Clinical Reports

Amniotic fluid embolism and isolated disseminated intravascular coagulation

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Purpose: Disseminated intravascular coagulation (DIC) is a well-known complication of an amniotic fluid embolus. However, clinical experience has shown that, in some patients, clinical hemorrhage may be the initial presentation. Mortality in this subset of patients is high. This report describes a patient with a suspected amniotic fluid embolus who survived and in whom the initial presentation was post partum hemorrhage.

Clinical Features: During the post delivery repair of a fourth degree perineal tear a 29 yr old prima gravida was noted to have excessive vaginal bleeding despite a well contracted uterus. Laboratory investigations revealed a decrease in hemoglobin from 126 g·L⁻¹ to 86 g·L⁻¹ and a severe disseminated intravascular coagulopathy (PT 27.5 sec, APPT 149 sec, direct fibrinogen < 0.6 g·L⁻¹, FDP > 640 g·L⁻¹). Treatment included massive blood component therapy and, eventually, total abdominal hysterectomy. The patient subsequently recovered without sequelae. In the absence of any other explanation, the coagulopathy was considered to be secondary to amniotic fluid embolus.

Conclusion: Amniotic fluid embolus remains an important cause of maternal mortality. Clinical reports, reviews of the literature, and the development of the national registry have all contributed to our understanding of this syndrome. Contrary to earlier beliefs, not all patients present with sudden cardiorespiratory collapse. As the present case illustrates, a less common presentation is the development of an isolated DIC in the peripartum period without antecedent hemodynamic or respiratory instability. Therefore, anesthesiologists must maintain a high index of suspicion for the disorder in order to facilitate early recognition and treatment.

Objectif : La coagulation intravasculaire disséminée (CIVD) est une complication connue de l'embolie du liquide amniotique. Cependant, l'expérience a démontré que, chez certaines patientes, l'hémorragie peut être le premier signe clinique. La mortalité est élevée dans ce sous-groupe de patientes. Nous décrivons ici le cas d'une patiente qui a survécu à une embolie probable du liquide amniotique dont le premier signe était une hémorragie post-partum.

Éléments cliniques : Pendant la réparation post-partum d'une déchirure périnéale du quatrième degré chez une primigeste de 29 ans, on a noté des saignements excessifs en dépit d'un utérus bien contracté. Les examens de laboratoires ont révélé une baisse de l'hémoglobine de 126 g·L⁻¹ à 86 g·L⁻¹ et une sévère coagulopathie intravasculaire disséminée (TP 27,5 sec, TPA 149 sec, le fibrinogène direct < 0,6 g·L⁻¹, PDF > 640 g·L⁻¹). Le traitement a consisté en transfusions sanguines massives et, finalement, en une hystérectomie abdominale complète. La récupération a été sans complications. En l'absence d'autre explication, la coagulopathie a été considérée secondaire à une embolie du liquide amniotique.

Conclusion : L'embolie du liquide amniotique demeure une cause importante de mortalité maternelle. Les résumés cliniques, les revues de littérature et la production d'un registre national ont tous contribué à nous la faire connaître. Contrairement à ce que nous pensions auparavant, les patientes ne présentent pas toutes un collapsus cardio-respiratoire soudain. Comme dans le cas actuel, une présentation plus rare est le développement d'une CIVD en période péripartum sans antécédents d'instabilité hémodynamique ou respiratoire. En conséquence, les anesthésiologistes doivent être vigilants face à ce problème afin d'en faciliter le diagnostic et le traitement précoces.

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AMNIOTIC fluid embolus can be one of the most frightening and devastating complications of pregnancy. Traditionally, the syndrome has been thought to consist of sudden, profound cardiorespiratory failure in a labouring or post-partum patient.¹ However, more recently, many of our beliefs regarding amniotic fluid emboli have come into question. The following case report describes a patient with probable amniotic fluid embolus in whom the presenting feature and the ongoing physiological abnormality was a consumptive coagulopathy.

Case Report

A 29 yr old para 0, gravida 1 woman was admitted at 39 wk gestation for induction of labour. She had a history of insulin dependent diabetes since the age of six years. She had not suffered any complications related to her diabetes. Specifically, there was no history of cardiovascular, renal, neurological or ophthalmic involvement. The present pregnancy had been uncomplicated. Throughout gestation her progress was followed closely by both the endocrine and high risk obstetrical services. She had been able to maintain good blood sugar control and, during the latter part of her pregnancy, had normal biweekly biophysical profiles. Following admission she was given 2 mg prostaglandin E2 intravaginal gel. Five hours later she had spontaneous rupture of membranes followed by rapid progression of labour. She was transferred to the labour floor requesting epidural anesthesia. However, before this could be arranged she developed a tetanic uterine contraction accompanied by deep variable decelerations, necessitating transfer to the operating room. On arrival, vaginal examination revealed that she was fully dilated and pushing spontaneously. Therefore, the obstetrician elected to deliver the infant by a vacuum extraction. This was complicated by a fourth degree perineal tear. Neonatal Apgar scores were 5 and 8 at one and five minutes respectively. Seven minutes later the placenta delivered spontaneously, intact and with no evidence of a retroplacental clot. Following completion of the third stage of labour, she was given 10 units syntocinon *im* and the perineal tear was repaired under local anesthesia. During this time excessive vaginal bleeding was noted, although the uterus was felt to be well contracted. Intravenous access was established and the anesthesia service was summoned to provide assistance with resuscitation as well as pain relief to allow further examination into the source of bleeding. Following intravenous sedation, exploration confirmed a well contracted fundus but revealed profuse oozing from

the lower uterine segment and upper vagina, both of which were subsequently packed. Simultaneously, ongoing resuscitation included supplemental oxygen by nasal prongs, additional *iv* access, crystalloids and packed cells. Hemodynamically, the patient had a sinus tachycardia of 110-130 bpm, SpO2 98% and a systolic blood pressure 100 mmHg. Urine output was decreased to approximately 15 ml·hr⁻¹. Laboratory investigations 40 min post delivery, prior to the exploration, revealed a decrease in hemoglobin from 126 g to 86 g·l⁻¹ and coagulation studies consistent with a severe coagulopathy (PT 27.5 sec, APPT 149 sec, direct fibrinogen < 0.6 g·l⁻¹, FDP > 640 g·l⁻¹). While resuscitation continued, arrangements were made to transfer the patient to a nearby hospital for embolization of the internal iliac arteries. However, since pelvic angiograms revealed extravasation of blood bilaterally from the vaginal arteries, the radiologist elected to embolize only these branches. This appeared to be successful and the patient was transferred to the ICU for further observation.

Over the next several hours, despite ongoing correction of her coagulopathy, the patient continued to bleed. As well, she developed a hematoma extending into the right gluteal fold and thigh. Therefore, 15 hr post delivery she was taken to the operating room for exploration of the vagina and possible laparotomy. Intraoperatively, the patient was found to have a large ischiorectal hematoma as well as bleeding from the lower uterine segment, cervix and vagina. Despite suture ligation of the internal iliac arteries she continued to bleed, eventually necessitating total hysterectomy and resuturing of the vaginal mucosa. The trachea was extubated the following day and she had no further complications. In total, correction of her coagulopathy had required 18 units of packed red blood cells, 10 units of platelets, 8 units of cryoprecipitate, and 5 units of fresh frozen plasma. After exclusion of other causes of DIC it was felt that this patient's coagulopathy was secondary to an amniotic fluid embolus.

Discussion

The entrance of amniotic fluid into the maternal circulation was first described by Meyer in 1926.¹ However, it was not until 1942 that the clinical importance of this entity was appreciated. At that time, Lauchbaugh and Steiner reviewed the presentation and pathological findings in eight cases of unexpected death in pregnant patients.²⁻³ By the 1950s the coagulation changes that frequently accompany this entity were becoming appreciated.¹ Since then, clinical experience has shown that DIC is a common sequel of amniotic fluid emboli. In 1979, Morgan's¹

review of the literature revealed that in 12% of patients the initial presentation was a bleeding diathesis, as opposed to cardiorespiratory compromise. Of those who survived the initial insult, an additional 37% subsequently developed a consumptive coagulopathy. Similarly, in 15 cases of fatal amniotic fluid emboli associated with induced abortion, two patients presented with a coagulopathy. Among those who survived more than one hour, 75% had DIC.⁴ More recently, in an analysis of the national registry for amniotic fluid emboli, Clarke found that 83% of patients had either laboratory or clinical evidence of a consumptive coagulopathy.⁵

Although there is general agreement that DIC almost invariably accompanies amniotic fluid emboli, either clinically or subclinically, the etiology is unknown.^{6,8-13} Despite the presence of activated factors II, VII and X in amniotic fluid, their concentrations are well below those in maternal serum at term and, therefore, probably do not contribute to the coagulopathy.⁷ On the other hand, amniotic fluid has been shown to have a direct factor X activating property and thromboplastin like effect, both of which increase with gestational age. Lockwood *et al.* found substantial quantities of tissue factor in amniotic fluid and have suggested that this might be the triggering agent.⁷ Potential sources include fetal epithelial cells from the respiratory, gastrointestinal, and genitourinary tract mucosa as well as sloughed skin. If tissue factor is indeed the causative agent, it activates the extrinsic pathway by binding with factor VII. This complex can then trigger the clotting mechanism by activating either factor IX or X. The authors further speculated that once clotting was triggered in the pulmonary vasculature, local thrombogenesis could then cause vasoconstriction and microvascular thrombosis.⁷ Moreover, thrombin may stimulate secretion of vascular endothelin. Animal studies have shown that this vasoactive peptide can depress both myometrial and myocardial contractility. This in turn can contribute to both the hemodynamic changes and uterine atony frequently observed in this syndrome.

Despite inconclusive evidence regarding the cause of the coagulopathy in amniotic fluid emboli, clinically, activation of the coagulation system may lead to life threatening hemorrhage due to consumption of the coagulation factors and activation of the fibrinolytic system. For those patients who survive the initial hemodynamic changes or in whom these changes are subclinical, uncontrolled bleeding may become the chief cause of maternal mortality. Treatment of the coagulopathy consists of blood component therapy including packed red blood cells, fresh frozen plasma, platelets and cryoprecipitate. Although some authors¹⁴

have suggested the use of low dose heparin once the bleeding has been controlled, there is not sufficient evidence to support this practice.^{12,13} Similarly, antifibrinolytics should not be given in DIC as they may precipitate severe intravascular thrombosis.

Summary

The patient described represents one of the atypical presentations of amniotic fluid emboli in which an isolated coagulopathy causing maternal hemorrhage was the first indication of the problem. Unlike other patients presenting in a similar manner, she did not develop hemodynamic instability secondary to the cardiovascular effects of an amniotic fluid embolus.^{6,9} The differential diagnosis of DIC in this population includes severe abruptio placenta, prolonged retention of a dead fetus, septic shock, ruptured uterus, preeclampsia, transfusion reaction, multi-organ failure and massive trauma or blood loss. Careful consideration of all of these possibilities revealed that none were applicable to this patient. Therefore, the diagnosis of an amniotic fluid embolus as the underlying etiology was primarily clinical and one of exclusion.

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