Thrombotic Thrombocytopenic Purpura: overview on the last 10 years of plasma exchange treatment in Hospital São José, Lisbon

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INTRODUCTION

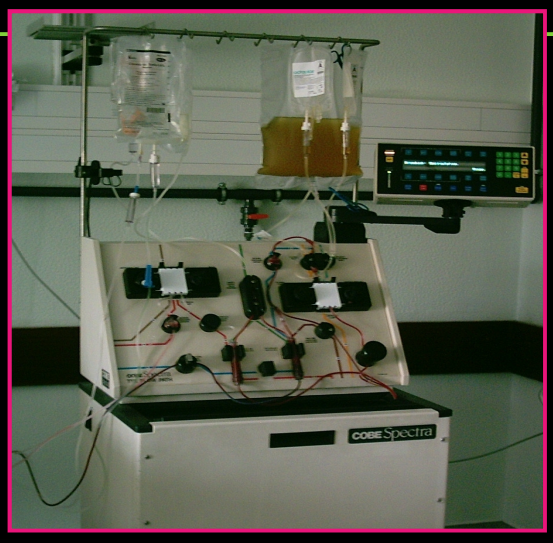
- Thrombotic Thrombocytopenic Purpura (TTP) was first described by Moschowitz in 1924.
- It is a rare disease.
- In the era before therapeutic plasma exchange (PE) 90% of patients died from systemic micro vascular thrombosis.
- PE removes the causative antibody to von Willebrand factor cleaving metalloprotease (ADAMTS13) and replaces ADAMTS13. Recognition of TTP can be difficult because of the variety of presentations and lack of specific diagnosis criteria.

A diagnosis of TTP may be made in the presence of a microangiopathic haemolytic anaemia and thrombocytopenia in the absence of any other identifiable cause.

- PE is the only treatment for which there are firm data on its effectiveness in TTP in adults.

MATERIAL AND METHODS

- Patient's Hospital medical records were rewied since January 2000 until December 2010.



- Daily PE with replacement of 1,0 to 1,5 times the predicted patient's plasma volume was initiated at least 24h after diagnosis , for a minimum of two days after platelet count and lactate desidrogenase returned to normal.
- CobeSpectra® (Caridian) was used; the replacement fluid administrated was solvent/detergent-treated plasma (Octaplas®, Octapharma); central venous access was used in all patients.

RESULTS

- In the last ten years we performed PE in 14 adult patients (11 females and 3 males).
- 21 episodes of TTP were observed: 17 were idiopathic and 4 were related to HIV infection (1) or first trimester pregnancy (3).
- No measurements of ADAMTS13 activity or antibody were made.
- Clinical presentation was heterogeneous and differential diagnosis was almost exclusively made with acute leukaemia.
- The most frequent symptom was muco cutaneous bleeding.
- Adjuvant corticosteroid therapy was instituted in all patients.
- PE was effective in all TTP episodes except one.
- Four patients relapsed .
- Four patients were submitted to immunosuppressive agents (rituximab, azatioprin and cyclophosphamid) when exacerbations or relapses occurred.
- Complications associated with PE were *minor* (allergic reaction and high blood pressure).

CONCLUSIONS

- A high index of suspicion of TTP is required for rapid diagnosis and prompt initiation of PE treatment.
- PE should be instituted within 24 hours of presentation of TTP. Plasma infusion remains appropriate when there may be a delay until PE is available.
- PE is a safe and effective treatment associated with excellent clinical outcomes.
- The optimal duration of therapy is unknown and once a patient is in remission the efficacy of any treatment to prevent relapses is uncertain.