

Acknowledgements

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A rare cause of maternal death: liver and inferior vena cava rupture due to previously undiagnosed Ehlers-Danlos Syndrome type IV

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EDITOR:

A 23-yr-old primigravid female died 8 days post Caesarean section due to liver decapsulation and inferior vena cava rupture due to previously undiagnosed Ehlers-Danlos Syndrome type IV (EDS IV). Her past medical history included many features suggestive of a connective tissue disorder but the diagnosis was only made *post mortem*.

Case report

The patient was a 23-yr-old primigravid female. Her booking summary documented an operation for 'protruding eyes' and that her 18-yr-old brother had previously had a heart transplant.

She was admitted at term for induction of labour for intrauterine growth retardation. An epidural was sited and after an 8 h labour a Caesarean section was

performed for fetal distress. The epidural was topped up and anaesthesia deemed adequate. However, 5 min into the procedure she was given a general anaesthetic as she was experiencing pain. A healthy female baby was delivered. There was 700 mL blood loss and tissue friability was noted. The possibility of a collagen disorder was raised. Postoperative recovery was uneventful and she was discharged home 3 days later.

The patient was readmitted 5 days postoperatively with dull chest pain and shortness of breath on exertion. Arterial blood gas analysis revealed a mild respiratory alkalosis with a normal PaO₂ on air. Dalteparin was commenced for a presumptive diagnosis of pulmonary embolus. She then developed sharp right shoulder pain and signs of respiratory distress with hypoxia and tachypnoea. The admission blood results were documented at this stage, haemoglobin 10.6 g dL⁻¹ and elevated serum transaminases, bilirubin and alkaline phosphatase. The obstetric registrar noted an increasing abdominal girth but a portable ultrasound scan revealed no ascites. The differential diagnoses of HELLP syndrome and acute fatty liver of pregnancy were discussed with the regional liver

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unit. Transfer was agreed for when a bed became available.

An hour later there was a sudden deterioration in her condition. She became clammy, tachycardic, hypotensive and tachypnoeic. She developed right upper quadrant pain and was admitted to the ICU. Fluid resuscitation was commenced after difficulty in obtaining adequate intravenous (i.v.) access due to venous fragility. A diagnostic peritoneal tap was performed and frank blood was aspirated. Her haemoglobin was now 6.3 g dL^{-1} and there was further increase in her abdominal girth.

Laparotomy 21 h after admission revealed a large subcapsular liver haematoma and several deep hepatic tears. The liver was packed and blood loss estimated at 3000 mL. Despite transfusion of large volumes of blood, fresh frozen plasma and cryoprecipitate, she remained cardiovascularly unstable but was returned to the ICU while awaiting the arrival of the hepatobiliary surgeon from the regional liver unit.

A second laparotomy revealed ongoing bleeding due to a decapsulated liver secondary to the subcapsular haematoma. There were also two splenic tears and subserosal haemorrhages throughout the small bowel. Intraoperatively she developed pulseless ventricular tachycardia, which degenerated into asystole. After 2 mg of i.v. epinephrine and 6 min of cardiopulmonary resuscitation, the rhythm changed to a narrow complex tachycardia, which was successfully cardioverted with a single 200 J DC shock. Haemostasis was achieved with packing and she was returned to the ICU. By this stage she had received 42 units of packed red cells and large volumes of platelets, cryoprecipitate and fresh frozen plasma. She had been given vitamin K, tranexamic acid and aprotinin. She required high levels of inotropic support and was profoundly acidotic.

Over the course of the night her cardiovascular stability improved, the lactate normalized and the acidosis began to resolve. The following morning she was transferred to the regional liver unit where her liver was inspected and repacked. She developed bilateral pneumothoraces and chest drains were inserted. There was no further active bleeding from the liver but lengths of necrotic small bowel were resected. Overnight she was successfully weaned from all inotropic support.

The following morning the decision was made to perform a fourth laparotomy for removal of packs and reinspection of the ischaemic gut. Upon reopening the abdomen, the liver was found to be completely decapsulated and bleeding uncontrollably. A Pringle manoeuvre was unsuccessful in achieving haemostasis and she became asystolic. This was followed by 30 min of cardiac massage and transfusion of 18 L of blood. There was a brief episode of ventricular

fibrillation, which was electrically cardioverted to pulseless electrical activity. Hepatectomy was performed but at this point it was noted that the suprahepatic inferior vena cava had been avulsed from the right atrium and cardiac massage was abandoned.

Discussion

EDS is a heterogeneous group of connective tissue disorders characterized by skin and joint laxity and tissue fragility [1,2]. However, type IV is further characterized by the risk of arterial, intestinal and uterine rupture [1,3], resulting in a median survival of 48 yr in those affected [3].

EDS IV results from mutations in the gene for type III procollagen (COL3A1) and is inherited in an autosomal dominant manner. However, 50% of cases result from spontaneous mutation [1], which makes diagnosis of an uncommon disease even more difficult. As a result, diagnosis is often only made at *post mortem* or after major complications [3].

The four major clinical diagnostic criteria on which the diagnoses are based are uterine, intestinal or arterial rupture, easy bruising, thin skin with visible veins and characteristic facial features (thin lips and philtrum, small chin, thin nose and large eyes [3]). Minor clinical diagnostic criteria include acrogeria, hypermobility of small joints, tendon or muscle ruptures, carotid-cavernous sinus fistulae, pneumothorax, chronic joint subluxations or dislocations, talipes equinovarus, gingival recession and cervical spine instability [3,4]. The diagnosis is confirmed by demonstrating that cultured fibroblasts synthesize abnormal type III procollagen or by identification of a mutation in the gene for type III procollagen (COL 3A1) [3].

Review of this patient's hospital notes (not available until after her transfer to the liver unit) contained many clues about her diagnosis. The patient was born with bilateral talipes and had flexion deformities of her fingers. She had displayed retarded motor development. At the age of 13 she fell down the stairs and the severity of the periorbital swelling and bruising caused a consultant ophthalmologist to question the mechanism of injury. At the age of 15, she had a magnetic resonance (MR) scan in order to investigate her dysmorphic appearance, primarily bilateral proptosis. This was normal, as was karyotyping. She subsequently underwent corrective surgery to her eyelids. Friability of the tissues was noted during her Caesarean section. Her unusual facial features and the difficulty in obtaining i.v. access were commented on while an inpatient on the ICU.

It is not possible to be didactic about best practice in management of pregnant women with this condition due to lack of evidence. Evidence that

exists is case report or patient cohort based. It has been reported that pregnancy should be avoided in those affected by EDS type IV, even to the extent of recommending termination [2,4,5].

Vaginal delivery may be hazardous as uterine contractions and pushing may increase the risk of uterine rupture. The increase in intra-abdominal pressure and blood pressure associated with pushing may result in hollow viscus or vessel rupture respectively [4,6]. Instrumental delivery may help reduce these risks but may result in an increased risk of perineal trauma. Haemostasis and repair may then pose major problems [6].

The use of epidural analgesia may reduce excessive maternal effort and also the sympathetic responses to pain but neuroaxial blockade may itself be harmful. Firstly although standard coagulation tests are usually normal, clotting factor deficiencies and platelet abnormalities have been described [4,6]. Epidural veins are likely to be more fragile and there is a theoretical increased risk of epidural haematoma.

Caesarean section may be advocated as a more controlled manner of delivery but there may be difficulty in achieving haemostasis due to tissue fragility. The choice of anaesthesia is also not clear. Spinal anaesthesia may not provide sufficient duration of block if problems with haemostasis are encountered and so combined spinal and epidural may be beneficial [4]. However, if major haemorrhage is encountered, profound sympathetic blockade may have a deleterious effect on the outcome for the patient. A general anaesthetic may therefore provide a more controlled environment. A hypertensive response to intubation should be avoided

and manipulation of the neck must be done with care due to potential cervical spine instability [4,6].

This case is reported to raise awareness of a rare and potentially devastating disease that may first present during pregnancy. It is not possible to offer recommendations as to the best management of a pregnant woman with EDS IV but awareness of the issues concerning delivery may help a multidisciplinary team achieve a plan that minimizes potential risks and is acceptable to the mother and her partner.

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Regional anaesthesia for Caesarean section in an ankylosing spondylitic patient with twin pregnancy

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EDITOR:

Ankylosing spondylitis can pose several anaesthetic and obstetric problems during pregnancy. This disease presents a unique scenario of difficult airway and difficult 'back'. Ankylosing spondylitis is a pre-

dominantly axial form of arthritis that usually begins in the sacroiliac joints and slowly progresses to spinal fusion. It presents initially with chronic lower back pain or stiffness. The estimated male to female ratio is between 4:1 and 6:1 [1–3] with HLA-B27 antigen association. As the peak incidence is in the young adult population [2,3], pregnancy may occur.

Case report

A 33-yr-old female patient with a 5-yr history of ankylosing spondylitis who was taking sulphasalazine

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