Abstract: PB3390

Title: ATYPICAL PRESENTATIONS OF PEDIATRIC ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA

Abstract Type: Publication Only

Topic: Thrombosis and vascular biology

Background:

Immune Thrombotic Thrombocytopenic Purpura (iTTP) in children is a rare, severe thrombotic microangiopathy. This condition is characterized by microangiopathic hemolytic anemia, severe thrombocytopenia, and organ ischemia due to reduced activity of the von Willebrand factor-cleaving protease ADAMTS13.

Aims:

To evaluate underlaying etiology and clinical course amonog children diagnosed with

iTTP

Methods:

A retrospective case series evaluating data collected from the medical files of four children diagnosed with iTTP.

Results:

The presented case series depicts a variety of iTTP presentations: one case of primary iTTP, one case induced by Shiga toxin, one associated with RAS-associated autoimmune leukoproliferative disease (RALD), and one initial manifestation of systemic lupus erythematosus (SLE). Notably, two patients recovered without undergoing plasma exchange.

Patient	Treatment at diagnosis	Time to resolution of thrombocytopenia (days)	Relapses	Current therapy	Outcome	Length of follow-up (months)
1	Plasma transfusion, CS	7	no	no	Complete remission	6
2	TPE, CS, Rituximab	4	yes	Sirolimus	Complete remission	60
3	CS, IVIG, Rituximab	9	no	Belimumab, HCQ, Coumadin	Complete remission	19
4	TPE, CS, Rituximab, Calplacizumab	2	no	no	Complete remission	5

Abbreviations: CS, corticosteroids; HCQ, hydoxychloroquine; IVIG, immunoglobulins; TPE, Therapeutic plasma exchange

Summary/Conclusion: Early ADAMTS13 testing in children with unexplained hemolysis or thrombocytopenia is crucial. The diverse underlying causes, including infections and autoimmune disorders, underscore the complexity of iTTP in the pediatric population. These cases highlight the necessity for personalized treatment

approaches that consider each patient's unique clinical situation and potential alternatives or modifications to conventional therapeutic regimens.

Keywords: ADAMTS13, Thrombotic thrombocytopenic purpura (TTP)