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Splenectomy for refractory thrombocytopenia in juvenile systemic lupus erythematosus

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Background

The prevalence of autoimmune thrombocytopenia in juvenile systemic lupus erythematosus (JSLE) ranges from 7–52% (mean 14,5%) but profound rates (<50,000/mL) are uncommon. Although the exact mechanism is unknown, the spleen has been implicated either as the source of antiplatelet antibodies or the site of sensitized platelets destruction. Splenectomy is rarely indicated due to the increased risk of severe infections and controversial effectiveness.

Materials and methods

From 1983 to 2007, 5079 patients were followed at the Pediatric Rheumatology Unit and JSLE occurred in 228 (4.5%). We report three female JSLE patients with refractory thrombocytopenia to whom splenectomy was indicated.

Results

Data are described in table 1. Thrombocytopenia preceded JSLE diagnosis in a mean of 30 months. All patients had initial normal bone marrow and positive antiphospholipid antibodies.

Conclusion

Splenectomy should be considered for the treatment of thrombocytopenia refractory cases. Results are variable. Bone marrow hypoplasia should be excluded.

Table 1: Clinical and laboratorial manifestations and treatment of refractory thrombocytopenia

Patient	SSR	ACS	MJS
Age (years)	16	12	12
Months between JSLE/splenectomy	1	38	10
Previous treatment	Pd, MP, CYC	Pd, MP, HCQ, IVG, AZA, C, RTX	Pd, MP, HCQ, IVG, AZA, C, MMF, RTX
Platelet count	3,000/mL	7,000/mL	10,000/mL
Splenectomy	Held	Held	Indicated (cancelled due to bone marrow hypoplasia-toxicity?)
Platelet count (time after surgery)	107,000/mL (2 w) 469,000/mL (2 m)	140,000/mL (1 d) 396,000/mL (3 d)	-
Recurrences	none	1 month after	-

Pd = prednisone; MP = Methylprednisolone pulse; CYC = Cyclophosphamide; HCQ = Hydroxychloroquine; IVG = Gammaglobulin; AZA = Azathioprine; C = Cyclosporine; MMF = mycophenolate mofetil; RTX = Rituximab