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SPLENECTOMY IN A PATIENT WITH REFRACTORY LUPUS-ASSOCIATED THROMBOCYTOPENIA - CASE REPORT

Romão Augusto Alves Filgueira Sampaio
Serviço de Reumatologia, Hospital Universitário Walter Cantídio, Universidade Federal do Ceará (UFC)
Fortaleza - CE
http://lattes.cnpq.br/5390537109696094

Mailze Campos Bezerra
Serviço de Reumatologia, Hospital Universitário Walter Cantídio, Universidade Federal do Ceará (UFC)
Fortaleza - CE

Abstract: Thrombocytopenia in Systemic lupus erythematosus (SLE) is frequent and sometimes difficult to manage. The efficacy of splenectomy in this group of patients is controversial. We present the case of a patient with SLE-associated thrombocytopenia refractory to multiple pharmacological therapies and who achieved therapeutic response only after splenectomy.
Keywords: Systemic lupus erythematosus, immune thrombocytopenia, splenectomy.

BACKGROUND

Systemic lupus erythematosus (SLE) is a multisystemic inflammatory disease and hematologic manifestations are frequent and sometimes difficult to manage. Thrombocytopenia may be the first sign of SLE and precede other clinical manifestations by years. We present the case of a patient with SLE-associated thrombocytopenia refractory to multiple pharmacological therapies and who achieved therapeutic response only after splenectomy.

CASE REPORT

Female patient, diagnosed with primary immune thrombocytopenia at age 42. Thrombocytopenia was mild, with no need for immunosuppression. At age 45 she was diagnosed with SLE after the clinicolaboratory findings: ANA+, anti-DNA+, malar erythema, alopecia, polyarthritis. She started hydroxychloroquine, resulting in remission of all manifestations (including platelet levels). At age 52 there was recurrence of thrombocytopenia (platelet levels below 50,000/mm^3), spontaneous ecchymoses, and hypermenorrhea. It was prescribed azathioprine 2 mg/Kg/day, with no response after months of use. Successively failed mycophenolate mofetil 2g/day, thrombopoietin receptor agonists (eltrombopag, romipolostim) and infusion of rituximab 2g. She evolved with severe thrombocytopenia (platelets < 1 mil/mm^3) unresponsive to methylprednisolone pulse therapy. Transient response with human immunoglobulin (platelet levels dropped sharply after 2 weeks of infusion). Bone marrow biopsy and immunohistochemistry ruled out malignancy. Antiphospholipid antibodies negative. Abdominal ultrasound showed no splenomegaly. A laparoscopic splenectomy was performed resulting in sustained normalization of platelet levels (surgery 8 months ago). She is currently with SLE out of activity (SLEDAI: 0).

CONCLUSION

Immune thrombocytopenia in SLE is frequent and sometimes difficult to manage. Splenectomy is reserved for severe and refractory cases. The efficacy in this group of patients is controversial. Some authors suggest limited effect and others point to similar efficacy and safety in patients with primary immune thrombocytopenia and those with SLE. Having associated antiphospholipid syndrome seems to be a risk factor for relapse after surgery. Literature data show sustained response (with or without associated pharmacological therapy) ranging from 64% to 85% of patients after the procedure. More studies are needed to better define the efficacy and safety of splenectomy in patients with immune thrombocytopenia associated with SLE.

REFERENCES

1. Kado R, McCune WJ. Treatment of primary and secondary immune thrombocytopenia. Curr Opin Rheumatol. 2019 May;31(3):213-222. doi: 10.1097/BOR.0000000000000599. PMID: 30920453.

2. Barron N, Arenas-Osuna J, Medina G, Cruz-Dominguez MP, González-Romero F, Velásques-García JA, Ayala-López EA, Jara LJ. Splenectomy in systemic lupus erythematosus and autoimmune hematologic disease: a comparative analysis. Clin Rheumatol. 2018 Apr;37(4):943-948. doi: 10.1007/s10067-018-3979-4. Epub 2018 Jan 16. PMID: 29335897.

3. Galanopoulos N, Christoforidou A, Bezirgiannidou Z. Lupus thrombocytopenia: pathogenesis and therapeutic implications. Mediterr J Rheumatol. 2017 Mar 28;28(1):20-26. doi: 10.31138/mjr.28.1.20. PMID: 32185250; PMCID: PMC7045921.