INTRODUCTION

Vernix caseosa is a cheese-like white cutaneous substance covering the skin of a newborn which was observed to serve as protective layer inhibiting growth of staphylococcus aureus and klebsiella on the nutrient agar, thus it was suggested to be left dried on newborn skin. Some authors reported that VCP could be initiated by antenatal or intrapartum leakage of amniotic fluids from utero tubal reflux and or unrecognized uterine perforation. Spillage of amniotic fluids into the peritoneal cavity was almost inevitable during caesarean section and is usually insignificant, although some rarely progress to peritonitis. Theoretically, a hypersensitivity reaction might occur from previous pregnancy delivery, or from a previous antenatal event. However, as there were previous cases which occurred in primiparous patients, the mechanism is likely the latter. Higher concentration of vernix caseosa in amniotic fluid, observed in difficult labour or in oligohydramnios cases might had pathogenic significance. We present a case of VCP diagnosed in a post vaginal delivery who developed septic shock with atypical presentation of peritonitis, which was eventually diagnosed by histopathological examination. To date, this is the first case to be reported in Malaysia that we know of.

CASE PRESENTATION

Mrs M, a 29 years-old, Siamese, Para 3, with underlying bronchial asthma presented on day 6 post vaginal delivery which was complicated by a first degree tear. The antenatal period was uneventful. Her initial complaint was shortness of breath for 2 days preceded with lethargy and poor oral intake; otherwise she denied any history of fever or any infective symptoms. She had a history of contact with her husband who was diagnosed with covid-19 category 1. She was however tested negative. At the emergency department (ED), she was septic looking with her vital signs as follows: temperature 38°C, blood pressure of 93/50mmHg, heart rate 123-130 bpm, respiratory rate of 40/min but maintaining oxygen saturation of 100% on nasal prong 3L/min. She was resuscitated with 10ml/kg of crystalloids and then started on IV infusion noradrenaline requiring up to 0.15mcg/kg/min. Systemic examination noted that her lungs were clear on auscultation and her abdomen was mildly distended; however it was soft, not tender and not guarded on
palpation. Arterial blood gases showed severe metabolic acidosis (pH 7.36/pCO₂ 23.6/pO₂ 103/HCO₃ 16.7/BE -12.2/lactate 4.6) with acute kidney injury (AKI) as evidenced by urea 22.5mmol/L, creatinine 340umol/L and her septic parameters were: WCC 6.4, CRP 318. Her liver function was otherwise normal. This patient was then admitted to ICU and treated as Acute Exacerbation Bronchial Asthma (AEBA) secondary to Hospital Acquired Pneumonia (HAP) and was covered with IV Ceftriaxone 1g BD. Bedside scan showed minimal left pleural effusion with contracted gallbladder and presence of sludge was noted in the gallbladder.

CTPA was ordered in view of persistent tachycardia and tachypnoea which revealed bilateral collapse consolidation with minimal pleural effusion, presence of ascites, and no evidence of pulmonary embolism. She had persistent temperature spikes up to 41°C and haemodynamic instability requiring double inotropic support with fast AF (heart rate up to 160bpm) which did not resolve despite being loaded with IV amiodarone. She was then electively intubated as her condition further worsened with increasing inotropic support and worsening septic parameters (WCC 31.17x10⁳/uL, CRP 318mg/L). None of the cultures sent came back with growth. Since her condition showed no improvement after 2 days on Meropenem, she was started on Intravenous Immunoglobulin (IVIG) of 0.3g/kg due to the fact that her condition was thought of as toxic shock syndrome.

After 2 days course of IVIG, her clinical condition seemed to improve and she was weaned off the inotropic and ventilation support and resolving AKI was noted as well. However, her WCC was static at 27.3x10³/uL, CRP 301mg/L with low grade fever of 37.3°C. Repeated bedside scans noted free fluids in the pelvic region, bulky uterus with thin ET, and dilated gallbladder with sludge still present. The case was referred to the surgical team as septic parameters did not resolve despite improvement of clinical condition and the team decided to proceed with CT abdomen. The radiologist reported that there were thickened small bowel loops with gross ascites, the gallbladder was well distended with no stones seen within and other organs were normal.

Consequently the team decided for bedside diagnostic peritoneal tap and 200ml of cloudy yellowish peritoneal fluid was aspirated and sent for investigation. Fluids cultures came back as no growth and cytology shows suppurative inflammation with no malignant cells seen. Other investigations were as follows: (protein 6736 mg/L, glucose 3.4mmol/L, LDH 7338u/L, PMN 468) with APRI score 0.4 which fulfilled the Runyon’s criteria (protein>10 mg/L, glucose <2.8mmol/L, LDH>serum LDH, PMN>250ml). As all these diagnostic measures were exhausted, she was then taken for a diagnostic laparoscopy which was converted to midline laparotomy and peritoneal washout. Intraoperatively, the existence of 1.2 Litre pus in the abdomen was noted over all quadrants with moderate
sloughs at the pelvis. The pancreas appeared to be unhealthy and bulky, the retroperitoneum appears oedematous and sloughy, with the jejunum being mildly dilated. Pus and omental biopsy were taken and sent for histopathology. Histopathological examination of the peritoneal fluids sent showed the presence of anucleate squamous which raised a possibility of vernix caseosa peritonitis. Anucleate squamous cells might be present along with lanugo hair and foreign body giant cell reaction as mentioned in previous literatures depending on the time since the triggering inflammation, but not in this case.  

Antibiotics therapy. Adjuvant steroid therapy was used in two cases with resistant symptoms after infection had been excluded, postulating that it could significantly enhance the suppression of the inflammatory response facilitating the recovery in their case. However, all the treatment options remained empirical and controversial as none of them were really evidence based. A case reported in 1998 even cautioned that aggressive treatment with broad spectrum antibiotics might trigger an acute Clostridium Perfringens infection.  

As in our case, the patient came in with haemodynamic instability and organ involvement and IVIG was used in the treatment course instead of steroids; which helped to suppress the inflammatory response and calm the haemodynamic instability enabling us to proceed with the operation under lower mortality risk of general anaesthesia. IVIG is a pooled antibody and a biological agent used to manage various immunodeficiency state and other conditions including autoimmune, infections and inflammatory states. It was used to normalize a compromised immune system. Different IVIG doses (low vs high) were administered based on the indicated medical condition. Low dose immunoglobulins serve merely as a passive replacement in immunodeficiencies (category I). High dose immunoglobulins played an active part and modulate the immune functions with additional anti-inflammatory activity (category II). Adverse effects reported in 5% of patients and could occur within 30 to 60 minutes of the infusion, attributable to the excipients and stabilizers contained in the preparation. Fortunately in our case, none of the adverse effects were observed. VCP is the inflammatory response to the amniotic spillage, and a short course of IVIG has been shown to suppress the inflammatory reaction rather than the use of broad spectrum antibiotics. More studies are needed to help us unveil more of its potential in treating VCP thus providing a way to treat this conservatively in the future.  

CONCLUSION

Currently VCP is a rarely reported complication as it is mostly unknown to parts of the world. It may not only

Figure 4: diagnostic laparoscopic findings of pus over all quadrants of the abdomen

This patient was well enough to be discharged from intensive care 3 days post operation. She was cared for in the surgical ward for a week and was finally discharged home.

DISCUSSION

The principal symptoms of VCP are generalized severe abdominal pain, pyrexia, peritonism and elevated WCC with inconclusive or normal imaging. The case we presented however presented with nonspecific complaints, and the diagnosis was only made via histopathological examination. This kind of cases has been reported almost every year by different countries, hence the authors strongly feel it is important to raise awareness of the clinical signs of VCP particularly among surgeons which hopefully will help in avoiding unnecessary extensive excisional procedures. A study published in 2011 suggested that radiological imaging may identify lesions in the abdominal wall or peritoneal cavity, and the diagnosis can be made via image-guided biopsy. Various options of conservative treatment have been contemplated as reported before and some do advocate postoperative antibiotics therapy.
happen antenatally, but also during post vaginal delivery as reported by this case. The diagnosis is most often missed as it tends to mimic other types of acute peritonitis. The symptoms could be vague and patient might present at a later stage in overwhelming state and/or other complications. Late or missed diagnosis might lead to unnecessary procedures and morbidities. Further study will help to determine the risk factors contributing to its progress to peritonitis. As in our case, we too had not thought that the cause could be peritonitis triggered by post-delivery inflammation of vernix spillage. This patient not only had to undergo additional series of procedures as it was not identified during operation itself; her operation was subsequently converted to midline laparotomy and she ended up with complications that required longer hospital stay. Therefore, VCP should be kept in mind as one of the provisional diagnoses in antenatal or postnatal women with severe and unusual abdominal symptoms regardless of the mode of delivery.

REFERENCES