**Vascular micro-thrombotic disease in pregnancy**

Sir,

Vascular micro-thrombotic disease (VMTD) includes conditions like thrombotic thrombocytopenic purpura (TTP) and TTP-like syndrome and is associated with adverse outcomes if not promptly diagnosed and treated. Challenges in diagnosing TTP-like syndrome stem from a vague presentation and overlapping features with conditions like pre-eclampsia, HELLP and disseminated intravascular coagulation (DIC). We discuss a case of a pre-eclamptic patient with thrombocytopenia in the postnatal period.

A 39 weeks pregnant patient with controlled pre-eclampsia was admitted for labour. Assisted foetal delivery resulted in third-degree perineal tear which was promptly repaired. Placenta was delivered intact. Slow bleeding continued post-delivery despite uterotonic drugs and a firm fundus. Haemoglobin dropped to 6.6 g/dL and platelets to 101 × 10⁹/L. Coagulation studies showed prolonged prothrombin time (PT), partial thromboplastin time (PTT) and fibrinogen dropped from 200 to 92 mg/dL.

Thromboelastography showed no fibrinolysis. Hepatic function panel was normal. Packed red blood cells, fresh frozen plasma and platelets were given. Blood loss continued, prompting emergent dilatation, curettage and Bakri balloon insertion. Persisting bleeding prompted bilateral uterine artery embolisation. Bleeding was controlled and haematocrit stabilised. The next day, platelets dropped from 58 to 31 × 10⁹/L without active bleeding. Patient remained afebrile and haemodynamically stable. Peripheral blood smear showed few schistocytes. LDH measured >3000 IU/L while fibrinogen and fibrin degradation product levels were normal. Platelets reached a nadir of 13 × 10⁹/L.
Immediate plasmapheresis and steroid yielded improved platelet counts.

Von-Willebrand factor cleaving protease: A disintegrin and metalloproteinase with thrombospondin type 1 motif, member 13 (ADAMTS13) level, and antibodies were normal. Plasmapheresis with steroids was continued till the platelets reached a goal of $>100 \times 10^9/L$.

VMTD is an emerging category of microvascular and macrovascular systemic thrombotic diseases. It includes TTP, TTP-like syndrome, haemolytic uremic syndrome (HUS) as well as other conditions like DIC. TTP/HUS is a diffuse intravascular microthrombotic (DIT) condition exhibiting thrombocytopenia and microangiopathic haemolytic anaemia (MAHA) due to ADAMTS13 protease deficiency. This can arise from congenital deficiency or from acquired antibody formation against ADAMTS13. The kidneys and the brain are the most commonly affected organs.[1]

Pregnancy can be associated with several thrombotic microangiopathies like HELLP, Acute Fatty Liver of Pregnancy (AFLP), DIC, pre-eclampsia and HUS with similar haematomorphology and overlapping features making diagnosis difficult.[2,3]

Endotheliopathy associated with insults like trauma, surgery or infection leads to inflammation and diffuse microthrombotic pathway activation. Platelet activation leads to exocytosis of endothelial ultra-large Von Willebrand Factor (eULVWF) release and formation of microthrombi.[1]

In TTP, the ULVWF are released into circulation affecting kidneys and brain.[1] In TTP-like syndrome, ULVWF are endothelium synthesised (eULVWF) which stay attached to the endothelial cells and, hence, can affect a variety of organs. TTP-like syndrome has the same haematologic manifestations of DIT, MAHA and thrombocytopenia but with minimal haemolysis and, hence, scant schistocytes on peripheral blood smear making it easy to miss the diagnosis.[1]

Plasmic score is a simple, validated clinical scoring system that assists in early diagnosis of TTP; however, it may not diagnose TTP-like syndrome where renal involvement is not always observed, and aetiology is multifactorial.[6]

The atypical presentation and the lack of involvement of typical organs alongside scant schistocytes may cause the diagnosis to be missed and delay treatment.[5]

While plasmapheresis is effective in TTP, in TTP-like syndrome, only early plasmapheresis is effective.[6]

Considering the urgency of plasmapheresis initiation in TTP-like syndrome, the diagnosis may rely heavily on the clinical picture and a high index of suspicion. While TTP diagnosis is well known and scored, TTP-like syndrome is a condition with varied risk factors and subtle presentation with poor outcomes. Unlike other conditions, haematopathology may not be reliable for diagnosis and a low threshold for clinical diagnosis is mandated.

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Conflicts of interest
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Airway management of a near obstructive vallecular cyst in an infant

Sir,

Congenital valleular cyst is a rare, benign condition but carries a potential threat of hypoxia and death if not managed appropriately. It is associated with laryngomalacia in 90% of cases. Airway management in these patients is very challenging because of difficult mask ventilation, laryngoscopy, supraglottic device insertion, and fiberoptic intubation.

A 3-month-old male child weighing 5 kg presented with complaints of cough, noisy breathing, and chest retractions for the last 2 months. On examination, the patient was tachypnoeic and having decreased air entry, inspiratory stridor, and intercostal recession. X-ray was done, which showed a hyperdense protrusion above the epiglottis [Figure 1, Panel a]. Nasendoscopy showed a large vallecular cyst abutting the epiglottis, which was completely obscuring the view to the larynx [Figure 1, Panel b]. The laryngeal inlet was opening for less than a second when the patient was trying to cry [Figure 1, Panel c, Video 1]. Patient was posted for vallecular cyst excision under general anaesthesia. Premedication was avoided in this patient due to the risk of airway obstruction in the preoperative room. Anticipating difficult airway, a backup plan of tracheostomy with the surgeon and resuscitation equipment was kept ready. Sedation was started with intravenous dexmedetomidine 1 mcg/kg bolus over 10 min followed by an infusion at the rate of 0.5 mcg/kg/h. The patient was induced with sevoflurane with maintenance of spontaneous ventilation to achieve a minimum alveolar concentration (MAC) of 1 to 1.2. A nasopharyngeal airway of size 3 mm internal diameter (ID) was inserted from left nostril and anaesthesia circuit was connected to it with end tidal CO2 monitoring. Intubation was then tried from right nostril with a fiberoptic bronchoscope (FOB) of size 2.8 mm (outer diameter), loaded with a 3.5 mm ID cuffed endotracheal tube. However, the laryngeal inlet was not visualised due to falling back of the vallecular cyst under the effect of general anaesthesia. Maneuvers like jaw thrust and tongue pulling were tried during fibroscopy but in vain. Cyst aspiration was then done with 26 G Quincke spinal needle under light laryngoscopy. Plane of anaesthesia was maintained by achieving a MAC of 1.5 and depth of anaesthesia was monitored clinically by the variability of vital signs and immobility. After aspiration of 3 ml of cyst fluid, the laryngeal inlet was visible and we were able to intubate the trachea. After securing the airway, the cyst was marsupialised with cyst wall excision. Aryepiglottopexy was done in the same sitting to prevent laryngomalacia. After securing haemostasis, extubation was performed on table. Postoperative period was uneventful and the patient was discharged after 2 days.

Preoperative assessment of these patients is of utmost importance and should include a detailed history and examination to identify obstructive symptoms. Computed Tomography (CT) scan, Magnetic resonance Imaging (MRI), and nasendoscopy are valuable diagnostic methods to determine the size, location, and contents of the cyst to plan anaesthesia and surgical management. Prenatal diagnosis of vallecular cyst with ultrasonography and MRI has been reported and earlier diagnosis allows planning perinatal management. Various methods of securing the airway have been used in the past depending upon...