Fetomaternal Alloimmune Thrombocytopenia Presenting as Intracerebral Bleeding in Utero

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Abstract. Feto-maternal alloimmune thrombocytopenia is a serious fetal disorder resulting from platelet antigen incompatibility between the mother and the fetus. Intracranial bleeding is the most serious complication of alloimmune thrombocytopenia and can result in severe disability and death in utero. The authors report a case of intracerebral hemorrhage in utero resulting from alloimmune thrombocytopenia. [Indian J Pediatr 2005; 72 (4) : e35-e37] E-mail: amuchou@yahoo.com.

Key words: Intracranial bleed; Alloimmune; Thrombocytopenia.

Fetomaternal alloimmune thrombocytopenia (FMAIT) is a disorder caused by fetomaternal platelet incompatibility analogous to that in rhesus hemolytic disease, with maternal antiplatelet antibodies crossing the placenta and destroying the fetal platelets. Intracranial hemorrhage is the most serious complication of FMAIT and can result in severe disability and death in utero. Here is reported a case of intracerebral hemorrhage in utero resulting from alloimmune thrombocytopenia.

CASE REPORT

A healthy 21-year-old gravida 2 para 0 woman was referred to the maternal and fetal medicine clinic for an abnormal intracranial fetal ultrasound at 32 weeks, which was done for poor fetal growth. Prior ultrasound scans done at 11 and 19 weeks were normal. Ultrasound revealed an abnormal head size with a large hyper echoic space-occupying lesion measuring 3 X 3.5 cm at the right cerebral hemisphere extending to the posterior horn of lateral ventricle (Fig. 1). Rest of the detail ultrasound revealed no abnormality. Biophysical profile was 8/8. Repeat ultrasound done after one week did not show any change in the size or echogenicity of the mass. Possibility of an intracranial tumor or intracranial bleed was considered.

The patient had spontaneous abortion at 15 weeks in her first pregnancy. She had no history of bleeding or autoimmune disorders in the past. She was not a smoker, did not drink alcohol or use cocaine. There was no family history of bleeding disorders.

Investigation revealed hemoglobin (Hb) of 13.4 gm/dl, with normal white blood cell and platelet counts. Blood group was O-positive. Serology for syphilis, toxoplasma, cytomegalovirus, hepatitis B and human immunodeficiency virus, rubella was negative. In view of the above ultrasound finding, she was further investigated to rule out alloimmune thrombocytopenia.

Platelet antigen studies revealed absence of human platelet antigen (HPA-1a). She had antiplatelet antibody against HPA-1a. ELISA done on maternal serum revealed no HLA antibodies. Her husband was investigated and he was homozygous for human platelet (HPA-1a) antigen. HLA typing was not performed during antenatal period. Parental counselling was done; elective cesarean section was planned. She received 2 doses of betamethasone prior to cesarean section. Compatible platelet (HPA-1a antigen negative) was arranged.

A female baby was delivered by elective cesarean section at 34 weeks. She cried soon after birth with Apgars of 8 and 9 at 1 and 5 minutes. The birth weight was 2420...
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alloimmune thrombocytopenia had intracranial hemorrhage. Of the 12 babies, 11 had intracerebral hemorrhage; of which 5 occurred in utero. Only one baby had subarachnoid bleeding. Intracranial hemorrhage in the utero can occur as early as 16 weeks but the majority occurred at 35-37 weeks of gestation. Intracranial hemorrhage in alloimmune thrombocytopenia is typically intraparenchymal, unlike ICH due to prematurity, which is characterized by intraventricular bleed. The most common site for intracranial hemorrhage is beneath the cerebral cortex, with expansion to a subarachnoid bleed and dissection through the brain reaching ventricles. Intracranial hemorrhage can lead to death or permanent neurological sequelae, such as seizures, learning disabilities, cerebral palsy, hypotonia, cortical blindness, developmental delay, seizures, and psychomotor retardation.

The diagnosis of FMAIT is confirmed by demonstrating antibodies in maternal serum that react with both father’s and child’s platelets and not with mother’s platelets. Antenatal management of fetal alloimmune thrombocytopenia is controversial and includes a combination of maternal intravenous gamma globulin (IVIG) administration, intrauterine platelet transfusion and corticosteroid therapy, while monitoring fetal platelet count closely throughout the course of pregnancy. Antenatal treatment of alloimmune thrombocytopenia with weekly gamma globulin effectively improves the fetal platelet count and prevents intracranial hemorrhage. In a series of 54 women with thrombocytopenic fetus, given IVIG weekly, no intracranial hemorrhage occurred and 20% showed no increase in platelet count with therapy.

Postnatally, the treatment of choice for alloimmune thrombocytopenia is administration of maternal (or compatible) platelets. Maternal platelets for transfusion must be washed and free from antibody containing plasma and resuspended in AB plasma. It is preferable to arrange the platelets before delivery of the baby. Other treatment options are intravenous immunoglobulin with or without steroids. When matched platelets are not available in an emergency situation, IV immunoglobulin 1 gm/kg/dose for two days have shown to be effective in cases with severe thrombocytopenia. Steroids have been tried in situations with continued low platelet count and bleeding.

The natural history of FMAIT is one of the progressive worsening of the disease in subsequent pregnancies. Previous antenatal intracranial hemorrhage is a reliable predictor of severely low platelet counts and intracranial hemorrhage in the subsequent affected fetus.

In the present index case, the father was homozygous for platelet antigen. Therefore, there is 100% recurrent risk in future pregnancy. Parents have been counselled regarding the need for antenatal therapy with repeated platelet transfusion or weekly immunoglobulins therapy in subsequent pregnancies.
This case is reported to consider and to have a high index of suspicion for alloimmune thrombocytopenia in a differential diagnosis of a fetus presenting with hyperechoic intracerebral mass in utero.

REFERENCES