Klippel Trenaunay Weber syndrome in pregnancy

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Klippel Trenaunay Weber syndrome (KTW) is a rare disorder, which occurs, in one in 27500 live births.[1] It is a congenital neuroectodermal disorder[1] and is characterized by the triad of symptoms such as port wine stain, bone and soft tissue hypertrophy and varicose veins. It is also associated with arteriovenous malformation.[2] Due to the complexity of this disease, it is a real challenge to the anaesthetist and the obstetrician.

We report a case of a KTW syndrome patient in her first pregnancy who was transferred to us from a district general hospital for the delivery. She had all the features along with multiple vascular anomalies and thrombocytopaenia. She underwent an elective caesarean section under general anaesthesia. Her intraoperative period was uneventful. At her post-operative day one she had a sudden cardiac arrest due to massive pulmonary embolism. This case highlights the high risk of thromboembolic events in patients with KTW syndrome.

Keywords: Klippel Trenaunay Weber syndrome (KTW); thrombocytopaenia; thromboembolism

Case report

An 18-year-old teenage primigravida was transferred to our tertiary centre from a district general hospital at POA of 40+2 for delivery. She had a large port wine stain on the left side of the neck. She had a lymphohaemangiomatous malformation on the right leg with soft tissue hypertrophy. It was associated with varicose veins which were extending from ankle to pelvis.

She also had disproportionate growth of digits along with macrodactyly of the right foot since birth. But her activities of daily living were not restricted due to disease. Her systemic and airway examination was normal.

She also had severe kyphoscoliosis along with cephalopelvic disproportion. She was never investigated for neurological or vascular malformations. Therefore, with input from the neurology and orthopaedic teams, we planned to proceed with elective caesarean section. Her cardiovascular assessment done during the first trimester was normal. She didn't have any other medical problems. However, she was not investigated for neurological or vascular malformations as she had defaulted antenatal clinic visits.

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She was found to have thrombocytopenia (platelet count <50,000) during pregnancy and her blood picture showed large giant platelets and rest of the coagulation profile was normal.

After obtaining high risk-informed consent and routine preoperative preparation with fasting and antacid prophylaxis (ranitidine 50mg and metoclopramide 10mg orally 8hrs and 2hrs preoperatively) and ensuring the availability of blood and blood products, she was taken up for surgery. Her preoperative platelet count was 35000 and six units of platelets were transfused prior to surgery according to haematology advise.

After establishing monitoring, modified rapid sequence induction was done with fentanyl 100µg, propofol 120mg and suxamethonium

100mg. Size 7.5mm endotracheal tube was inserted and anaesthesia was maintained with oxygen, N_2O and isoflurane. A live baby girl weighing 3.1kg was delivered by lower segment caesarean section and syntocinon 5 units bolus followed by infusion was started after delivery. Analgesia was provided with morphine 7.5mg intravenously. Due to the high risk of bleeding tranexamic acid 1g bolus was given during surgery.

The intraoperative period was uneventful. Only minimal blood loss was noted during surgery. The patient was extubated and transferred to the intensive care unit for post-operative observation.

In the intensive care unit, mild oozing was noted from the surgical site and a tight dressing was applied. Syntocinon infusion 40u was given over 4 hours to minimize bleeding. The patient had persistent thrombocytopaenia with a platelet count of 10,000. Blood picture showed giant platelets. Her coagulation profile was within the normal range. However, thromboprophylaxis with low molecular weight heparin was not started due to the risk of bleeding.

Next day she was complaining of persistent abdominal pain. Her haemoglobin count dropped from 11.2g/dl to 8.2g/dl. Examination revealed a left-sided abdominal mass. Ultrasound scan showed multiple cystic spaces in subhepatic and multiple cysts in the spleen. Since her clinical picture was suggestive of bleeding, a decision was made by the surgeon to go ahead with splenectomy to minimize bleeding.

When trying to mobilize, the patient's eyes rolled up and she developed sudden haemodynamic instability followed by a cardiac arrest and she passed away in spite of efforts for resuscitation. An autopsy revealed a massive acute pulmonary embolism resulting from acute right leg thrombosis.

Due to the conflicts in the family, they refused to take over the baby and she was looked after in the hospital for 6 months and was handed over to the Department of Child care and probation.

Discussion

Only a few case reports have been published in patients with KTW syndrome in pregnancy and none of the cases reported are from Sri Lanka. Usually, in these patient's pregnancy is discouraged due to the risk of thromboembolism and haemorrhage.[3]

The exact pathophysiology of this disease is not elucidated. Bliznak and Staple suggested that damage to sympathetic ganglia or intermediolateral tract during the intrauterine period can lead to anomalies in deep veins. McGrory believes that both ectodermal and mesodermal dysplasia will lead to KTW syndrome.[1]

This disease has many implications for anaesthetists. We avoided central neuraxial block in this patient as we have not excluded haemangiomas, spinal arteriovenous malformations prior to surgery. She also had unexplained thrombocytopaenia which can lead to spinal haematoma.[4] However, KTW syndrome is not an absolute contraindication for central neuraxial block after excluding pathological anomalies.

These patients can have difficult intubation due to associated facial anomalies such as soft tissue hypertrophy and upper airway angioma. Since our patients' airway examination was normal we proceeded with general anaesthesia. Another major issue is that they are at high risk of bleeding due to associated factor deficiencies, consumptive coagulopathy and thrombocytopaenia.

Persistent thrombocytopaenia was noted prior to pregnancy. Even though this is possible in this syndrome, we could not investigate further as she had defaulted clinic visits.

Other than the three typical clinical features they will also present with abnormalities in deep veins such as hypoplasia, aplasia, and aneurysmal dilatation and absent or incompetent valves.[1] This usually affects a single extremity. The most common site is the lower limb followed by arms, trunks and rarely the head and neck.[1] Our patient had a hypertrophied right leg along with varicose veins.

They can have haemangiomas of variable depth. It can be either limited to the skin or can extend deeper to tissues including muscle and bone.

Visceral organs such as spleen, liver, bladder and colon also can be affected. These organ involvements can cause greater morbidity due to internal bleeding. This can manifest as haematuria and haematochezia. Our patient had oozing along with dropping of haemoglobin during the postoperative period. Her ultrasound examination showed multiple subhepatic cysts and multiple cysts possibly in the spleen. Therefore, internal bleeding was suspected and emergency laparotomy was planned to arrest bleeding.

Pregnancy and the puerperium are established risk factors for venous thromboembolism. This occurs in one in 1600 pregnancies.[5] 50% of these patients are at high risk of venous thrombophlebitis and 22% experience pulmonary thromboembolism and subsequent pulmonary hypertension and right ventricular failure.[4]

Acute pulmonary embolism can lead to catastrophic cardiovascular collapse. Platelet activation and the platelet aggregation are the key events in thrombus formation and vasoconstriction. Ultimately once thrombus is formed it will lead to disseminated intravascular coagulation due to increased fibrin degradation. Occasionally patients with massive acute venous thromboembolism (VTE) will develop thrombocytopaenia.[6]

RCOG guidelines on reducing the risk of thromboembolism highlight the importance of prophylactic low molecular weight heparin in women with high-risk factors. However, it was a debatable issue in the index case during the perioperative period due to associated thrombocytopaenia and risk of bleeding.

Conclusions

Syndromic patients bring out a major challenge to the anaesthetist throughout delivery. Therefore, to get a better result the underlying problems of these patients should be evaluated extensively prior to surgery. It is important to bring the patient's problems into multidisciplinary team discussion and very careful handling should be done with necessary precautions prior to any interventions.

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