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Case Report

Anaesthetic management of type II abernethy malformation posted for endovascular device closure

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ABSTRACT

Abernethy malformation is a rare congenital vascular malformation in which anomalous communication is seen between portal and systemic circulation. While treating the main goal is to prevent shunting of portal blood into the systemic circulation and preserving hepatic blood flow. This can be achieved surgically or non-surgical method. We report anaesthetic management of non-surgical closure of type II Abernethy malformation of 10 year old female child who had undergone ostium secundum ASD (atrial septal defect) patch closure.

1. Introduction

John Abernethy in 1793 described this malformation first time, when 10 month old child died of unknown cause. During post mortem examination he could find multiple congenital anomalies including, dextrocardia, polysplenia, portal vein joining inferior vena cava and transposition of great vessels.[1] Two variations are reported in this anomaly.[2] Type I includes congenital absence of portal vein with complete diversion of portal blood into inferior vena cava (IVC). This furthered classified as, type Ia in which superior mesenteric (SMV) and splenic vein (SV) separately drains into systemic veins and in Ib both SMV and SV join to form a short extra-hepatic portal vein (PV) and ultimately draining into IVC. Type II includes Presence of hypoplastic PV and portal blood is diverted into IVC through a side-to-side, extrahepatic communication.[3] The main key is to preserve portal blood flow to liver as much as possible while closing the shunt between portal and systemic circulation.[4] Very few pediatric cases have been reported from India.[5,6] As this is a rare entity and no reports are available on anaesthetic management of such a case for endovascular device closure hence it was challenging. Here we report anaesthetic management of type II abernethy malformation posted for endovascular closure who had undergone ASD correction.

2. Case presentation

A 10-yr-old 25 kg child presented to our hospital, Dr Vithalrao Vikhe Patil Pravara Rural Hospital, Maharashtra, India, with the chief complaints of breathlessness on exertion suggestive of grade II NYHA (New York Heart Association) class and bluish discoloration of all fingers and

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toes since 4-yrs. She was diagnosed as having large ostium secundum ASD when she was 3-yr-old and pericardial closure was done in different hospital. At the age of 5-yr she started developing dyspnea, bluish discoloration of fingers and toes, which was insidious in onset and gradually progressive and developed jaundice at the age of 8yr. On arrival, she was conscious, oriented, respiratory rate of 30/min, heart rate of 120 beats/min, blood pressure was 90/60mm of Hg, room air saturation was 75% with central and peripheral cyanosis, pandigital clubbing of grade2, yellowish discoloration of sclera and no peripheral edema noticed. On cardiovascular system examination S1, S2 heard with no murmur. Other systems were within normal limits.

With history and physical examination a list of differential diagnosis viz, TAPVC (Total anomalous pulmonary venous connection), IVC draining to LA, Severe PAH with opening of PFO (patent foramen ovale), Pulmonary AV fistula and communication between right or left pulmonary artery to LA was made.

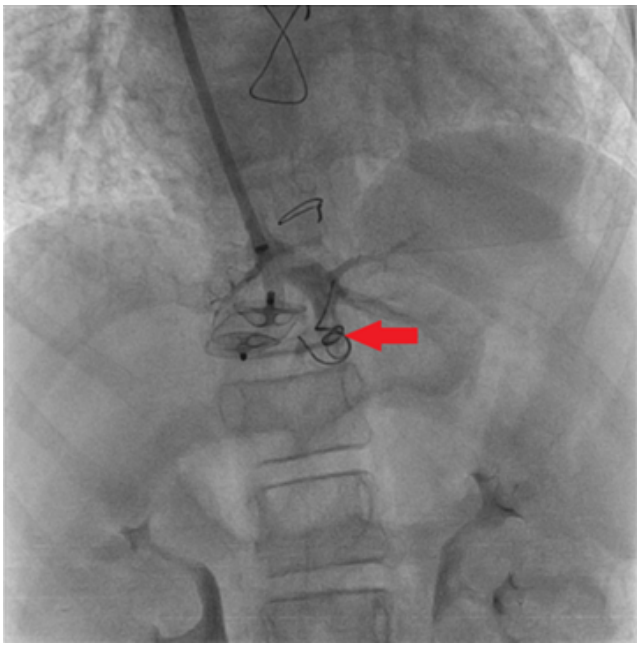

Fig. 1: A small branch connecting IVC and portal vein which was occluded with coil (Red arrow)

Blood investigation showed haemoglobin of 12 gm%, normal renal function test. Liver function test showed minimal rise in AST 48 IU/L and ALT 46 IU/L; alkaline phosphatase 111 IU/L; total bilirubin and indirect bilirubin were 3mg/dl and 2.2 mg/dl respectively; prothrombine time 15sec (test) 16sec (control), INR of 1.3. Chest x-ray showed artifacts of sternal wire. Echocardiogram showed an intact atrial septum, no leak, no patent ductus arteriosis, and good biventricular function. Agitated saline was injected through peripheral vein and by 4 cardiac cycles it was detected in left (LA) and left ventricle (LV) suggestive of pulmonary AV communication.

Ultrasound abdomen showed a small hemangioma of spleen; congenital portosystemic shunt connecting left portal vein to inferior vena cava, CT abdomen angiogram confirmed the diagnosis of abnormal porto-systemic shunt. CT brain showed early changes of encephalopathy.

Patient was electively posted for endovascular device placement to close the shunt between portal vein and IVC through right internal jugular vein approach. Child received premedication of Tab.Ranitidine 75 mg, Tab.Midazolam 3.5 mg in the night before surgery and kept nil by mouth for 6 hour before procedure.

All the standard monitors like electrocardiogram, noninvasive blood pressure (NIBP), pulse oximetry were connected. A 20 G intravenous cannula was secured and ringer lactate was used for the maintenance fluid. Inj Glycopyrrolate 0.004mg/kg, Inj. Fentanyl 1mcg/kg, Inj Midazolam 0.03 mg/kg were given. After the preoxygenation for 3 min, child was induced with Inj Ketamine 2mg/Kg, Inj. Atracurium 0.5mg/Kg and intubated with 6mm cuffed endotracheal tube. Anaesthesia was maintained with O2: air in 1:1 ratio with sevoflurane end tidal concentration of 2%. Invasive lines; right femoral artery, femoral vein and right internal jugular vein were secured by cardiologist and pressures were monitored. Throughout the procedure the systolic B.P of 80-100 mm Hg, diastolic B.P of 60-80 mm Hg and CVP of 9 mmHg were maintained. Many a times ECG showed ventricular ectopic which were due to guide wire manipulation and resolved on its own. Nasopharyngeal temperature was monitored and maintained between 36-37^{o}C.

Through right internal jugular vein (IJV) approach porto-systemic shunt was identified and the large communication at the level of RA and occluded with device. A small branch was also identified connecting IVC and portal vein which was occluded with coil (Figure 1). Procedure was completed in 1hr with minimal blood loss. Throughout the procedure the saturation was maintained 96% before and 100% after the device closure.

Patient was extubated after giving reversal Inj. Glycopyrrolate 0.2mg, Inj. Neostigmine 1.25 mg I.V and after meeting all extubation criteria and shifted to high dependency unit. Post-procedure patient was stable.

Patient maintained saturation of 88 % with room air and 100 % with FiO2 of 30%. The cyanosis disappeared from all the digits after the closure of shunt. With subsequent days her room air saturation improved to 92%. On follow up after one month, work tolerance of the child was improved; there was absence of icterus; cyanosis and clubbing. The room air saturation improved to 95%. This case is presented after obtaining written informed consent from the patient.

3. Discussion

Abernethy malformation is a congenital porto-systemic shunt that results from persistence of embryonic vessels which is an extremely rare anomaly.[7] There are two types of Abernethy malformation and treatment option depends upon the type of malformation. In patients with type I malformation shunt closure is not an option as it is the only drainage route for the mesenteric venous blood which requiring meticulous biochemical monitoring or liver transplantation.[8] In patients with type II anomaly, surgical or percutaneous trans-catheter endovascular closure is recommended to arrest mesenteric blood entering systemic circulation.[4–9]

It is associated with hepatic encephalopathy, due to porto-systemic shunting, hepatic masses, focal nodular hyperplasia (due to absence of the portal vein), hepatocellular carcinoma, hepatoblastoma. This can develop due to decreased clearance of neurotoxins from liver, e.g., ammonia.[9,10] Due to lack of facility we could not measure the serum ammonia levels. The opening up of hepatopulmonary arterio-venous (AV) communication is the route for systemic embolism. In our case we could diagnose it by detecting bubbles in LA and LV (4^{th} cardiac cycle) by echocardiography after the injection of agitated saline into peripheral vein. It is important to prevent introduction of air while giving drugs or fluids intravenously as there is a high risk of systemic embolism hence both anaesthesiologist and cardiologist took maximum care throughout the procedure.

Appearance of pandigital cyanosis and clubbing are due to shunting of deoxygenated blood through opened pulmonary AV communication supplying to periphery and may be indicative of chronic hypoxemia. Congenital abnormalities like, cardiac defects, biliary atresia and polysplenia are associated with this malformation.[6] Our patient was a diagnosed and operated case of ostium secundum ASD patch closure when she was 3 year old. Post operatively no adverse event noted and was doing well till the age of 6years. We could find early changes of encephalopathy, cardiac anomaly, HPS, draining of left branch of PV to the IVC in this patient which meets criteria to diagnose Abernathy malformation.

We did choose general anaesthesia as a preferred technique as it helps to keep patient immobile, for better haemodynamic control and finally it was comfortable for cardiologist to do such a rare procedure. It is wise to reduce the dose of benzodiazepines as premedicant due to prolonged duration of action.[10] Ketamine was used as induction agent as to avoid fall in systemic vascular resistance. We did avoid nitrous oxide while maintaining anaesthesia to prevent rise in pulmonary vascular resistance and to avoid enlargement of air emboli if any that would have occurred during the procedure. Even though the isoflurane is volatile agent of choice in historical aspect as it is compared to the enflurane or halothane, the use of any one modern volatile agent is not an absolute contraindication. Most of the opioids are metabolized in liver and the half-life of the drugs is prolonged. Single dose fentanyl in impaired liver function doesn't alter the outcome.[10] In our patient we used 1mcg/kg Inj Fentanyl.

During the procedure arterial blood gas (ABG) analysis showed PaO2 of 106 mmHg and PaCO2 of 40 mmHg with FiO2 of 70%. Total duration of the procedure was 60 min, with minimal blood loss. Post operatively the ABG showed PaO2 of 90 with FiO2 of 50%. On 4^{th} post-op day she was maintaining room air saturation of 88-90% and was comfortable.

The improvement in the saturation was only transient with supplementation of oxygen. The cause of opening of pulmonary AV communication is mainly due to unmetabolized products entering in the circulation as products are acting as vasodilators. Once the shunt at portal level is occluded, toxic products gets metabolized and pulmonary AV communication closes gradually also cyanosis and clubbing disappears. The time taken to close these communications is variable. It took 6 weeks in our patient to improve in work tolerance, disappearance of clubbing and cyanosis, increase in room air saturation from 88 to 95%. Hence one should never expect the persistent rise in PaO2 and room air saturation immediately after the closure of porto-systemic shunt.

4. Conclusion

Multidisciplinary approach with high index of suspicion helps in diagnosing rare condition. One should not expect immediate improvement in room air saturation immediately after the shunt closure instead it improves only after closer of pulmonary AV communications after toxic products gets metabolized.

5. Source of Funding

None.

6. Conflict of Interest

None.

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