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Case Report

Lichen planus with IgA vasculitis: a rare association

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ABSTRACT

Lichen planus is an inflammatory skin disorder which has been shown to be associated with thyroid disorders, hepatitis B, hepatitis C. IgA vasculitis is a form of cutaneous small vessel vasculitis (CSVV) characterized by palpable purpura, abdominal pain and joint pain usually seen in children. We report a case of 45-year-old female patient came with violaceous itchy lesions which were distributed over trunk and extremities of 1 month duration. Complete blood count (CBC) and serum biochemistry tests were normal. Histopathology of violaceous lesions showed changes of lichen planus. Purpuric lesions showed mild perivascular and transmural inflammatory infiltrate, composed mainly of neutrophils and few lymphocytes in the superficial dermis. Direct immunofluorescence was positive for IgA (granular pattern along with vessel walls of small vessels). Based on clinical, histopathological findings, a diagnosis of lichen planus with IgA vasculitis was made. The patient was treated with 30 mg prednisolone, gradually tapered over 6 weeks and anti-histamines. Satisfactory response was observed at follow-up at 6 weeks. Our case highlights the rare association of lichen planus with IgA vasculitis. Thorough evaluation of the patient helped in reaching correct diagnosis.

Keywords: Lichen planus, IgA vasculitis, Hepatitis-B virus

INTRODUCTION

Lichenoid disorders are inflammatory dermatoses characterized clinically by flat-topped, papular lesions and histologically by a lymphocytic infiltrate with a band-like distribution in the papillary dermis.[1] It is an idiopathic inflammatory skin disease affecting the skin and mucosal membranes, often with a chronic course with relapses and periods of remission. Its prevalence is approximately 0.5% of the population.[2] Lichen planus (LP) is thought to be a T-cell-mediated autoimmune disease, possibly targeting the basal keratinocytes, which can be triggered by a variety of situations, thyroid disorders, viruses (hepatitis B and hepatitis C), drugs and contact allergens.

IgA vasculitis, previously called Henoch–Schönlein purpura (HSP), is an immune complex vasculitis characterized by IgA1- dominant immune deposits affecting small vessels (predominantly capillaries, venules or arterioles). It often involves the skin and gastrointestinal tract, and frequently causes arthritis.[3] It a form of cutaneous small vessel vasculitis (CSVV) characterised by palpable purpura, abdominal pain and joint pain usually seen in children. Patients with IgA vasculitis are typically younger and have more extrarenal manifestations. Association of LP with IgA vasculitis is very rare. We report a case of lichen planus who presented with concurrent lesions of palpable purpura, which on investigation was found to be IgA vasculitis.

CASE REPORT

A 45-year-old female patient came with complaints of violaceous itchy lesions which were distributed over trunk and extremities of 1 month duration and red raised lesions over both upper and lower limbs for 15 days. The patient was apparently normal 1 month ago. Then she developed

violaceous itchy lesions which started over lower limbs. Later, she developed similar lesions over forearm, dorsum of hand and trunk. Later she developed red raised lesions starting over both lower limbs which gradually progressed to both upper limbs and trunk of 15 days duration. Violaceous lesions were associated with itching, while red raised were associated with pain. No history of any drug intake prior to eruption of skin lesions was reported. There was no history of trauma over affected area. There was no history of associated co-morbidities and autoimmune diseases like diabetes, hypertension, autoimmune thyroiditis and connective tissue diseases. There was no history of joint pains, abdominal pain and upper respiratory tract infection.

General physical examination was normal. There was no evidence of any pallor, icterus, cyanosis, clubbing, lymphadenopathy or pedal edema. Examination of respiratory, cardiovascular, central nervous system and gastrointestinal system was normal. On dermatological examination, multiple discrete violaceous papules were seen over the flexors of wrist, upper and lower extremities and trunk (Figure 1). The lesions are closely studded and distributed diffusely over above-mentioned areas. Post-inflammatory hyperpigmentation was seen over dorsum of ankles bilaterally. Koebners phenomenon was seen at wrist, trunk, upper and lower extremities. On magnification, Wickham striae was seen over lesions at wrist. There were no lesions over scalp, palms and soles. Oral and genital mucosa was normal. No nail abnormality was noticed.

Multiple discrete purpuric lesions were seen over bilateral upper and lower extremities (Figure 1). Haemorrhagic crust formation and scaling/desquamation were seen over most of the lesions. Urticarial plaques and smaller purpuric lesions were seen over thighs (Figure 2). Diffuse oedema was seen over dorsum of left hand with overlying purpuric lesions (Figure 3). No large bullae were seen. Livedo reticularis and atrophy/scarring were not present. Purpuric lesions or erosions are not seen over oral and genital mucosa. On diascopy lesions were not blanchable.

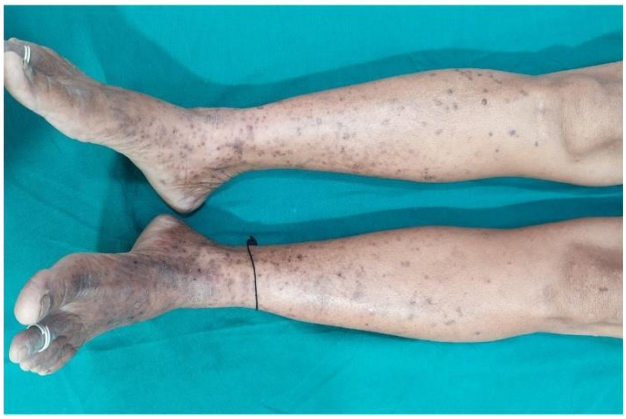

Figure 1: Discrete violaceous papules and purpuric lesions seen over both lower extremities with PIH.

Based on clinical presentation, violaceous papules were clearly suggestive of lichen planus. For the purpuric lesions, cutaneous small vessel vasculitis, HSP, erythema multiforme and livedoid vasculopathy were considered. Patient was subjected to detailed laboratory investigations. Complete blood picture and serum biochemistry profile including random blood sugar (RBS), liver function test (LFT), blood urea and serum creatinine were within normal limits. Viral screening showed HIV, HCV were negative but hepatitis-B surface antigen (HbsAg) was positive.

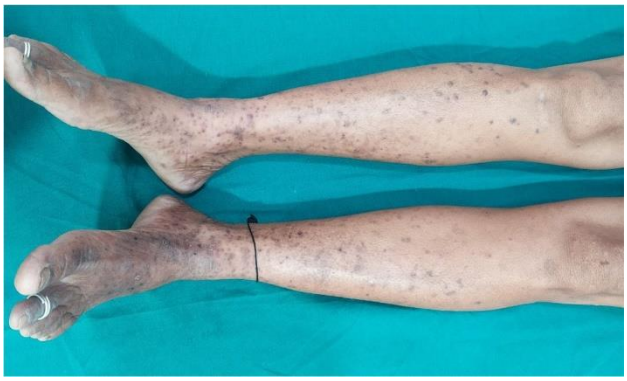

Figure 2: Urticarial plaques and smaller purpuric lesions were seen over thighs.

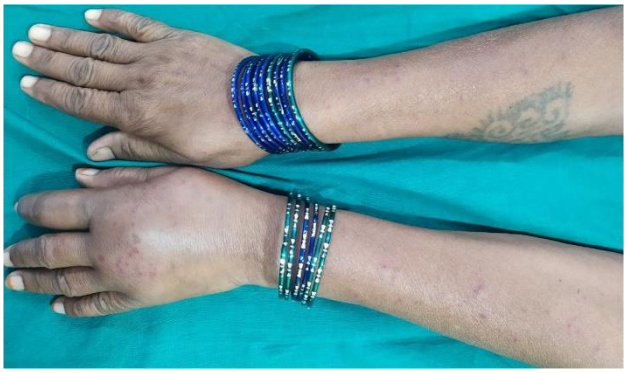

Figure 3: Diffuse oedema was seen over dorsum of left hand with overlying purpuric lesions.

Skin biopsy samples from two lesions were sent for histopathology and direct immunofluorescence. Histopathology of violaceous lesions showed moderate compact hyperkeratosis in stratum corneum, prominent stratum granulosum, mild acanthosis in stratum spinosum and lymphocytic degeneration in stratum basale (Figure 4). Upper dermis showed moderate interstitial lymphohistiocytic infiltrate, which is seen abutting the basal layer of epidermis. Small vessels in the upper dermis show transmural neutrophilic infiltrate and extravasation of RBCs along with melanophages. The above features were suggestive of lichen planus. Histopathology of purpuric lesions showed mild basket weave hyperkeratosis in stratum corneum, stratum granulosum was preserved, mild acanthosis of stratum spinosum and intact stratum basale. Superficial dermis showed mild perivascular and

transmural inflammatory infiltrate (Figure 5), composed mainly of neutrophils and few lymphocytes and moderate extravasation of RBCs. Deep dermis and subcutis were unremarkable. Direct immunofluorescence - positive (+2) for IgA – granular pattern along the walls of small vessels (Figure 6). The above features were suggestive of IgA associated vasculitis.

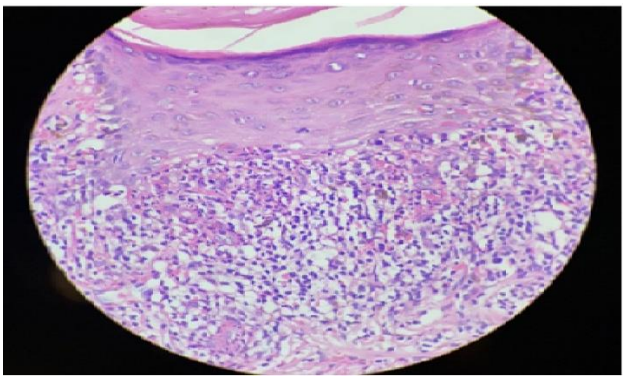

Figure 4: Lichenoid basal layer degeneration.

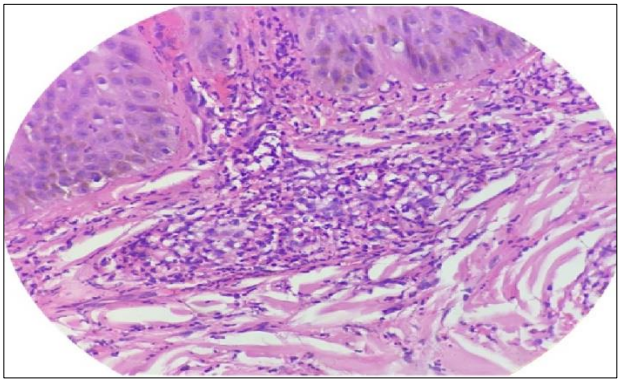

Figure 5: Transmural neutrophilic infiltration.

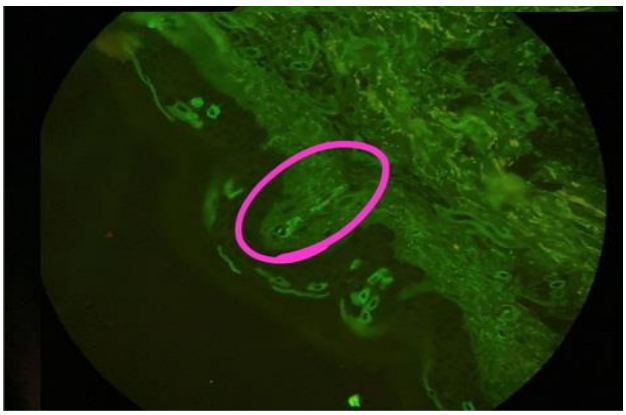

Figure 6: Granular pattern along the walls of small vessels.

Based on clinical, histopathological and direct immunofluorescence findings, a diagnosis of lichen planus with IgA vasculitis was confirmed. The patient was treated with oral steroids (30 mg prednisolone daily for 3 weeks and gradually tapered over 6 weeks), topical clobetasol 0.05% cream twice daily for 4 weeks, tablet dapsone 100 mg daily at night for 4 weeks and anti-histamines. Marked improvement was seen in lichen planus lesions with residual pigmentation. Purpura lesions resolved completely at follow-up at 4 weeks and no new crops of lesions were seen.

DISCUSSION

The lymphocytic infiltrate of LP is composed of CD8+ T cells with a significant proportion of γ-δ T cells, which is unusual in the skin.[1] Gene expression studies of LP have demonstrated an up-regulation of type I interferon-inducible genes and the presence of a specific chemokine signature; CXCL9, the ligand of the receptor CXCR3, was significantly increased in this study.[4] T cells, both CD4+ and CD8+, accumulate in the dermis, whereas CD8+ T cells infiltrate the epidermis. It has been proposed that CD8+ cytotoxic T cells recognize an antigen (currently unknown) associated with major histocompatibility complex (MHC) class I on lesional keratinocytes, and lyse them. Activin A – a transforming growth factor β (TGF-β) family member induced by pro-inflammatory cytokines and which promotes Langerhans cell differentiation and activation – is increased in the upper epidermis and dermis in LP.[5] Mast cell degranulation, and T-cell secretion of matrix metalloproteinase 9 (MMP-9), may contribute to basement membrane disruption, enabling migration of CD8+ T cells into the epithelium. Idiopathic LP has been reported in association with diseases of altered or disturbed immunity, including ulcerative colitis, alopecia areata, vitiligo, dermatomyositis, morphoea and lichen sclerosus, systemic lupus erythematosus, pemphigus and paraneoplastic pemphigus. In addition, LP has been observed in association with thymoma, hypothyroidism, myasthenia gravis, hypogammaglobulinaemia, primary biliary cirrhosis, especially in those treated with penicillamine, and primary sclerosing cholangitis.

Increased levels of IgA in the serum (in 50% with active disease), circulating immune complexes containing IgA, and the deposition of IgA in blood vessel walls and in the renal mesangium are associated with IgA vasculitis. In IgA vasculitis, IgA1 rather than IgA2 is the main IgA subclass deposited in skin lesion.[6,7] Diminished glycosylation of the proline-rich hinge region of the IgA1 heavy chain is thought to be an important factor in allowing the IgA to be deposited in the mesangium and in activating the alternative pathway of complement in IgA, as it makes such IgA1 molecules more prone to forming macromolecular complexes.[8] Other IgA antibodies that occur in HSP include IgA ANCA, although this finding is very variable between studies. IgA rheumatoid factor and IgA anticardiolipin antibodies are also sometimes present, as are IgA anti-endothelial cell antibodies (AECAs). TGF-β is of particular interest as blood levels of T cells that produce this cytokine are increased in HSP, and it is known to enhance IgA1 responses. Neutrophil activation, elevated nitric oxide levels, reactive oxygen species and increased

urinary leukotriene are all documented. Underlying etiology responsible for the development of HSP Include upper respiratory tract infections (streptococcal, RSV, influenza), hepatitis B and seasonal tendency. IgA vasculitis is predominantly seen in childhood and occurrence in adults is uncommon.

Hepatitis B virus (HBV) is a small enveloped DNA virus. The icosahedral nucleocapsid, the virus core, contains the partially double-stranded genomic DNA plus a polymerase that has reverse transcriptase activity. It infects humans causing an acute hepatitis before potentially establishing chronic persistent infection. Chronic infection can lead to cirrhosis and hepatocellular carcinoma resulting in over 600 000 deaths per year worldwide. Skin manifestations can appear in both acute and chronic infection-small vessel vasculitis, urticarial vasculitis, polyarteritis nodosa, serum sickness-like reaction, urticaria, GCS, porphyria cutanea tarda, pruritus, sarcoidosis (with interferon and/or ribavirin therapy, erythema multiforme and erythema nodosum.

Concurrent occurrence of IgA vasculitis with lesions of lichen planus is extremely uncommon with few cases reported in literature. In our case, hepatitis B positivity was detected, which may be responsible for triggering the cell mediated immunity resulting in development of lichen planus. Rarely viral infections give rise to CSVV. It is possible that Hepatitis B infection had a role in pathogenesis of IgA vasculitis. All cases of LP should be thoroughly evaluated to find out other co-morbidities such as autoimmune diseases and chronic disorders which are components of metabolic syndrome like DM, hypertension, hyperlipedemia, and obesity. All patients of IgA vasculitis should be thoroughly evaluated to find out involvement of other visceral organs including renal, inflammation of joints and GI involvement.

CONCLUSION

Our case highlights the rare association of lichen planus with IgA vasculitis. Common trigger factors (hepatitis B infection) could be responsible for eliciting the inflammation in both conditions. Thorough evaluation of the patient helped in reaching correct diagnosis. Complete investigations should be done to rule out associated co-morbidities of both lichen planus and IgA vasculitis.

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