Intra-operative vascular injury and its management in a case of Ehler-Danlos syndrome

Sir,

We report a case of 10-year-old male child of Ehler-Danlos syndrome with bilateral undescended testes (cryptorchidism), scheduled for bilateral orchidopexy, whose surgery got complicated due to vascular rupture.

Patient (25 kg) had facial dysmorphism with slender face, prominent bones, sunken cheeks, thin nose and bulging eyes. Past history of delayed cry at birth and being extremely floppy was noted. and patient had hypermobility of joints and kyphoscoliosis in D4 to L3 vertebrae. Investigations, including echocardiography was unremarkable. His coagulation profile was also within normal limits and there was no history of abnormal bleeding after minor trauma.

Patient was accepted for the procedure with informed consent and half tablet of midazolam 7.5 mg was given a night before. Intravenous (i/v) access was secured by 20 G cannula and ringer lactate started. Non-invasive blood pressure, electrocardiogram (lead II and V) and pulse oximeter were attached.

Injection glycopyrrolate 0.2 mg i/v and injection fentanyl 50 μg i/v were given. Patient was induced with propofol 50 mg i/v and was intubated without difficulty with 6.0 mm (ID) cuffed endotracheal tube with 15 mg i/v atracurium. Patient was maintained on isoflurane (0.4-1%), N_2O (66%) and O_2 (34%) with top ups of atracurium (2.5 mg), when required.

When surgeon mobilised the vas and applied traction whole surgical field was filled with blood which was due to lacerations of both femoral vessels, which were then clamped. Immediately second IV line was secured with 18G cannula, blood sent for cross match and patient was catheterised. Vascular surgeons were summoned and in the meantime central venous pressure (CVP) line was placed. On table it was decided by vascular surgeons for end-to-end anastomosis of femoral artery. 2000 U of heparin was given but procedure failed twice due to increased friability of the vessel. Thereafter, it was decided to take a graft from the saphenous vein and anastomosis was performed with the help of this graft. Furthermore, due to increased friability of femoral vein, it was decided to ligate it. Three units of blood and one packed red blood cell were given and CVP was kept within the normal limits in 8 h long surgery. Patient was extubated fully awake after reversing neuromuscular blockade with inj. neostigmine and glycopyrolate. Heparin 1000 U was given and patient shifted to intensive care unit for further monitoring and post operative period was unventful. At 1 month follow-up, the patient had normal limb

perfusion and no vascular sequelae. His limb mobility was preserved with good function.

Ehlers-Danlos syndrome is a disorder of connective tissue and can be classified into at least ten types on the basis of clinical, genetic and biochemical information.[1] The fragile skin and loose joints is often a result of abnormal genes that produce abnormal proteins which confer an inherited frailty of collagen. Among the different forms of Ehlers-Danlos syndrome, type IV has a high incidence of vascular damage and is also known as vascular Ehlers-Danlos syndrome, possibly present in the present case, though without the history of excessive bleeding or bruising.

Patients with Ehlers-Danlos syndrome type IV have been classically described to have four distinctive features:[1] characteristic faces (long thin nose and lips, sunken cheeks and bulging or protruding eyes), very thin and translucent skin, vascular fragility (tendency to bruise easily and rupture of vessels, especially the middle sized arteries) and rupture of viscera such as intestine and uterus.[2,3] Spontaneous rupture of arteries is the most common presenting symptom.[2] Most patients develop these complications before the age of 40. Median age in these patients is 48 years.[4] Cryptorchidism is frequently associated with connective tissue disorder as the guiding descending track is defective.[5]

Venous fragility can cause excessive blood loss, hence adequate venous access, preferably elective central line should be ensured before induction. If regional anaesthesia is chosen, spinal anaesthesia has to be performed carefully to reduce chances of vascular trauma and it is possible that epidural anaesthesia also can lead to significant bleed in these cases. During general anaesthesia, there should be gentle laryngoscopy and intubation. Laxity of ligaments may increase chances of atlanto-axial dislocations during forceful laryngoscopy. Laryngeal mask airway can cause increased pressures in pharyngeal areas, which can lead to soft-tissue damage and vascular ooze.[6] The positioning of patients is important with emphasis on adequate padding of joints as skin trauma and also nerve palsies can occur.

Careful and precise surgical technique with meticulous haemostasis and availability of a vascular surgeon is mandatory in these cases. High risk consent for vascular complications and subsequent critical care should be taken. Good venous access before induction and extreme care during regional anaesthesia is important, especially avoiding epidural anaesthesia.

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