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Rheumatology

IGA VASCULITIS SECONDARY TO LEPTOSPIROSIS: A RARE CASE REPORT

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ABSTRACT IgA vasculitis, formerly called Henoch – Schonlein purpura or HSP, is an inflammation of the small blood vessels in the skin, GI tract & the kidneys. As IgA is driven in response to external stimuli, especially microorganisms, its assumed that various microorganism has the capacity to induce abnormal IgA autoimmune reactions to the host. Herein we report a case of a 48year old female with IgA vasculitis secondary to leptospirosis. The patient presented with fever, purpuric rash over upper & lower limbs, anemia & pedal edema. IgG & IgM were positive for leptospirosis with low complement C4 & high IgA levels. C- ANCA was positive whereas Total ANA profile was negative. Along with treatment of Leptospirosis, patient was started on i.v. steroids, upon which fever subsided as well there was marked improvement in the rash.

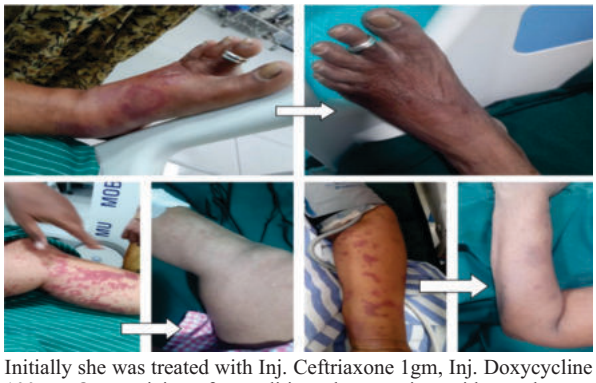
KEYWORDS : IgA vasculitis, leptospirosis

INTRODUCTION -

IgA vasculitis (IgAV) is an inflammation of small vessels caused by perivascular deposition of IgA and activation of neutrophils. It may present as systemic vasculitis (IgAV - Henoch-Schönlein purpura) or as a variant restricted to the skin (skin-limited IgAV), while IgA nephropathy presents a variant restricted to the kidneys. Systemic IgAV affects children more frequently than adults. In the latter, although rare, cases in adults have already been described in a worldwide incidence varying from 3 & 14.3 cases per 1 million inhabitants. The dominant clinical features include round or oval and retiform palpable purpura predominantly on the lower legs, arthralgia or arthritis, gastrointestinal bleeding or pain and glomerulonephritis with mesangial IgA deposits (IgAVN) depending on the variant. Pulmonary, cardiac, genital and neurological involvement occurs, but is rare. In case of self-limited disease, only symptomatic treatment is recommended.

Case Report:

A 48 years old female, with H/O T2DM & HTN presented with complaints of fever for 2 weeks, swelling of bilateral lower limbs along with Rash over bilateral lower, upper limbs & arthralgia of knee, ankle joints for 4 days. On examination, she had pallor, pedal edema, & Erythematous, coalescent & discrete patches & macules on legs, thighs, forearms & feet. Fever workup was done showing leptospirosis IgG & IgM positive. The typical rash was observed to be consistent with that of vasculitis. On further investigations, we observed, microscopic hematuria, proteinuria of nephrotic range (77mg/dl), mild indirect hyperbilirubinemia, thrombocytosis (5.16 lakhs/cumm), anemia (Hb – 7.3), Coombs – mild positive, hypoalbuminemia (3.2). On further investigated we observed low C4 levels (4), high IgA levels (611) & C-ANCA positive as well as negative Total ANA profile. Renal function & parameters were within normal limits with normal urine output.



Initially she was treated with Inj. Ceftriaxone 1gm, Inj. Doxycycline 100mg. On suspicion of vasculitis and supportive evidence she was started on Inj. Methyl Prednisone @ 1mg/kg bd wt. & other supportive management. Later fever subsided & rash improved. Patient was discharged with oral steroid on tapering doses.

DISCUSSION:

In the above case, the patient was clinically diagnosed by the presence of palpable purpura over the upper & lower limbs & presence of arthralgia, hematuria, proteinuria supported with presence of C-ANCA pattern over immunofluorescence indicating vascular deposits of IgA. In clinical practice or large trials, a biopsy is not necessarily always performed & a diagnosis is often made based on clinical features in combination with positive ANCA serology. Blood cultures & Urine Cultures, malaria antigen, triple markers, Antibody for scrub typhus were negative except, the IgG & IgM for leptospirosis were positive. This microorganism possibly triggered the development of IgA vasculitis by creating IgA immunocomplexes and deposition on small vessels.

Kidney involvement is the most important organ dysfunction in determining the therapeutic options for IgA vasculitis and influencing its prognosis. In our case, the patient is having mild IgA vasculitis without marked involvement of kidney or GI tract. IgA vasculitis patients show significant improvement of cutaneous purpura symptoms with the treatment of Prednisolone 1 mg/kg daily.

CONCLUSION:

In this case of Fever presenting with rash of acute onset, it was vital to suspect this rare association of Leptospirosis & IgA Vasculitis depending upon observable clinical and laboratory pearls. This outcome is a wakeup call, warranting a broad perspective into the pathophysiology of any type of rash during examination to prevent possible multiorgan involvement which is evident by the dramatic clinical response to the timely intervention by steroid therapy.