

From idiopathic thrombocytopenic purpura to systemic lupus erythematosus

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A B S T R A C T

Thrombocytopenia is one of the cardinal features of Systemic Lupus Erythematosus (SLE). It is recognized as the earliest and rarely the sole manifestation of SLE at the time of diagnosis. This report narrates the case of one such patient who had an abrupt onset of life-threatening thrombocytopenia and was subsequently diagnosed as having SLE.

Keywords: Idiopathic thrombocytopenic purpura, systemic lupus erythematosus, thrombocytopenia

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Introduction

SLE is an autoimmune disease in which a person's immune system mistakenly attacks its own tissues itself. It can affect any organ system of the body like skin, brain, cardiovascular system, gastrointestinal tract, kidney and locomotor system and hematological system. We will discuss a case report of severe thrombocytopenia which was due to SLE.

Case report

A 25-year-old single lady from Gujrat, with no previous pre-morbid illness, was admitted in Medical ward of a tertiary care hospital of Islamabad through emergency. Her chief complaints were gums and nasal bleeding for 3 days along with a petechial rash for 01 day. She was taken to a local doctor in Gujrat who gave her Proton Pump Inhibitors and Multivitamin tablets. Two days later she developed generalized petechial rash more prominent on limbs. At this point she was taken to District Head Quarter (DHQ) hospital Gujrat where her blood complete picture showed severe thrombocytopenia. She was

transfused with 06 units of Platelets and then referred to a tertiary care hospital.

When she was seen in our hospital, she had petechial rash all over her body while her nasal and gum bleed had temporarily stopped. However, next day she again developed nasal and gum bleed along with macroscopic hematuria. She did not have any history of fever or any other systemic upsets except history of arthralgia. Her past medical history was insignificant as was her personal and family history. On examination her vital signs were stable. General physical examination only revealed widespread petechial rash on her body most prominent on limbs with no palpable lymph nodes, pallor or jaundice. She had sub-conjunctival hemorrhage in her left eye with normal vision. Systemic examination was also unremarkable.



Figure 1: Petechial rash on legs.



Figure 2: Sub-conjunctival hemorrhage.



Figure 3: Petechial rash on upper limbs.

Keeping her symptoms in view our list of differential diagnosis for the patient included Idiopathic Thrombocytopenic Purpura (ITP), Dengue Fever, Malaria, Hematological malignancy and Systemic lupus erythematosus. Her investigations showed a normal white cell count, Hemoglobin of 10.7 with MCV 76 and markedly low platelets count of 9000. Her electrolytes, liver and kidney function tests were normal as was her urinalysis. Malarial parasite and Dengue fever screening were negative. Chest imaging by X-ray and ultrasound abdomen were also normal. Results of clotting profile and

D-Dimers also came out negative. To evaluate the cause of thrombocytopenia her bone marrow biopsy was performed and report showed a hyperplastic marrow with the impression of peripheral destruction of platelets. At this point a diagnosis of ITP was considered, however, before labeling patient as a case of ITP an autoimmune screen was run along with complement levels. Both ANA (performed by indirect immunofluorescence method) and Anti ds DNA antibodies were strongly positive and complement levels were markedly low – C3= 50 (Ref: 83-140), C4= 5 (Ref: 10-20). Hence, on the basis of these test results, the patient was diagnosed as having SLE.

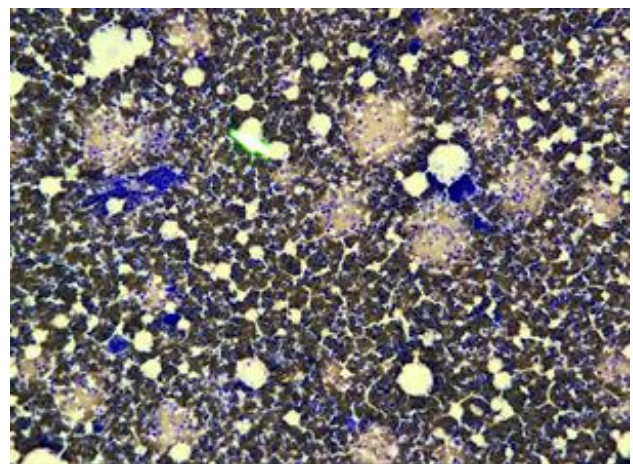


Figure 4: Bone Marrow biopsy – Megakaryocytes can be clearly seen

The patient was transfused with platelets to prevent active bleeding. She was given pulsed Methylprednisolone 1 gram for three consecutive days followed by oral Prednisolone at 1mg/kg body weight. Calcium and Vitamin D supplements were also prescribed to prevent Glucocorticoid induced osteoporosis. She was also given Hydroxychloroquine to prevent flares of her disease. Iron supplement was also started as she had a mild microcytic anemia. Her Platelets began to increase on day 3 and on discharge her blood CP showed a Platelet count of 1, 14,000. She has been advised a regular follow up in outpatient department.

Discussion

SLE is an autoimmune disease involving different organ systems of the body.¹ The spectrum of clinical features in patients of SLE is very wide and varies from a mild skin rash to severe organ or life threatening disease.²

Based on such variable presentation of symptoms it can be easily said that no two patients of SLE are alike. Hematological manifestations of SLE include anemia, thrombocytopenia and low white cell count or a combination of any of these. Thrombocytopenia is one of the criterions mentioned in the American College of Rheumatology (ACR) classification criteria for SLE.³ This case report highlights the importance of screening patients with thrombocytopenia for connective tissues diseases particularly SLE before labeling them as a case of ITP. Traditionally a diagnosis of SLE and other connective tissue diseases is thought of only when a patient presents with a particular set of symptoms. However, from literature review it is clear that sometimes low platelet count may be the only manifestation for some time before development of overt SLE.

The prevalence of thrombocytopenia in patients of SLE has been shown to be 7% to 30% according to studies conducted in the past.⁴ Also it has been seen that thrombocytopenia is associated with increased risk of lupus nephritis, neuropsychiatric manifestations of SLE, hematological involvement and over all a high prevalence of thrombocytopenia in patients of SLE has been shown to be 7% to 30% according to studies conducted in the past.⁴ Also it has been seen that thrombocytopenia is associated with increased risk of lupus nephritis, neuropsychiatric manifestations of SLE, hematological involvement and over all a high disease activity and more chances of end organ damage compared with SLE patients who had normal platelet count.⁵ However, on the other hand these patients are seen to have low prevalence of skin involvement.

Thrombocytopenia is sometimes the sole manifestation of SLE. Such cases are often erroneously labeled as ITP till the time they develop other symptoms of SLE. Studies conducted in early nineties^{6, 7} demonstrated that low platelet count can be one of the earliest manifestations of SLE. Such patients also often have history of arthralgia or arthritis and sometimes presents with skin rash too. It can take between 4 and 14 years for the patients to fully develop manifestations of SLE.⁵ Our patient too gave history of arthralgia involving both knee joints and sometimes small joints of hands. Other than the musculoskeletal system rest of the systems were found to be uninvolved in our patient both

in past and also during her admission in hospital. Literature review also shows that presence of Anti Ro antibodies in patients of thrombocytopenia can be used as a predictor of development of SLE at a later stage.⁷ None of the true ITP patients are positive for this antibody.⁶ Because of affordability issue ENA profile could not be done in our patient to look for Anti Ro antibody positivity but we plan to get it done in future when she comes for follow up visits.

Conclusion

Thrombocytopenia in a young female should not be ignored. SLE should be included in the list of differential diagnosis in all patients presenting with thrombocytopenia. By timely diagnosing SLE effective measures can be taken to prevent life threatening complications of this disease.

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