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CASE REPORT

Systemic Lupus Erythematosus Presenting as Coagulopathy in a Male Child

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Abstract

Systemic lupus erthematosus (SLE) are rare presentations in children and comprise about 10-20% of all cases. SLE is an multisystem disease with wide variety of clinical manifestations and when occur they are most commonly seen in the females but are relatively rare in males. We present here a case report of 14 year old male child with SLE who presented primarily with changes in coagulation pathways.

Key Words

Childhood SLE, Coagulopathy

Introduction

SLE is an episodic and multisystem disease, which is characterized by the presence of Antinuclear antibodies (especially those directed to dsDNA) and of other autoantibodies. SLE causes immunologically mediated tissue damage that can affect every organ but commonly affects the skin, musculoskeletal system and hematological system. [1] On comparing with adult onset SLE, childhood-onset is more severe. [2]

Hematological presentations of SLE includes involvement of cell lineages and alterations in the coagulation system. Because of the rarity of the disease and varying presentations in children, diagnosis may be difficult. Early diagnosis and appropriate treatment is essential to provide a favorable outcome for normal development in both childhood and adolescent.

Case report

An 14 year old developmentally normal male child born to a non consanguineous couple was brought with complaints of multiple joint pain, associated with swelling of both ankle and knee joint with Waxing and waning course for the past one month. He also had associated features of fever, facial puffiness, decreased urine output and reddish blue patches present over the right arm and trunk for past 3 days. History of swelling in the right buttock following an injection 3 days back. No history of epistaxis, hematuria, gum bleeds and malena. No history of similar episodes in the past. No history of jaundice.

On examination, child was conscious, oriented. Vitals were stable. The child had pallor with ecchymotic patches noted over the right arm (3x4cms) and trunk (6x4cms) (*Fig1*), swelling of ankle and knee joint present, multiple petechiae noted in the palate (*Fig 2*) and few purpuric spots noted over face, hematoma noted in the right gluteal region and in left lower limb. Per abdomen shows no organomegaly. Other system are within normal limits. The differential diagnosis were acquired coagulopathy

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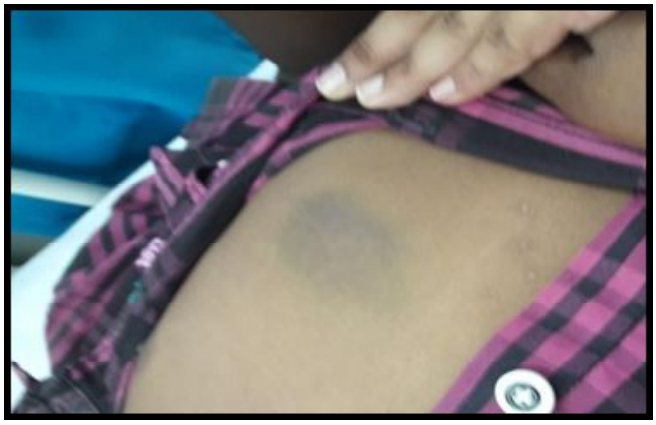

Fig 1. Ecchymotic Patches Seen Over Trunk

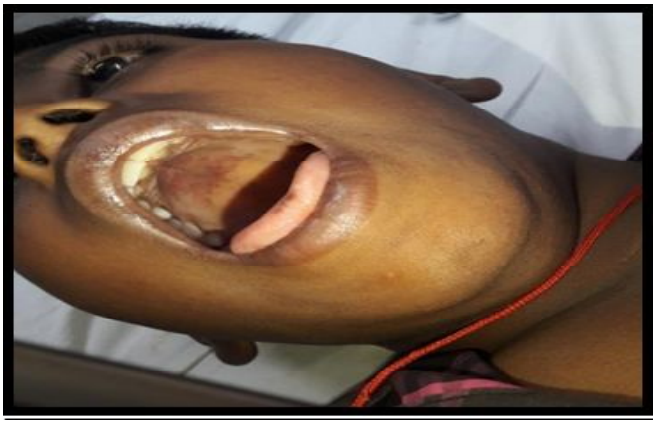

Fig 2. Multiple Petechiae Seen Over Palate

secondary to infection, malignancy and SLE.

Child was investigated CBC shows Hb:8.9gm/dl, TLC:3400cells/mm3 and Platelets:23,000/mm3. Peripheral smear sho ws normocytic normochromic anaemia with thrombocytopenia and leucopenia without evidence of hemolysis/ atypical cells. ESR:130 mm/hour, CRP:2.4mg/dl, Urine routine shows plenty of RBCs, albumin +++, and granular casts, Urea:25mg/dl and Creatinine:0.5mg/dl, SGOT:33 IU/L and SGPT: 29 IU/L, RA factor: negative. In view of fever with thrombocytopenia, Dengue and Scrub typhus done which was negative. As the child had skin bleeds, Coagulation profile done (PT:24 sec, INR:2.1 and APTT:>100 sec) was prolonged however FDP and D dimers done for DIC was negative.

The conclusion of the Investigations revealed that the child had pancytopenia, coagulopathy (increased PT, APTT), thrombocytopenia, renal involvement (albuminuria and hematuria), raised acute phase reactants (Raised ESR and CRP) and multisystem involvement (renal, hematological). possibility of collagen vascular disease was suspected, hence further work up were done which shows urine spot PCR >2mg/dl, ANA and dsDNA were positive with low C3 levels and also Antiphospholipid antibodies was negative . Depending upon the clinical and laboratory findings, the child was diagnosed as a case of SLE . Hence child was given with pulse dose of methylprednisolone and continued with steroid and hyroxychloroquine. Now currently on regular follow up.

Discussion

SLE is an chronic complex autoimmune disease of unknown cause. Pediatric onset represent 10-20 % of all cases. The prevalence of SLE children and adolescents are (1-6/100,000) which predominantly affects females with reported 2-5:1 ratio prior to puberty and 9:1 ratio during reproductive years. [3] Though the cause of SLE remains unknown. Genetic, hormonal, environmental and immunologic factors may play a role. The unique feature of childhood SLE is the identification of monogenic forms of SLE, mainly due to defects in the complement system or abnormal B cell development. [4,5] As the child fulfilled the ACR/EULAR classification criteria SLE 2019; the clinical and laboratory findings that met the criteria were fever, thrombocytopenia, athritis, protienuria, low C3 and anti dsDNA antibody. Total score was 25 (a score of 10 or more fulfilled the criteria for SLE).

Hematological manifestations are commonly noted in patients with SLE. The manifestations are primarly involvement of cell lineages and coagulation abnormalities. Among alteration in cell lineages, anemia contributes to 80-90 % of cases. Anaemia may be due to anemia of chronic disease or iron deficiency anemia or hemolytic anemia in the order of its occurrence. Among white cell lineage, leukocytopenia observed in 46 to 64 % of cases. neutropenia or lymphopenia are observed of which latter is a marker of disease activity. Thrombocytopenia is noted in 7-30% of cases and commonly observed in 15% of pediatric cases at its onset. [1]

Alterations in the coagulation system can be thrombotic or hemorrhagic of which thrombotic are more prevelant and is noted to have high morbidity and mortality. The thrombotic manifestations have a prevelance of 10 - 15% depending on the presence of risk factors. These includes of antiphospholipid antibodies (APL). The prevelance of lupus anticoagulants is 20-30%, beta2glycoprotien 40 % and anticardiooipin antibodies 42%. [6] High levels of homocysteine are considered as an independent risk factors for atherosclerosis, arterial and venous thrombosis. [7] Hemorrhagic manifestations are less frequent. Alveolar haemorrhage was a rare manifestations but

carries a high mortality. Hemorrhagic manifestations are also related to acquired defects of coagulation factors with respect to presence of inhibitors. [8] SLE can be treated with high dose steroid, cytotoxic drugs, IV immunoglobulin and plasmapheresis.

Conclusion

SLE should be promptly considered in the differential diagnosis of a child or adolescent with unexplained multiorgan involvement associated with fever and increased ESR. Although there is a striking female predominance in SLE, diagnosis must be considered even in a male child with atypical presentation. Although involvement of cell lines are commonly noted in SLE Coagulation abnormalities also needs detailed workup. So that specific therapy can be initiated early to decrease the morbidity and mortality associated with this disease.

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Conflicts of Interest

There are no conflicts of interest.

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