Splenectomy and/or cyclophosphamide as salvage therapies in thrombotic thrombocytopenic purpura: the French TMA Reference Center experience: SALVAGE THERAPIES IN SEVERE TTP

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Résumé en anglais	BACKGROUND: The objective was to assess the efficacy and safety of splenectomy and cyclophosphamide as salvage therapies in severe thrombotic thrombocytopenic purpura (TTP).STUDY DESIGN AND METHODS: During a 10-year period, patients who did not improve with plasma exchanges, steroids, vincristine, and/or rituximab were considered for splenectomy or cyclophosphamide. Patients with a documented severe (<10% of normal value) acquired ADAMTS13 deficiency are reported here. RESULTS: Eighteen patients with a severe acquired ADAMTS13 deficiency required a salvage therapy. Thirteen patients had a splenectomy 19 (interquartile range [IQR], 10-51) days after TTP diagnosis. One patient died the day after splenectomy. The remaining patients improved platelets (PLTs) until Day 6, along with a rapid and major lactate dehydrogenase improvement. Six patients, however, subsequently experienced a transient worsening. Durable PLT count recovery in survivors was observed within 13 (IQR, 11.5-25.5) days. Postoperative complications included thromboembolic events (two cases) and infections (five cases). Five patients received pulses of cyclophosphamide 12 (IQR, 12-15) days after TTP diagnosis. All patients recovered PLTs 10 (IQR, 9-24) days after the first pulse and two experienced a transient worsening. Three patients experienced infections. Three relapses occurred 5 months, 2.5 years, and 4.5 years after splenectomy and one relapse occurred 3.5 years after cyclophosphamide. After a 2.5 (IQR, 0.75-6.2)-year follow-up, the overall survival was 94%. CONCLUSION: Cyclophosphamide and splenectomy provide comparable high remission rates in severe TTP with acceptable side effects and should be considered in the more severe patients who do not improve with other therapies.
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