June 2018

Amniotic fluid embolism

Nadia Nawaz
Aga Khan University

Ahmed Raheem Buksh
Aga Khan University, ahmed.raheem@aku.edu

CASE REPORT

Amniotic Fluid Embolism

Nadia Nawaz[1] and Ahmed Raheem Buksh[2]

ABSTRACT

Amniotic fluid embolism is a rare and severe problem in obstetric patients. We experienced a 21-year primigravida who underwent emergency cesarean section due to sudden collapse and fetal distress after rupture of membranes in labour. Subsequently, she developed intraoperative coagulopathy, hemorrhage, hypotension, and respiratory collapse requiring ventilation. Both maternal and neonatal lives were saved with full recovery and discharged in stable condition. The clinical diagnosis of amniotic fluid embolism (AFE) was made, which is a very rare complication of pregnancy/puerperium with varying presentation, ranging from cardiac arrest, circulatory collapse and death through mild degrees of organ system dysfunction with or without coagulopathy. AFE has no definitive test for its diagnosis; the detection of fetal elements in the maternal vasculature is non-specific. The treatment is largely supportive and there is no specific therapy. However, mortality of this condition remains high, if not timely intervened.

Key Words: *Amniotic fluid embolism. Cesarean section. Complication. Cardiac arrest.*

INTRODUCTION

Despite its recognition as a distinct entity for almost 100 years, the syndrome commonly referred to as Amniotic Fluid Embolism (AFE) remains one of the most devastating conditions in obstetrics.[1] AFE is a rare condition and a lethal complication of pregnancy and puerperium. Because of this very rarity, and the fact that it is a diagnosis of exclusion, the true incidence is difficult to determine. Combined with internationally differing definitions of AFE as well as differing methodologies for reporting and recording data, this results in a great range of reported incidence rates between countries. The International Network of Obstetric Survey Systems (INOSS) has recently produced a consensus definition of AFE as 'an acute cardio-respiratory collapse within 6 hours after labour, birth or ruptured membranes, with no other identifiable cause, followed by acute coagulopathy in those women who survive the initial event,' and has recommended its use in future.[2]

Its presentation varies from cardiac or circulatory shutdown to sudden death with varying degrees of multiple organ dysfunction. Approximately 47% of women who experience AFE have a prior sign or symptom before collapse. Commonly reported premonitory symptoms include tingling sensation or numbness, agitation or breathlessness, lightheadedness, chest pain or complain of feeling cold. Almost 36% of patients had fetal compromise before any change in maternal condition, likely due to compensated or unrecognised maternal hypoxia and hypotension. Cardiac arrest as a presenting feature is recorded in 60-80% of registered cases.[2]

AFE appears to involve an abnormal activation of humoral and immunologic mechanisms leading to a massive inflammatory reaction following the entry of fetal cells (antigens) into the maternal circulation.[3] It is largely a clinical "diagnosis of exclusion" and, therefore, requires high level of suspicion. At present, there is no any specific antemortem test for diagnosis of this syndrome. It is commonly diagnosed retrospectively and is primarily clinical diagnosis. Postmortem presence of fetal debris or squamous cells in the maternal pulmonary circulation remains a key diagnostic requirement, but is not found in every case.

The management of pregnant women with suspicion of AFE remains largely supportive and is directed at reversing any observable physiological abnormalities, with primary correction of oxygenation, coagulopathy and circulatory support. Once the patient presents with collapse, the basic principles of ABCd resuscitation should apply, with "d" representing delivery of the fetus in this case. A multidisciplinary team approach including senior obstetricians, midwives, anaesthetists, critical care team, intensivists, and haematologists is the key to success. Several studies have now highlighted the association between neonatal outcome and timing of delivery in relation to the onset of AFE. The overall UK perinatal mortality rate in AFE is reported as 67 per 1000. The morbidity of AFE for the baby is also reflected in high rates of NICU admission, ventilation, encephalopathy, and long-term morbidity.[2]

The recurrence rate of AFE is unknown because of the extremely low incidence and high mortality rate of the condition. No cases of recurrence have been reported in the literature, but many cases of straightforward

Department of Obstetrics and Gynaecology[1] / Pathology and Laboratory Medicine[2], The Aga Khan University Hospital, Karachi.

Correspondence: Dr. Nadia Nawaz, Senior Resident, Department of Obstetrics and Gynaecology, The Aga Khan University Hospital, Stadium Road, Karachi.
E-mail: dr.nadianawaz@hotmail.com

Received: August 30, 2017; Accepted: December 28, 2017.

subsequent pregnancies have been reported. So, whilst patients can be reassured to some degree when planning future pregnancies, they should also be counselled that the available sample size does not allow definitive conclusions regarding the risk of recurrence.[2] A recent, retrospective review by Dildy *et al*. found that out of 105 women who had survived AFE, only 22% had gone on to conceive again; and of those, who had not conceived, 61% had chosen not to do so due to a fear of recurrent AFE.

CASE REPORT

A 21-year healthy primigravida, height 163 cm, weighing 62 kgs, presented at 36 weeks of gestation in labour room with rupture of membranes since morning and mild labour pain. When received in our triage area, she was well oriented and vitally stable (blood pressure 120/80 mmHg and pulse 82 bpm). Fetal heart beat at the time of presentation was 140bpm and fetus showed a reactive cardiotocograph. On vaginal examination, it was 2 cm dilated, draining clear liquor.

Suddenly, patient had a generalised tonic-clonic fit with upward rolling of eyes and frothing from mouth. Fit was self-limiting, followed by sudden maternal collapse. She became pulseless and no blood pressures were recordable. Immediate cardiopulmonary resuscitation (CPR) was initiated and patient intubated. She revived but soon crashed again. A second episode of CPR was started and patient revived. Simultaneously, on per abdominal examination, there was sustained uterine contraction and fetal heart beat showed bradycardia of 65 bpm, which necessitated the need for cesarean section. Patient was rushed to operating room for emergency cesarean section with continuous bagging; and a limp baby boy of 2.9 kgs was born with an Apgar score of 2/1 and 7/5 min. Baby was handed over to pediatrician for resuscitation. The vital signs of the patient in the operation room (OR) were 100/60 mmHg of blood pressure, 150 bpm of heart rate, and 100% of oxygen saturation on high ventilatory parameters. Her arterial blood gases at the time of intubation were pH 6.78, PCO2O, 11.8, PO_2 13, and O_2 saturation 48%. She was administered norepinephrine infusion at 0.15 µg/kg/min for hypotension intraoperatively. Intra-operatively, patient developed uterine atony and started bleeding from uterine stitch line and developed excessive oozing, secondary to disseminated intra-vascular coagulation (DIC). Further laboratory tests were performed that showed the presence of severe coagulopathy with values of prothrombin time (PT) >120 s (normal 10.5-13.5 s), activated partial thromboplastic time (APTT) >120 s (normal: 21-36 s), INR >9.0, D-dimer 3.3 mg/L (normal <0.2 mg/L) and fibrinogen, 0.5 g/dl (normal; 1.5-4.0 g/dl), fibrinogen degeneration product (FDP) >20 µg/L (normal; 0-4 µg/L). Her platelet count was initially 104 x 109/l. Massive transfusion protocol was initiated and a total of 6 packed cell volumes (PCV), 10 fresh frozen plasma (FFP), 10 platelet, and 6 cryoprecipitates were transfused intraoperatively, in the immediate post-operative period. With immediate and rapid blood products replacement, uterine atony and bleeding from stitch line improved markedly and a condom catheter uterine tamponade was inserted with 400 ml of distilled water and vagina was packed with 2 ribbon gauzes. The total duration of operation was 3 hours; the urinary output was 800 ml with 3000 ml of blood loss. The fluid input was 2000 ml; and 3650 ml of blood was transfused. Her chest radiography showed increased hilar markings and possibility of pulmonary edema.

Postoperatively, patient was shifted to intensive care unit (ICU). Hemodynamic stability was achieved and her urinary outputs were satisfactory after mild initial hematuria. She remained in ICU for one day and was extubated on 1st postoperative day. Transthoracic echocardiography was performed which revealed normal cardiac chambers, ejection fraction of 50%, mildly reduced right ventricular systolic function, and moderate tricuspid regurgitation. Next day, she was shifted to special care unit. During ICU stay, MRI brain was performed showing no evidence of posterior reversible encephalopathy syndrome (PRES), cerebral venous sinus thrombosis (CVST). No additional significant abnormality was noted. CT did not demonstrate a pulmonary embolism. A diagnosis of AFE was made. Both baby and mother were discharged in stable condition on 6th postoperative day. On 2 weeks post-natal follow-up in clinic, both mother and baby were healthy and stable. There were no observed neurological sequelae as well.

DISCUSSION

AFE is notified among the five major causes of direct maternal deaths in the UK with other including eclampsia, pulmonary embolism, severe sepsis and peripartum haemorrhage.[4,5] It is a rare but devastating complication of pregnancy with incidence of approximately 1 in 40,000 deliveries.[1,3,6] The reported mortality rate of the condition ranges from 20% to 60%.[1,3] The diagnosis of AFE is largely a "diagnosis of exclusion".[3] Its pathogenesis can be broadly classified into three subtypes: (1) The mechanical obstruction subtype - which is the actual embolization of the maternal pulmonary vasculature,[3] (2) The anaphylactic subtype - in which there is anaphylactic reaction with idiosyncratic adverse response and massive postpartum hemorrhage, (3) The DIC subtype with a massive post-partum haemorrhage. Each subtype has a separate pattern of clinical signs and symptoms with varying severity of the disease. Wide range of mortality and survival is affected by the specific subtype.[7-9]

Management is largely supportive with an inter-disciplinary team effort that comprises prompt and

aggressive hemodynamic resuscitation including timely fluid and blood component replacement with correcting hemostatic disorders and provision of end-organ support with timely delivery of the fetus to aid in resuccitation and decreasing fetomaternal compromise.[3]

A team effort and multidisciplinary care can have profound effect on the outcomes. For the past 10 years, medical error and patient safety in the course of patient care has gained significant attention. Obstetric emergencies usually involve low risk and young patients and require a very efficient chain of multidiciplinary team effort to improve outcomes. A key to success is the timely intervention, sometimes referred to as "The Golden Hour" in the management. As in this case, timely recognition of the collapse with urgent involvement of the anesthesia team and cesarean delivery of the fetus within 5 mins of CPR were the hallmarks.

To be more precise, a very well coordinated and time-sensitive response to an obstetric emergency is required for improving outcomes. The role of local training programmes on individual basis in the management of maternal collapse cannot be denied and are conducted frequently. However, more recently, there has been a huge debate on simulation training in obstetric emergencies and the emerging results are worth it. It involves training of a multiprofessional team involved in the care rather than on individual basis and simulates an emergency without any harm to real patients.

Few articles, evaluating the effect of teamwork training with simulation models, have been published; and even fewer have examined objective measurements of improvement in the management of acute obstetric emergencies.[4] In a multicentre randomised clinical trial in The Netherlands, they found a significant improvement in team performance and a significant increase in the use of new medical technical skills 8 months after obstetric, multiprofessional team training in a medical simulation centre.[5]

There is paucity in the literature on the role of effective teamwork in the management of AFE owing to the rarity of its occurence. However, some case reports have been published internationally showing successful outcomes with prompt detection and aggressive management. A case reported in India in which a 27-year primipara had AFE during the peripartum period, in which due to expeditious CPR, both the mother and the newborn survived.[6]

Many survivors do present with some degree of neurological impairment. The rate of perinatal death with AFE approximates 25%, and 50% of surviving neonates are neurologically intact. A study in UK included women with a diagnosis of AFE from February 2005 to January 2014. It concluded that for every one woman who dies because of AFE, four survive. Among the surviving women with AFE, 7% display permanent neurologic injury. The women who survived with AFE without any permanent neurologic injury, 17% had other major morbidity.[9] For counselling regarding subsequent pregnancies, it is worth knowing that to date a total of 12 cases of successful pregnancy after survival from AFE have been reported in the literature with no case of recurrence of AFE.[10] A retrospective case study in Japan, including 10 AFE patients, showed a maternal mortality rate of 70%.[11]

AFE is rare but potentially devastating syndrome that manifests mainly during or around delivery. Immediate resuscitation and interdisciplinary approach, with active cardiopulmonary stabilization, hemodynamic correction, early vasopressor use, correction of hemostatic disorders and undelayed cesarean delivery, plays a important role in the outcome.

REFERENCES

1. Clark SL. Amniotic fluid embolism. *Obstet Gynecol* 2014; **123**:337-48.
2. Tuffnell DJ, Slemeck E. Amniotic fluid embolism. *Obstetrics, Gynaecol Reprod Med* 2017; **43**:627-32.
3. Balinger KJ, Lam MTC, Hon HH, Stawicki SP, Anasti JN. Amniotic fluid embolism: despite progress, challenges remain. *Current Opinion Obstet Gynecol* 2015; **27**:398-405.
4. Merién A, Van de Ven J, Mol B, Houterman S, Oei S. Multi-disciplinary team training in a simulation setting for acute obstetric emergencies: a systematic review. *Obstet Gynecol* 2010; **115**:1021-31.
5. Fransen A, Van de Ven J, Merién A, de Wit-Zuurendonk L, Houterman S, Mol B, *et al*. Effect of obstetric team training on team performance and medical technical skills: a randomised controlled trial. *BJOG* 2012; **119**:1387-93.
6. Meshram P, Maneesha HU, Doval J. Successful resuscitation following amniotic fluid embolism during emergency lower segment caesarean section: A Case Report. *J Thrombo Cir* 2016; **2**:106.
7. Nair M, Kurinczuk J, Brocklehurst P, Sellers S, Lewis G, Knight M. Factors associated with maternal death from direct pregnancy complications: a UK national case-control study. *BJOG: Int J Gynaecol Obstet* 2015; **122**:653-62.
8. Sadera G, Vasudevan B. Amniotic fluid embolism. *J Obstet Anaesth Crit Care* 2015; **5**:3.
9. Fitzpatrick K, Tuffnell D, Kurinczuk J, Knight M. Incidence, risk factors, management and outcomes of amniotic-fluid embolism: a population-based cohort and nested case-control study. *BJOG: Int J Gynaecol Obstet* 2016; **123**:100-9.
10. Buechel J, Berset A, Lehmann MA, Lapaire O. Unresponsive primipara after rupture of membranes. *BMJ Case Rep* 2015; bcr2015209765.
11. Yoneyama K, Sekiguchi A, Matsushima T, Kawase R, Nakai A, Asakura H, *et al*. Clinical characteristics of amniotic fluid embolism: an experience of 29 years. *J Obstet Gynaecol Res* 2014; **40**:1862-70.