SAFETY AND EFFICACY OF SPLENECTOMY IN THE TREATMENT OF ANTIPHOSPHOLIPID SYNDROME-ASSOCIATED CYTOPENIAS

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Background: Thrombocytopenia and autoimmune hemolytic anemia (AIHA) are common hematologic manifestations in primary antiphospholipid syndrome (APS). Although splenectomy is considered a second-line treatment in both primary immune thrombocytopenia (ITP) and idiopathic AIHA, its role in APS patients with either one of these manifestations has not been adequately defined, mainly because of the theoretically increased risk of thrombosis for patients with APS who undergo surgery.

Objectives: To determine the safety and efficacy of splenectomy for steroid-refractory thrombocytopenia or autoimmune hemolytic anemia in patients with primary APS, when compared to patients with ITP or idiopathic AIHA.

Methods: We performed a retrospective, single-center, case-control study. We included patients with primary APS and either thrombocytopenia, or autoimmune hemolytic anemia/Evans syndrome who underwent splenectomy between 2000 and 2018. The control group was made up by patients with primary immune cytopenias (ITP or AIHA) who also underwent splenectomy during that period. Cases and controls were adjusted by age, the hematologic manifestation and date of splenectomy. We recorded demographic, clinical and serologic characteristics at the time of surgery and during follow-up.

Results: We included 34 patients in each group. Thrombocytopenia was the indication for splenectomy in 53% of patients, with AIHA or Evans syndrome comprising the remaining 47%. Most patients were female (78%) and median age was 37 years. Among APS patients, 41% had triple-antibody positivity. There were no differences regarding comorbidities between groups. Patients with APS received more immunosuppressive treatment lines before splenectomy compared to controls (p=0.02), and there was a trend for more high-dose steroid cycles in the APS group (p=0.07). Median time to splenectomy was 54 months in APS patients and 18 months in controls, but without statistical significance.

Regarding splenectomy, most were laparoscopic (88%) and surgical complications were similar between groups (18%). However, patients with APS had a higher incidence of global non-surgical complications in the first month (50 vs 23%, p=0.04), most of them being infections (21 vs 3%, p=0.05). There was no difference in the incidence of post-surgical thrombosis, venous or arterial, between groups.

Most patients achieved a global response after one month (85% in APS group, 91% in controls, p=0.7). Complete response was observed in 65% and 79% of cases and controls, respectively (p=0.27). Median follow-up time was 52 months for APS patients and 41 months for controls. There were no differences regarding relapse which required any treatment adjustment between cases and controls (44% and 38%, respectively, p=0.8, Fig 1). However, 47% of APS patients received a prolonged maintenance immunosuppressive treatment, compared with 6% of controls (p<0.01). The incidence of infections and thrombosis during follow-up was similar between groups (p=0.15 and p=0.7, respectively; Fig 2).

Conclusion: Splenectomy is associated with adequate and long-lasting responses in APS patients with cytopenias, which do not differ from patients with non-APS-associated thrombocytopenia and AIHA. Thrombosis was not a common complication in our patients; however, there was a higher incidence of infections in APS patients. This could be related to the higher steroid doses and more intensive previous immunosuppressive therapies. Splenectomy could be considered an earlier treatment option for APS patients with refractory cytopenias, and this could reduce infection risk and post-surgical morbidity.

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HYDROXYCHLOROQUINE FOR THE PREVENTION OF RELAPSES IN A SERIES OF 812 PATIENTS WITH PRIMARY ANTIPHOSPHOLIPID SYNDROME: THE HIBISCUS RETROSPECTIVE STUDY

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Background: The relapse rate in antiphospholipid syndrome (APS) remains high, 20% at 5 years in thrombotic APS and 28% in obstetrical APS (1). Hydroxychloroquine (HQC) appears as an additional therapy, with immunomodulatory and antithrombotic effects (2-5).

Objectives: The main aim of this trial is to assess the efficacy of treatment with Hydroxychloroquine in preventing new events in primary anti-phospholipid syndrome patients.

Methods: We have performed a retrospective multicentre open-labelled study (2002-2018).

Results: 812 patients with APS from 53 international centres from 16 countries were included. In all cases, the previous standard treatment was inefficient. The mean follow-up was 20.2±10.8 months, the mean age 39.5 years old.

The type of clinical manifestations is described in figure 1.