Idiopathic Thrombocytopenic Purpura Masquerading Paediatric SLE

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Sir,

Idiopathic Thrombocytopenic purpura (ITP) is an auto-immune disease characterized by accelerated clearance of auto-antibody sensitized platelets and suboptimal platelet production. It is a diagnosis of exclusion established after ruling out secondary causes like medication, Auto-immune diseases, Lympho-proliferative disorders and chronic infection. We report a case of paediatric Systemic Lupus Erythematosus (SLE) which initially manifested as thrombocytopenia and was diagnosed as ITP. Development of cutaneous involvement followed the diagnosis of ITP a year later.

A 4 year old girl child was diagnosed as ITP at the age of 2.5 years when she presented with petechiae and thrombocytopenia (Platelet count = 34000). Her bone marrow biopsy revealed a hypercellular marrow with increased number of megakaryocytes. Direct Coombs and ANA at presentation were negative. She underwent multiple (eight) admissions in subsequent 1 year wherein she was treated with systemic steroids which led to fluctuating platelet levels (varying from 2000- 35000). ANA repeated twice during the treatment course was negative. After 1.5 years; child started developing asymptomatic multiple erythematous annular plaques on face and extremities which were misdiagnosed at several occasions with possibility of Tinea and Seborrhoic dermatitis. Six months after onset of skin lesions she started developing lesions of different morphology. She developed annular erythematous papules and plaques on bilateral malar area and petechiae, purpura on lower extremities. Many discrete papules on face and extremities developed central necrosis resembling targetoid lesions. Lesions healed with evidence of epidermal atrophy. She had cold extremities with dusky erythematous macules topped with ulceration on tips of fingers and toes [Figure 1]. Oral examination revealed erosions. Fundus examination revealed evidence of healed choroiditis. Rest of her systemic examination was normal. Based on these findings, her work up was done for probable diagnosis of Vasculitis in setting of SLE with or without lupus pernio and Rowell’s syndrome.

Investigations revealed mild anemia and thrombocytopenia (Platelet: 60000) without any evidence of hemolytic anemia. Her ANA was now positive in a speckled pattern. She had highly positive dsDNA (>1000) suggestive of high disease activity, positive anti Smith antigen (suggestive of disseminated LE) and positive AntiRib P protein. Her coagulation profile, ANCA, β2 microglobulin, complement levels and renal function were normal. Lesional biopsy revealed features of immune vasculitis. Since she fulfilled 4 out of 11 ARA criteria, she was diagnosed as a case of SLE and started on daily oral Prednisolone at dose of 1mg/kg and Hydroxychloroquine at 6mg/kg. Steroids were gradually tapered with response. Treatment led to stabilization of platelet count, clearance of skin lesions and fall in titre of dsDNA to 160 within 2 months of therapy.

Figure 1: Clinical photograph of patient showing targetoid lesions on face; healing with epidermal atrophy and purpuric lesions on extremities with central necrosis and ulceration
Thrombocytopenia is seen in 7-30% of SLE patients.\(^1\) Immune thrombocytopenia may precede SLE in up to 16% patients.\(^2\) Mestanza Peralta et al., followed up 115 patients of ITP, who underwent splenectomy for its treatment, for 7.2 years and found that 14 patients developed SLE.\(^3\) Similar results were found by Anderson et al., and Perez et al., However; the studies by Altintas and Kurata had contrasting results. Altintas studied 365 children and 108 adults for a follow up period ranging from 2.1-7 years. He found 9% children and 33% adults to be ANA positive. But none of the ANA positive patients developed SLE.\(^4\)

Older age, female gender and positive ANA are significant risk factors for development of ITP in patients of SLE.\(^5\) Presence of high titre ANA is a sensitive marker for future development of SLE in these patients.

Our case highlights the importance of a regular follow up of all patients of ITP and periodic screening with ANA. Any cutaneous changes in these patients should not be ignored and evaluated thoroughly in light of any connective tissue diseases as early disease detection can help in better patient management.

References


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