ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 11 | November - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PAPULONODULAR MUCINOSIS IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT –A CASE REPORT

Pathology

Dr. G. Nivas* Junior Resident, Dept Of Pathology, Bangalore Medical College And Research Institute, Bangalore, Karnataka, Pincode – 560002 *Corresponding Author

Dr. Dayananda S Biligi Professor, Dept Of Pathology, Bangalore Medical College And Research Institute, Bangalore, Karnataka, Pincode – 560002

ABSTRACT

Papulonodular mucinosis (PNM) associated with systemic lupus erythematosus (SLE) is a primary cutaneous mucinosis, given that its mucin deposition manifests as a clinically detectable lesion. Here we are reporting a case of Papulonodular mucinosis associated with SLE. A 23-year-old male presented with asymptomatic swelling over his face, body, and limbs with erythematous lesions and a burning sensation over his back and hand. On local examination, there are multiple well-defined skin color nodules and papules over the scalp, forehead, and trunk. On microscopic examination of skin biopsy shows dense mucin deposition between collagen bundles with evidence of sparse perivascular mononuclear cell infiltrate. On blood investigation, the Antinuclear antibody (ANA) was found to be positive and the patient was treated with glucocorticosteroids. In conclusion, the PNM is a rare and atypical manifestation of SLE. Differential diagnoses of PNM are other cutaneous mucinosis such as lichen myxedematosus, myxedema, scleroderma and reticular erythematous mucinosis; and other conditions such as lupus profundus, and rheumatoid nodules. Treatment with glucocorticosteroids (topical, intralesional, or systemic), antimalarial agents, retinoids, cyclophosphamide, methotrexate, plasmapheresis, or surgical options, such as dermabrasion, laser, or excision, have been shown to be of benefit.

KEYWORDS

papulonodular mucinosis, Systemic lupus erythematosus, Mucin deposition

INTRODUCTION

Papulonodular mucinosis (PNM) is characterized by abnormal deposition of mucin in the skin. It is also called cutaneous lupus mucinosis or Papular and nodular mucinosis of gold. [1,2] PNM is mostly associated with autoimmune connective tissue disease.[3] Papulonodular mucinosis was associated with SLE in approximately 76% of cases and DLE in approximately 20%.[4] Papulonodular mucinosis can cause the development of raised, often inconspicuous papules and nodules on the upper body and trunk. In individuals with darker skin tones, these lesions may appear more prominent, taking on an erythematous or violaceous hue.[5] The presence of papulonodular lesions in SLE patients coincides with elevated levels of circulating autoantibodies. This temporal association implies a potential causal link, whereby these autoantibodies may induce cutaneous fibroblasts to overproduce glycosaminoglycans, ultimately leading to lesion development.[6] Here we described an unusual case of Papulonodular Muciniosis (PNM) coexisting with Systemic Lupus Erythematosus (SLE), a rare clinical association.

Case Report

A 23-year-old, male presented with asymptomatic swelling over the face, body, and limbs with erythematous lesion and burning sensation over back and hand. On local examination, there are multiple well-defined skin color nodules and papules over the forehead, neck, trunk, and scalp. Figure 1 depicts the lesions.

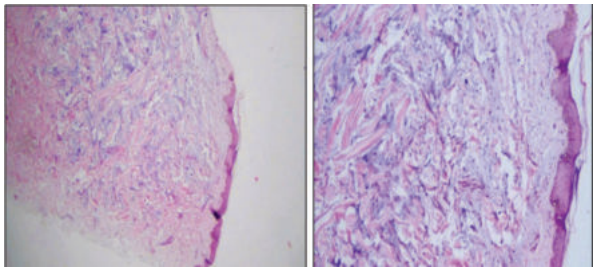
Figure 1- Papulonodular lesions over the forehead, neck, trunk, and scalp

Gross Examination -

A skin-covered 0.7x0.5x0.2cm bit of tissue was received.

Microscopy

The sections studied show skin with an epidermis and dermis. The epidermis appears near normal. The dermis shows dense mucin deposition between collagen bundles, with a sparse perivascular mononuclear cell infiltrate evident. Figure 2 depicts the microscopic features.



H&e, 10x H&e, 40x

Figure 2 – Microscopic features of papulonodular mucinosis

Alcian Blue

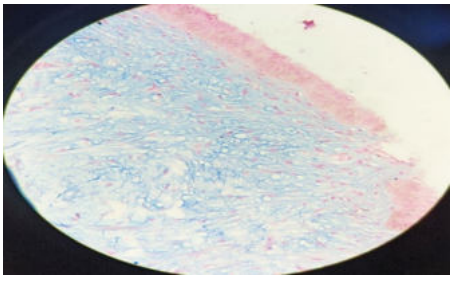


Figure 3 - Alcian blue stain shows mucin deposition in the dermis.

DISCUSSION

Papulonodular mucinosis associated with systemic lupus erythematosus is a primary cutaneous mucinosis, given that its mucin deposition manifests as a clinically detectable lesion. Pathogenesis of papulonodular mucinosis has not been fully elucidated, elevated cytokines and circulating autoantibodies have been noted in cutaneous mucinosis. One of these serum inflammatory markers may possibly increase glycosaminoglycan production. The increase in dermal mucin deposition would result in a clinically observable lesion such as a papule or nodule.

CONCLUSION:

In conclusion, the PNM is a rare and atypical manifestation of SLE. Differential diagnoses of PNM are other cutaneous mucinosis such as lichen myxedematosus, myxedema, scleroderma, reticular erythematous mucinosis; and other conditions such as lupus profundus, and rheumatoid nodules.[2] Treatment with glucocorticosteroids (topical, intralesional, or systemic), antimalarial agents, retinoids, cyclophosphamide, methotrexate, plasmapheresis, or surgical options, such as dermabrasion, laser, or excision, have been shown to be of benefit.[7]

REFERENCES:

1. Dallo C, Lee K, Cragun WC, Lee M. A Case of Papulonodular Mucinosis in a Patient With Systemic Lupus Erythematosus. Am J Dermatopathol. 2020 Apr;42(4):280-282.
2. Sonntag M, Lehmann P, Megahed M, Ruzicka T, Kuhn A. Papulonodular mucinosis associated with subacute cutaneous lupus erythematosus. Dermatology. 2003;206(4):326-9.
3. Ramírez Campo L, Londono˜ Salinas AM, Gómez Gómez LV, Ruíz Suárez AC, Eraso Garnica RM. Mucinosis papulonodular en el inicio de lupus eritematoso sistémico juvenil: asociación inusual. Rev Colomb Reumatol. 2019;26:282–286
4. Hansen E, Pettit C, Chung CG, Korman AM. Papulonodular mucinosis with features of discoid lupus erythematosus. JAAD Case Reports. 2024 Mar 1;45:59-61.
5. Rongioletti F, Rebora A. The new cutaneous mucinoses: a review with an up-to-date classification of cutaneous mucinoses. JAm Acad Dermatol. 1991;24:265-270
6. Pandya AG, Sontheimer RD, Cockerell CJ, Takashima A, Piepkorn M. Papulonodular mucinosis associated with systemic lupus erythematosus: possible mechanisms of increased glycosaminoglycan accumulation. Journal of the American Academy of Dermatology. 1995 Feb 1;32(2):199-205.
7. Fleischmajer R: Papular mucinosis; in Freed berg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Katz SI, Fitzpatrick TB (eds): Fitzpatrick's Dermatology in General Medi cine, ed 5. New York, McGraw-Hill, 1999, pp 2129–2131.