Gynecology & Obstetrics

Schepers et al., Gynecol Obstet 2013, 3:6
http://dx.doi.org/10.4172/2161-0932.1000184

Case Report Open Access

Excessive Bleeding due to Consumptive Coagulopathy after Delivery of an Intrauterine Fetal Death in a Patient known with Multiple Vascular Malformations; Mimicking the Kasabach-Merritt Syndrome

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Abstract

During the third trimester of pregnancy a physiological hypercoagulability state has been reported as a consequence of hormonal changes. This produces a vulnerable state for intravascular clotting. Some pathological obstetric conditions can modify this weak balance which can lead to hemorrhage and organ failure. We report a patient known with multiple Vascular Malformations and Localized intravascular coagulation causing a Chronic Mild Coagulopathy with excessive bleeding due to a high grade Disseminated Intravascular Coagulation at delivery of an intrauterine deceased full term fetus.

Keywords: Obstetric; Intrauterine deceased fetus; Hemorrhage, Consumptive coagulopathy

Case Report

We report a patient with excessive bleeding due to a high grade Disseminated Intravascular Coagulation (DIC) after delivery of an intrauterine deceased fetus.DIC is characterized by widespread activation of the clotting and fibrinolytic systems resulting in a hypocoagulable state due to consumption of clotting factors and platelets [1-3]. In addition to the clinical presentation the diagnose is confirmed with the characterized hematological findings as prolonged prothrombin time (PT), activated partial thromboplastin time (APTT), low platelet count, low fibrinogen and elevated products of fibrin breakdown [4]. During the third trimester of pregnancy a physiological hypercoagulability state has been reported as a consequence of hormonal changes. This produces a vulnerable state for intravascular clotting. Some pathological obstetric conditions can modify this weak balance which can lead to hemorrhage and organ failure [2].

We saw a 41-year-old woman with an at term pregnancy complicated by an intrauterine fetal death. The obstetric history mentioned a preeclampsia in the first pregnancy with induction of labor. At that time, the blood loss was 1480cc from a genital laceration. Relevant medical history included an uncomplicated removal of a retrosternal and orbital Vascular Malformation and a laparotomic myomectomy because of a subserosal fibroid. A blood transfusion was needed after both operations because of a hemorrhage. The family history mentioned Vascular Malformations which affected her father, two brothers, a daughter of a brother and patients first born son.

Two days after the establishment of the fetal death labor was induced. At that time the laboratory coagulation results were normal: The APTT was 31 sec (normal 0-32), the PTT was 12.9 sec (normal<14.5) and the fibrinogen level was 2.2 g/l (normal 2.0-4.5 g/L), with a platelet count of 196×10^9/l (normal 150-400). After a normal delivery a lifeless full-term son was born. The placenta was expelled quickly, however excessive bleeding occurred. Uterus massage and intravenous infusion with sulproston was started without effect. Because of refractory uterine bleeding the uterus was manually inspected in the operating room but no remaining placental tissue was found. The uterus, cervix and vagina were intact. A Bakri balloon was used for tamponade. However this did not reduce the fluxus. The laboratory coagulation results, determined only four hours after the previous normal results, were severely disturbed: The APTT was 317 sec, the PTT was 37.6 sec, the platelet count was 56×10^9/l with an undetectable serum fibrinogen level and a D-dimer of >20000 (normal<500) compatible with DIC. The bleeding eventually stopped after treatment with six fresh frozen plasma, one platelet concentrate, fibrinogen and tranexamic acid together with embolization of the uterine artery. Patient lost 7500cc of blood and received 13 packed red blood cells. A request for autopsy of the fetus was declined. The placenta did not show abnormalities and no microthrombi were found.

Two weeks later the patient had no clinical signs of hemorrhagia or other symptoms. Laboratory investigation showed normal APTT, PT and platelet count but low-normal fibrinogen (1.5 mmol/L) and an elevated high d-dimer (15000 ng/mL) persisted. In addition patient had a hemoglobin of 11.6 g/L (normal 11.5-15.2), reticulocytes of 26 promille (normal 5-25), a low haptoglobin (<58 mg/dL), a normal LD and an elevated unconjugated bilirubin. In the blood film 3/1000 schistocytes were noted. The laboratory data are compatible with a Chronic Mild Coagulopathy and a compensated low grade mechanical hemolysis. Magnetic Resonance Imaging (MRI) of the total body revealed Vascular Malformations at various parts of the body but not in the uterus (Figure 1).

This patient presented with excessive bleeding after delivery due to a high-grade DIC. A few weeks later the laboratory results still showed a Chronic Mild Coagulopathy with a compensated low-grade hemolysis. The family history mentioned several family members with vascular malformations. This family very likely has a hereditable vascular malformation predisposition. The combination of multiple vascular malformations with this coagulation disorder is based on consumption

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Received October 13, 2013; **Accepted** November 15, 2013; **Published** November 22, 2013

Citation: Schepers EM, Tom Vlasveld L, Snoep JJD, Loyson SAJ, Boers KE (2013) Excessive Bleeding due to Consumptive Coagulopathy after Delivery of an Intrauterine Fetal Death in a Patient known with Multiple Vascular Malformations; Mimicking the Kasabach-Merritt Syndrome. Gynecol Obstet 3: 184 doi:10.4172/2161-0932.1000184

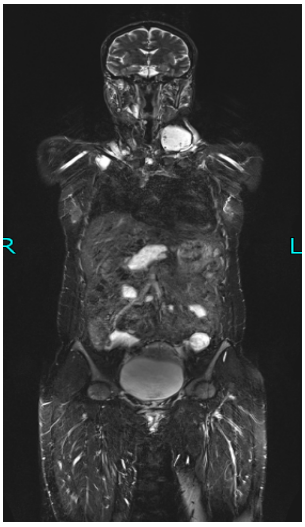

Figure 1: Magnetic resonance imaging (MRI) of the total body revealed vascular malformations at various parts of the body.

of clotting factors due to trapping of platelets by abnormally growing vascular tumors and hemolysis caused by damage of red blood cells [5]. This coagulation disorder with an autosomal dominant variant of multiple vascular malformations is mimicking the Kasabach-Merritt Syndrome (KMS). Typically the KMS is represented by a potentially life-threatening coagulopathy characterized by enlarging hemangiomas with thrombocytopenia and consumptive coagulopathy. KMS is associated with kaposiform hemangioendothelioma, tufted angiomas and rarely with congenital hemangiomas. In 1967 the first case of this coagulation disorder occurring during pregnancy was reported. This case and a few other cases, which were described in the subsequent years, showed Localized Intravascular Coagulation (LIC) due to KMS with consumptive coagulopathy after a normal pregnancy and a normal at term delivery [6,7]. In one patient with KMS, LIC occurred after a cesarean delivery, performed because of a large complex of varicosities extending along the vaginal wall and the labia majora which partially obstructed the introitus [8]. As is described in these cases the Kasabach-Merritt syndrome is exclusively associated with kaposiform hemangioendothelioma and tufted angioma.

In our case one may hypothesize that the physiologic activation of the coagulation system at the end of the pregnancy augmented the already consisting Chronic Mild Coagulopathy (due to the widespread Vascular Malformations and LIC) and resulted in placental circulation disturbances with intrauterine fetal death. Otherwise it may be argued that the intrauterine fetal death with subsequent delivery of a lifeless fetus resulted in activation of the coagulation, which superimposed on an already consisting Chronic Mild Coagulopathy, and resulted in a state of high grade DIC with consumptive coagulopathy [9]. In view of the fact that the coagulation parameters determined at the start of the delivery, two days after the intrauterine death, showed no signs of consumptive coagulopathy, and that pathological examination of the placenta revealed no microtrombi or massive bleeding, the latter sequence of events is most likely.

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Conclusion

We described a patient with multiple vascular malformations suffering from massive bleeding due to disseminated intravascular coagulation and consumptive coagulopathy mimicking the Kasabach-Merritt syndrome at delivery of an intrauterine deceased full term son.

The patient was advised not to pursue a new pregnancy any longer, given the complicated course of the delivery and the estimated risk of recurrent bleeding.

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