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CHRONIC IDIOPATHIC CUTANEOUS VASCULITIS: A CASE REPORT

Rheumatology

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ABSTRACT

We report a 25-year-old man presenting with cutaneous vasculitis. Cutaneous vasculitis is a rare condition that refers to a type of vasculitis that affects the blood vessels, which includes the arteries, veins or both and it also affects any part of the body. This pathophysiologic condition of the patient is presented clinically as recurrent cutaneous vasculitis, with the patient having absence of any other symptoms apart from swelling of ankle and erupted red spot like rashes all over the body specifically aggressive on the lower limbs. Our report is presented with the entire clinical manifestation of patient, differential diagnosis of patient and the underlying etiology which was determined to be idiopathic. Treatment undergone by the patient for the whole period of recurrences, and diagnosis for any other ailment which could have caused this pathophysiological condition is also elaborately discussed in this article.

KEYWORDS

Cutaneous vasculitis, Purpuric rashes, Blood vessels

INTRODUCTION

Vasculitis is a disease of variable causes in which immunologically mediated inflammatory reaction of the blood vessel wall leads to vessel wall damage and weakening (aneurysm, rupture) or obstruction of lumen, leading to infarction of tissue.[1] It can be primary (without an identifiable cause) or secondary to stimuli such as infections, drugs or systemic autoimmune diseases. It can be limited to the skin or manifest in other organs such as kidney, lung and heart, thereby implying varying prognosis.[2] It can be classified as large vessel type (giant cell arteries, polymyalgia rheumatica, takayasu arteritis), medium vessel (polyarteritis nodosa, Kawasaki disease) and small vessel type vasculitis. Small vessel type includes some of the most common types of vasculitis. They are often divided into the diseases where anti-neutrophil cytoplasm antibodies (ANCA) are found in the blood (ANCA associated vasculitis) and diseases where ANCA is not found. The ANCA associated vasculitides are Wegener's granulomatosis (now called Granulomatosis with Polyangiitis) Microscopic Polyangiitis and Churg-Strauss syndrome (now called Eosinophillic Granulomatosis with Polyangiitis).[3]

As a result the affected person tend to experience swelling, petechiae, itching in affected areas, sensation of pin and needles in petechiae areas, fatigue. Many of the small vessel type of vasculitis can cause damage to the kidneys, the typical symptoms of impaired kidney function would be fatigue, poor appetite, and swollen legs and breathlessness due to retention of fluid. The blood pressure may be very high which can cause damage to organs like kidneys, heart and brain. The presence of blood and protein in urine can indicate inflammation in the kidneys and requires urgent assessment by nephrologist. If not treated at right time, inflammation in the kidneys can cause severe damage leading to dialysis treatment or kidney transplantation. This paper deals with the clinical presentation and management of a case of chronic idiopathic cutaneous vasculitis.

CASE PRESENTATION

A 25 year old male presented with a history of severe knee pain, swollen ankle region associated with red spots on lower limb. He reported to a general physician on 3rd day from the development of pain and rash (Fig.1), the doctor suggested to observe the spread of rashes and prescribed tablet paracetamol 650mg twice daily for 5 days.

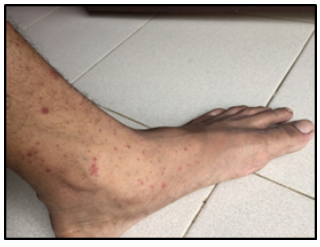

Figure 1 – Initial petechial rashes (1st week)

In between 4th – 7th day the rashes slowly faded away. Two weeks later pain was subsided, but again the rashes were increasing with pricking sensation on lower limbs (Fig.2). As the week progressed rashes were slowly reducing and at the end of 2nd week skin was clear without any rashes and no pain. With no other symptoms, all of a sudden at 3rd week when the patient was working at office on an afternoon, the red spots started appearing again and was seen all over the lower limbs (Fig.3).

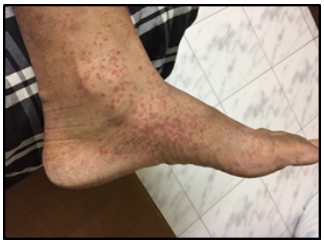

Figure 2 – 2nd Week

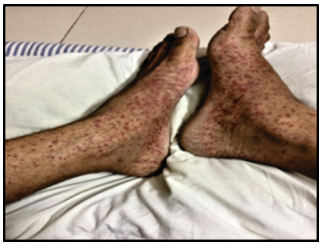

Figure 3 – 3rd Week

So the patient reported to a dermatologist, the findings seen was multiple bilateral erthematous petechial rash in feet upto middle leg with associated pain over the rash, ankle joint pains, no associated swelling. Patient was systemically healthy. On clinical examination, multiple palpable petechial rash, non blanching type was evident. The differential diagnosis was acute erythematous petechial rash secondary to viral illness. Medications prescribed was tablet naproxen 250 mg twice daily for 5 days. On 4th week, the rashes had reduced with no associated joint pain (Fig.4). On reviewing clinically fair improvement was seen, with petechial rash reduced upto 80%, minimal joint pain symptoms, no systemic symptoms and febrile spikes. Impression was secondary cutaneous vascultis due to viral etiology. Injection methylprednisolone acetate 80 mg intramuscularly was given and patient was asked to report back if condition is worsening. On 5th week the patient was referred to rheumatologist, there was few pus filled rashes seen and mild itching present (Fig.5). The patient was prescribed with tablet hydroxychloroquine 200 mg and fexofenadine 180 mg tablets once daily for 7 days, mupirocin 2% ointment for applying on pus filled rashes. Then blood and urine investigations was advised for the patient.

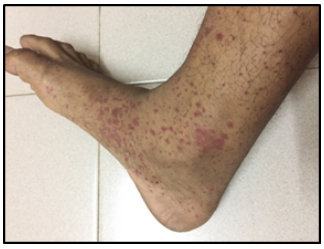

Figure 4 – 4th Week

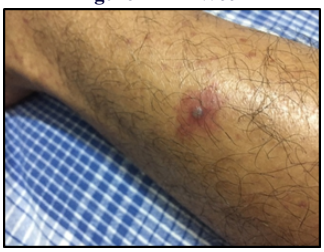

Figure 5 – 5th Week

The complete blood count results are as follows: hemoglobin was 16 g/dl, red blood cell count was 5.39 millions/cumm, packed cell volume was 47.2%, his platelet count 1.98 lakhs/cumm, white blood cell count was 6700 cells/cumm, neutrophils was 58.8%, lymphocytes was 31.5%, monocytes was 5.1%, eosinophils was 3.8%, basophils was 0.8%, erythrocyte sedimentation rate was 13 mm/hour and random blood sugar was 82mg/dl. Dengue profile IgG and IgM antibody, NS1 antigen was non reactive. Serum creatinine was 0.80 mg/dl. Glomerular filtration rate was 125ml/min. Antibody and complement levels are as follows, C4 complement was 22 mg/dl, C3 complement was 112 mg/dl. cANCA and pANCA immunofluorescence tests were negative. C-reactive protein was <6mg/L negative. Antistreptolysin O (ASO) titer value was 228.5 IU/mL which indicates recent group A streptococcus infection which could have been a trigger factor in the patient. Urine analysis was completely normal which showed absence of blood, casts and nil proteins. The petechial rash was seen recurring on weekly cycle and was even more severe after 6 months (Fig.6). Thus, this is a classic case of idiopathic type of cutaneous vasculitis. No biopsy was required. Tablet naproxen 250 mg and montelukast sodium 10mg was taken during acute painful rash attacks. The rashes was seen for a period of one year. After a year there was a complete disappearance of the petechial rash (Fig.7).

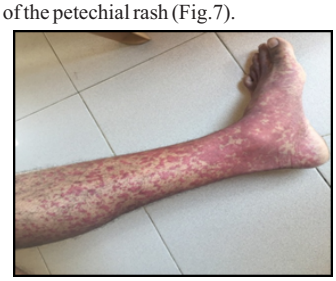

Figure 6 - 6 months

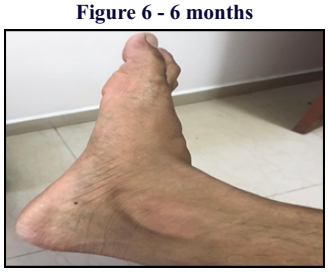

Figure 7 - After one year

DISCUSSION

Cutaneous vasculitis is a abjectly understood condition due to its variable clinical manifestation and it may overlap with various other infections, certain connective tissue disorders and malignancies. The skin is the organ most commonly involved, with typical presentation of a painful, burning rash predominantly in the lower extremities, with up to one-third of patients presenting with trunk and upper extremity involvement.[4] The most common skin manifestation is palpable purpura.[5] Other skin manifestations include maculopapular rash, bullae, papules, plaques, nodules, ulcers, and livedo reticularis. Some may also present with arthralgias or arthritis involving the knees or ankles.[6] In the present case report, we have evaluated the clinical features, relevant past medical history and various laboratory tests to reach a diagnosis of chronic idiopathic cutaneous vasculitis. The clinical presentation in patient was crops of nonthrombogenic palpable purpura, primarily involving dependent areas such as legs, ankles, feet. This is in accordance with studies done by Sais et al [7]and Ekenstam et al[8] where they observed purpura in 89.2 and 62% of cases, respectively. Gastrointestinal involvement in the form of pain in abdomen, diarrhea, frank blood in stools was seen in patient.

Vasculitis has various diagnostic challenges, primarlily cases could present with variable clinical manifestations as a isolated cutaneous vasculitis to multisystem involvement. Several medical conditions may mimic with the presentation of vasculitis, so the range of differential diagnosis is therefore extensive. It may develop as a primary disorder or be secondary to various medical conditions, thus it is essential to differentiate them for proper treatment planning. The main objectives when treating a patient with cutaneous vasculitis is to rule out the systemic organ involvement and followed by identifying the etiology. A meticulous correlation of the patients medical history and the clinical, serological and imaging findings aids in proper diagnosis.

CONCLUSION

vasculitis needs complete and exhaustive approach for its successful management. Proper detailed patient history taking, thorough cutaneous and systemic clinical examination along with relevant laboratory investigations will assist in arriving at diagnosis. Treatment should to be personalized based on intensity of involved cutaneous and systemic pathology. Some types are benign and are self limiting, while someothers have the probablity to affect various vital organ function and patients life. Idiopathic cases are also difficult ones to treat and patients should be continuoulsy monitored to avoid future complications.

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