48th ANNUAL DIAGNOSTIC SLIDE SESSION, 2007 DIAGNOSES AND REFERENCES

MODERATOR: E. Tessa Hedley-Whyte, M.D.

EDITOR: Leroy R. Sharer, M.D.

Case 2007-5

Submitted by: Leroy R. Sharer, M.D., Valerie A. Fitzhugh, M.D., and Debra S. Heller, M.D., UMD-New Jersey Medical School, Newark, NJ 07103

Diagnosis: Cerebral vasculopathy, with extensive and multiple hemorrhages in brain, old and recent, secondary to probable alloimmune thrombocytopenia of infancy

Comment: From the Presenter (Dr. Sharer): Alloimmune thrombocytopenia of infancy (ATI), a diagnosis first suggested for this case by Dr. Lucy B. Rorke-Adams, occurs when maternal antibodies are directed against paternal antigens expressed on platelets; and it can be a cause of intrauterine hemorrhage in brain, as in this case. Up to 10% of infants with this disorder have evidence of brain hemorrhage on ultrasound; and when it occurs, such hemorrhage is a major cause of morbidity and mortality. Clinical information suggested that this disorder was ruled out in this child, but there is a large number of platelet antigens, most commonly glycoproteins, that have been implicated in the causation of ATI, and not all of them are routinely tested. It is likely that the mother was sensitized during a previous pregnancy, which ended in spontaneous abortion. Few reports of neuropathological findings are available for this disorder, with none of them mentioning the vasculopathy seen in this case. It should be noted that the child survived for several weeks after platelet transfusions were discontinued, perhaps allowing for development of the more chronic cerebral vascular changes, with thickening of the intima and media. Appreciation is also expressed to Dr. Mohila. for pointing out her abstract that was presented at this same combined EB/ASIP/AANP meeting.

References:

Berkowitz RL, Bussel JB, McFarland JG: Alloimmune thrombocytopenia: State of the art 2006. Am J Obstet Gynecol 2006; 195:907-913.

Bussel JB, Zacharoulis S, Kramer K, et al: Clinical and diagnostic comparison of neonatal alloimmune thrombocytopenia to non-immune cases of thrombocytopenia. Pediatr Blood Cancer 2005; 45:176-183.

Dean LM, McLeary M, Taylor GA: Cerebral hemorrhage in alloimmune thrombocytopenia. Pediatr Radiol 1995; 25:444-445.

Mohila CA, Ornvold K, Kubicka ZJ, Harris BT: Cerebellar loss and brain stem atrophy associated with neonatal alloimmune thrombocytopenia in a discordant twin (abstract). Experimental Biology, April 2007, abstract #559.1, Washington, DC.