CASE REPORT

Thrombocytosis in a patient with systemic lupus

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Abstract

Systemic lupus is an autoimmune disease of worldwide distribution. The disease is characterized clinically by multisystem manifestations. Haematological manifestations are diverse. Thrombocytosis has rarely been reported in association with SLE and may occur as a result of active disease or reactive due to underlying inflammatory process. Our patient was a 14 years old female who was diagnosed as having systemic lupus and had thrombocytosis which persisted despite control of the underlying disease with corticosteroids. Persistent thrombocytosis raised the possibility of Hyposplenism which was confirmed by peripheral smears and radiological investigations. Patient was given appropriate vaccinations in order to prevent the risk of sepsis in a hyposplenic patient.

Keywords: Systemic lupus, Thrombocytosis.

Introduction

Systemic lupus is associated with various haematological manifestations either because of the active disease or reactive due to underlying inflammatory process.[1] Thrombocytosis is clinically significant when the platelet count is over 400,000/μl. The causes of thrombocytosis could be multiple such as myeloproliferative disorders ,reactive thrombocytosis, Post splenectomy and hyposplenism.[2] Hyposplenism (or asplenia or autosplenectomy) is defined as under functioning of the spleen which may or not be associated with a reduction in splenic size.[3]

Our patient had thrombocytosis which initially was thought to be reactive but later on persisted despite control of systemic lupus which raised the possibility of other etiologies like hyposplenism. Hyposplenism has been seen in systemic lupus due to vascultis changes[4] in the spleen or because of the associated Anti-phospholipid syndrome causing splenic infarcts.[5]

A case of Hyposplenism and thrombocytosis in systemic lupus is reported. The clinical consequences of hyposplenism specially the risk of infection and the preventive strategies are discussed.

Case Report

A 14 years old female with no significant past medical history was seen in 2012, in medical clinic of King Fahad Hospital, Hofuf, Saudi Arabia with the complaint of joint pains, fatigue and low grade fever since last two months. Patient had been treated symptomatically without much relief . On examination she had painful movements in most of the peripheral joints but no swelling or erythema. There was no evidence of skin rashes, hair loss, lymph adenopathy and hepatosplenomegaly. Laboratory investigations results are shown in (Table-1 and 2)

Patient was diagnosed as a case of systemic lupus and started on oral prednisone with good clinical response. Patient's CBC was persistently showing thrombocytosis which initially was thought to be reactive but it persisted despite a clinical response after starting steroids. Peripheral blood smear showed, Howell jolly bodies, target cells and spherocytes. Ultrasound examination revealed normal spleen. A 99m-technetium sulphur colloid scan and 99m-technetium labeled, heat-damaged erythrocyte scan revealed failure of splenic uptake. The patient was diagnosed as having

Table-1: Complete Blood Count.

Haematology Parameters	Before steroids treat	4 weeks post steroids	8 weeks post steroids
WBC	$6.08x10^9$/l	$11X10^9$/l	$10x10^9$/l
Haemoglobin	10.8 gm/dl	11.5gm/dl	12gm/dl
MCV	68fl	74fl	80fl
Platelets	$599X10^9$	$750x10^9$	$800x10^9$
ESR	60mm/hr	40mm/hr	35mm/hr

WBC: White Blood Cell Count. MCV: Mean Corpuscular Volume. ESR: Erythrocy to Sedimintation Rate.

Table-2: Immunology Profile.

	On initial visit	4 weeks Post steroids	8 weeks Post steroids
ANA	Strongly Positive(1:1240)	Strongly Positive	Strongly Positive
AntiDsDNA	Strongly Positive	Strongly Positive	Weakly Positive
AntiSSA	Positive		
AntiSSB	Negative		
Anti Sm	Negative		
C3&C4 complements	low	normal	normal
Anti-cardioplipin (IgG, IgM)	Borderline High	Positive in High titers	
Lupus anticoagulants	Negative		

ANA: Antinuclear Antibody. AntiDsDNA: Anti Double Stranded Deoxyribonucleic acid. AntiSSA, SSB, Sm are auto antibodies.

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functional hyposplenism secondary to systemic lupus.

In view of the higher risk of pneumococcal infections in hyposplenism, the patient was vaccinated with a polyvalent pneumococcal vaccine and she remained clinically stable until her last follow up in the clinic.

Discussion

Systemic lupus is associated with various haematological manifestations either because of the active disease itself or reactive due to an underlying inflammatory process.[1] Thrombocytosis is considered when the platelet count is over 400,000/μl. The causes of thrombocytosis could be multiple such as myeloproliferative disorders, reactive or secondary thrombocytosis or Hyposplenism or Post splenectomy.[2] Hyposplenism (or asplenia or autosplenectomy) is defined as underfunctioning of the spleen which may or not be associated with a reduction in splenic size.[3] The normal functions of the spleen are reduced to a varying degree in a number of illnesses including systemic lupus.[3] Hyposplenism is characterized clinically by leukocytosis, thrombocytosis, presence of Howell jolly bodies, erythrocytes pits and acanthocytes on peripheral smear. Radiologically non clearance of the sulfur colloid scan is also a feature of Hyposplenism.[3]

Our patient had thrombocytosis which initially was thought to be reactive but later on persisted despite control of systemic lupus which raised the possibility of other etiologies like hyposplenism. Abnormal sulfur colloid scan in addition to conventional ultrasonography along with peripheral smears findings were considered as fairly specific for Hyposplenism as seen in other series,[3,4] so the need for MRI and bone marrow examination was not considered in this patient.

Hyposplenism has been infrequently described in patients with systemic lupus. It is seen in about 5% of the cases and is thought to be caused by vasculitic changes within the spleen[4] and also has been reported in association with Antiphospholipid (APL) syndrome.[5] In a study by Castellino and colleagues[5] Platelet count was evaluated in 465 patients with SLE (387 women, 78 men). Seventeen (3.7%) patients with thrombocytosis were observed out of which 3 (17.6%) were diagnosed as having hyposplenism by characteristic peripheral smear and confirmed later by liver-spleen scans showing absence of splenic uptake (a finding of functional autosplenectomy). One satisfied criteria for Antiphospholipid syndrome (APS), and the other 2 patients had positive IgG Antiphospholipid antibodies (APL) at medium titer. It was concluded by this study that thrombocytosis or even correction of thrombocytopenia in patients with SLE should raise suspicion of hyposplenism or autosplenectomy, in particular if secondary APS is present. Our patient, though not fulfilling the criteria for anti-phospholipid syndrome, had borderline positive Antiphospholipid antibody titer which could be contributing to the development of thrombocytosis.

Hyposplenism is associated with significant risk of serious infections, associated with high mortality, an entity known as overwhelming post splenectomy infection (OPSI). The bacteria causing OPSI are predominantly capsulated bacteria like Pneumococci, Meningococci and Haemophillus.[6]

In hyposplenic patients, like ours, preventive measures are mandatory and it has been shown that multidisciplinary approach involving patient education, anti-pneumococcal vaccination and antibiotic prophylaxis using penicillin V, will considerably reduce the incidence of serious pneumococcal infections in these subjects.[7,8] The commonly used pneumococcal polysaccharide vaccine is ineffective in asplenic subjects, because it requires the presence of IgM memory B cells, and should be given before splenectomy. In hyposplenic subjects, like our patient, the pneumococcal conjugate vaccine is more effective, because it utilizes a T cell dependent mechanism, and should be the preferred vaccine in these circumstances.[9]

Conclusion

Thrombocytosis in a patient with systemic lupus is a serious issue and as recommended by Castellino,[5] that any patient with systemic lupus presenting with persistent thrombocytosis must have blood smear examined by a haematologist as this procedure can be done easily and can be helpful in making an early diagnosis.

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