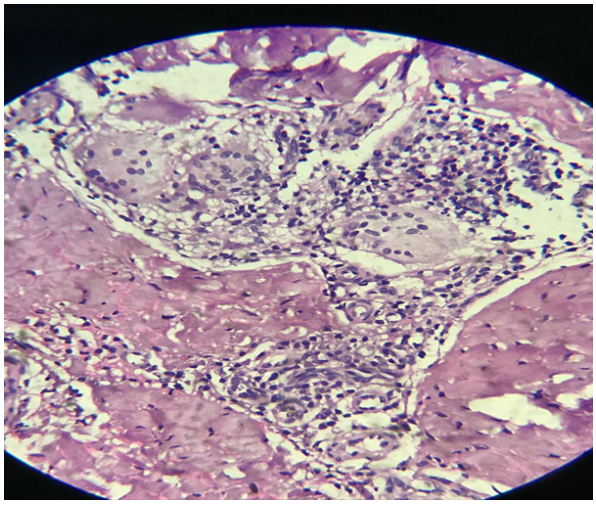

Figure 3. Epithelioid cells, Multinucleated giant cells.

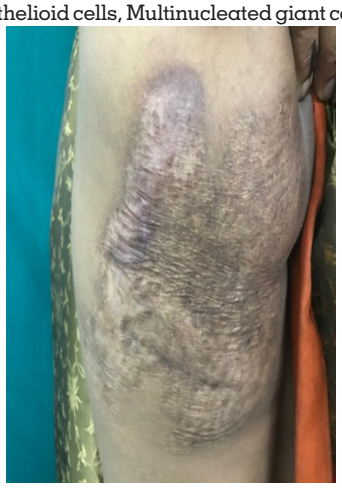

Figure 4. Good clinical response after starting Anti-tubercular Treatment.

Case 2:-

A 62-year-old female presented with erythematous skin lesions over the right upper eyebrow, bilateral cheeks, and dorsum of nose (Figure 5) for the past 3 years which were insidious in onset and gradually progressive.

She had no history of fever, photosensitivity, oral ulcers, and joint pains.

Preliminary workup revealed an elevation in ESR.

Considering the possibility of borderline tuberculoid leprosy, discoid lupus erythematosus, skin biopsy was carried out which showed features suggestive of lupus vulgaris (Figure 6,7) and the patient was started on ATT.

Figure 5

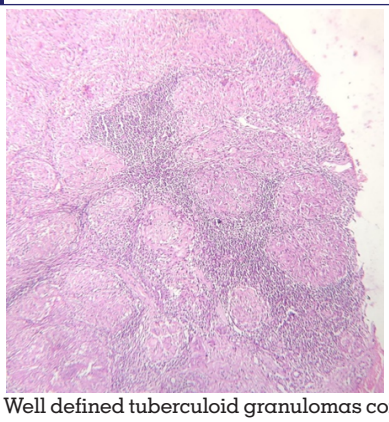

Figure 6: - Well defined tuberculoid granulomas composed of Langhans giant cells, mature epithelioid cells with dense lymphocytic infiltrate and plasma cells

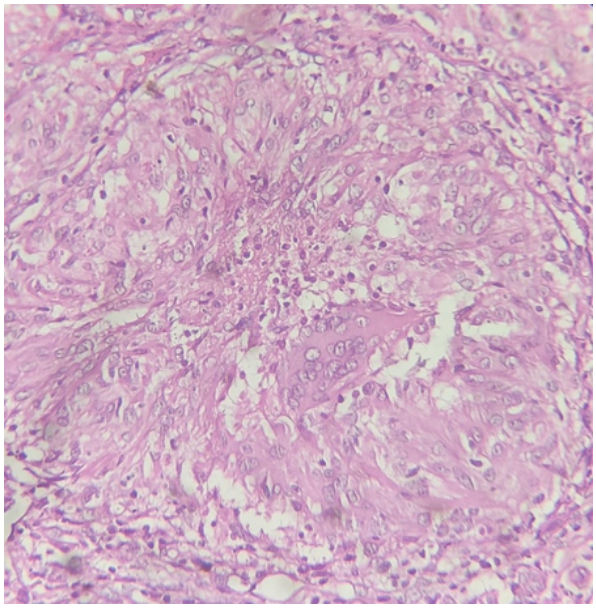

Figure 7: - Langhans giant cells, mature epithelioid cells.

Case 3: -

A 13 year old male presented with asymptomatic scaly lesions over the face. On examination 2 well demarcated scaly erythematous to skin coloured plaques were seen over the face of which one was towards the medial aspect of left eyebrow 1 x 1 cm and the other on the left cheek over the malar prominence measuring 1.5 x 2.5 cm. (Figure 8)

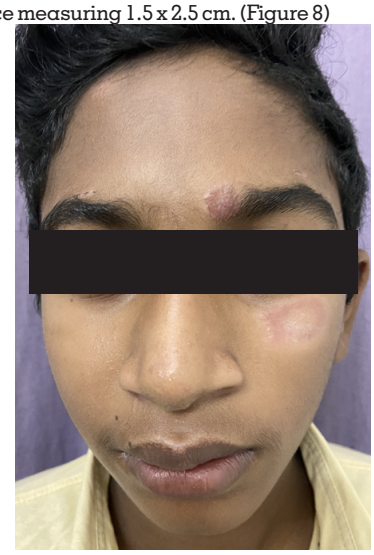

Figure 8

There was no history of discharge, trauma, loss of sensations, fever, paraesthesia, loss of weight, cough and atopy. Sensations over the lesions were intact.

Preliminary workup revealed elevated ESR. Considering the possibility of Hansen's disease, Seborrhoeic dermatitis and Sarcoidosis, skin biopsy and Slit Skin Smear was done which was negative. On skin biopsy (Figure 9,) upper and mid dermis shows granulomas comprising of epithelioid cells, langhans type giant cells, lymphocytic rich infiltrate. Subepithelium shows multiple hair follicles,sebacous glands, fibrocollagenous tissue.

Mantoux positive – 15mm. CBNAAT was negative. Patient was given ATT and showed good response. (Figure 10)

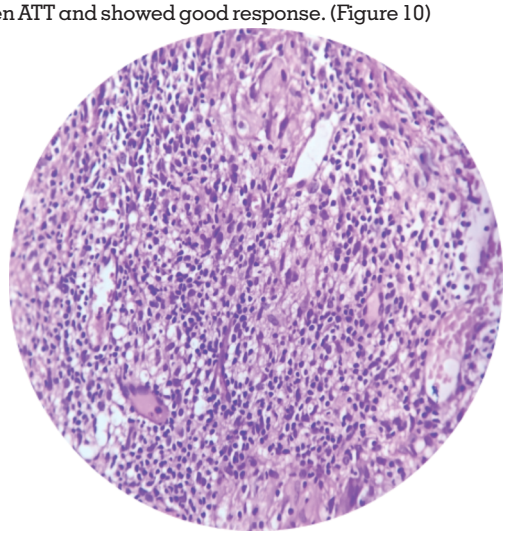

Figure9:- Upper and mid dermis shows granulomas comprising of epithelioid cells,langhans type giant cells, lymphocytic rich infiltrate.

Figure 10. Respone To Anti-tubercular Treatment One After Its Initiation.

Case 4: -

A 41 year old male presented with raised erythematous plaque over the face since 8 months which was insidious in onset with a tendency for scarring and associated with itching. No history of fever, cough, tingling, loss of sensation.

On examination two well to ill-defined erythematous plaque measuring 2x3cm was noted over the chin and a 1x1cm lesion over the philtrum.(Figure 11)

Preliminary work up revealed elevated ESR. CBNAAT-negative.

Considering the possibility of Sarcoidosis, Granuloma annulare and Lupus vulgaris, skin biopsy (Figure 12) was done which revealed keratinised stratified squamous epithelium showing focal atrophy. Superficial dermis shows fibro collagenous tissue and chronic inflammatory cell infiltrate. Mid dermis shows multiple granulomas comprising of epithelioid cells, multinucleated giant cells and lymphocyte infiltration suggestive of lupus vulgaris.

Mantoux test was positive. Patient was given ATT and showed good response. (Figure 13)

Figure 11

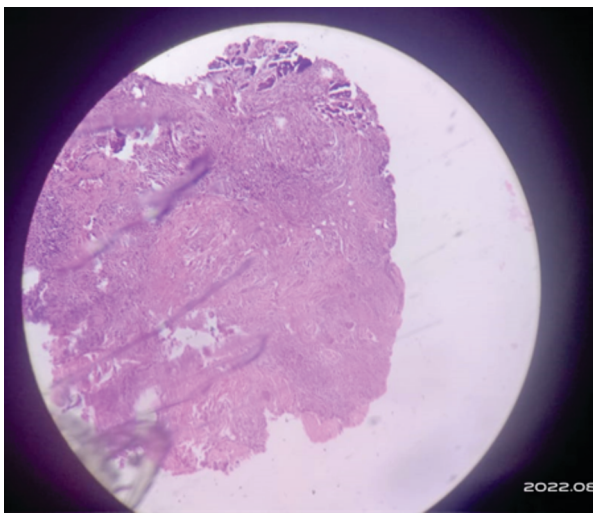

Figure12:- Keratinised stratified squamous epithelium showing focal atrophy. Superficial dermis shows fibro collagenous tissue and chronic inflammatory cell infiltrate. Mid dermis shows multiple granulomas comprising of epithelioid cells, multinucleated giant cells and lymphocyte infiltration suggestive of lupus vulgaris.

Figure 13:- Response one month after initiation of Anti-Tubercular Treatment.

Case 5: -
A 12 year old female presented with asymptomatic lesion over her right knee since 5 years which was insidious in onset and gradually progressive with a tendency for scarring. No history of fever, loss of appetite, cough, loss of sensations or tingling.

On examination – a well to ill defined erythematous to skin coloured plaque measuring about 6x8 cm was noted over the anterior aspect of right knee with hyperkeratosis and scaling on one side and wrinkling on the other side and areas of atrophy was seen. (Figure 14)

Preliminary workup revealed CBNAAT negative. Considering the possibility of Lupus vulgaris, skin biopsy was done which revealed mild hyperkeratosis, focal parakeratosis and spongiosis. Exocytosis of lymphocytes with plenty of lymphocytic infiltrate was seen in superficial dermis along with few epithelioid cells and giant cells suggestive of granulomatous dermatitis. (Figure 15)

Mantoux test was positive. Patient was started on ATT and showed good response. (Figure 16)

Figure 14

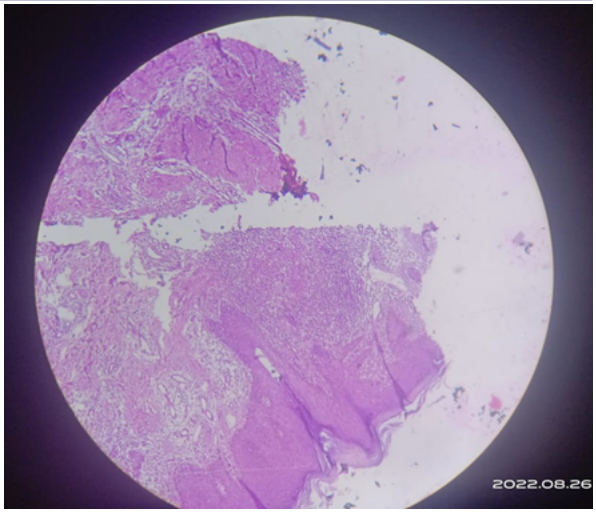

Figure15:- Exocytosis of lymphocytes with plenty of lymphocytic infiltrate was seen in superficial dermis along with few epithelioid cells and giant cells suggestive of granulomatous dermatitis.

Figure 16:- Clinical response 10 days after starting Anti-Tubercular Treatment.

DISCUSSION: -

Lupus vulgaris is a cutaneous tuberculosis seen in individuals who had previously encountered with tuberculous bacilli and those with moderate and high tuberculin sensitivity.[4,5]

It can manifest via endogenous (hematogenous/ lymphogenous) route, exogenous infection on BCG vaccination site, scrofuloderma scar or direct spread from tuberculosis infection. Characteristic lesions are reddish brown plaques spreading to periphery leaving a scar tissue in centre.[6]

We presented 5 cases of Lupus vulgaris some of which mimicked other dermatological conditions. The protean manifestation of the disease,[7,8] the lack of easily available confirmatory tests and the multitude of diseases presenting as chronic scarring skin lesions could be reasons for low level of detection and frequent late diagnosis.

Early diagnosis and prompt treatment helps in preventing disfiguring and destructive lesions. Conventional culture may be negative due to sparse bacilli in the lesion. Hence, histopathological examination plays a key role in diagnosis.

CONCLUSION:-

Lupus vulgaris, arguably a great mimic can present in a

multitude of forms.

Strong clinical suspicion, histopathological diagnosis and response to ATT are sufficient to diagnose this condition especially when culture and PCR are not available or non-contributory. The various clinical forms and its association with systemic tuberculosis make it essential for one to have a clear understanding of Lupus vulgaris.

Today, when tuberculosis threatens to burst into a serious health issue, early diagnosis and treatment are more important than ever to control and prevent morbidity.

Statements and Declarations: - Compliance with Ethical Standards

Ethics Approval: -

This is an observational study. The Kamineni Institute of Medical Sciences Research Ethics Committee has approved the ethics clearance for this observational study.

Informed Consent: -

All the patients in this case series gave informed consent for participation in this study and publication of their medical data , including their images and investigations done. Informed consent was obtained from all individual participants included in the study. Informed consent was obtained from legal guardians in cases where the patients were less than 18 years of age.

The authors affirm that human research participants provided informed consent for publication of the images in Figures 1 to 16.

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Conflict Of Interest:- The authors declare that they have no conflict of interest.

REFERENCES:-

1. Behera B, Devi B, Patra N (2010) Mutilating lupus vulgaris of face: an uncommon presentation. Indian J Dermatol Venereol Leprol 76: 199-200.
2. Marcoval J, Servitje O, Moreno A, Jucglà A, Peyrí J (1992) Lupus vulgaris. Clinical, histopathologic, and bacteriologic study of 10 cases. J Am Acad Dermatol 26: 404-407.
3. Mukta V, Jayachandran K. Lung and lupus vulgaris. Lung India. 2011 Apr;28(2):127-9. doi: 10.4103/0970-2113.80327. PMID: 21712924; PMCID: PMC3109836.
4. James W., Berger T., Elston D. *Mycobacterial Diseases from Andrew's diseases of the skin*. 11th. Elsevier; 2011.
5. Tappeiner G. *Tuberculosis, from Dermatology in General Medicine*. 7th. McGraw-Hill; 2008.
6. Yates VM, Rook GAW. Mycobacterial infections. In: T Burns, S Breathnach, N Cox, C Griffiths, eds. *Rook's Textbook of Dermatology*, 7th edn. London: Blackwell Science Limited, 2004: 28.1– 28.39.
7. Gopinathan R, Pandit D, Joshi J, *et al* . Clinical and morphological variants of cutaneous tuberculosis and its relation to mycobacterium species. *Indian J Med Microbiol* 2001; 19: 193– 196.
8. Tappeiner G, Wolff K. Tuberculosis and other mycobacterial infections. In: *Fitzpatrick's Dermatology in General Medicine*. 6th ed. Freedberg IM, Eisen AZ, Wollf K, Austen KF, Goldsmith LA, Katz S, editors. New York: McGraw-Hill; 2003. pp. 1933–49.