Perioperative Challenges in an Infant with Uncorrected “Pink-Tetralogy of Fallot”—for Excision of Choroid Plexus Papilloma: A Case Report

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Abstract

Perioperative management of an infant with intracranial tumor and congenital cyanotic heart disease poses a lot of challenges to the neuroanesthesiologist and neurointensivist. Maintaining both the cardiovascular and cerebrovascular homeostasis are of paramount importance for a successful outcome. In this case report, we have mentioned the perioperative challenges of a 7-month-old baby with tetralogy of Fallot who underwent craniotomy and excision of choroid plexus papilloma.

Keywords
► infant
► choroid plexus papilloma
► craniotomy
► tetralogy of Fallot

Introduction

Perioperative management of an infant with intracranial lesion and underlying cyanotic congenital heart disease (CHD) poses unique challenges to the neuroanesthesiologist and neurointensivist because of their complex hemodynamics. While managing these cases, both cardiovascular and cerebrovascular homeostasis have to be maintained for a successful outcome. An infant with CHD coming for noncardiac surgery is a common event.1‑3 Children with cyanotic heart diseases such as tetralogy of Fallot (TOF) coming for stereotactic aspiration or craniotomy for abscess drainage have also been reported commonly.4,5 However, an infant with TOF coming for craniotomy and excision of an intraventricular tumor is an uncommon event. Here we report the anesthetic challenges of an infant with TOF coming for craniotomy and excision of a choroid plexus papilloma.

Case Report

A 7-month-old male child (weight 7 kg, height 68 cm) was admitted with the diagnosis of choroid plexus papilloma for craniotomy and excision of the tumor. At 3 months of age, he was diagnosed with TOF with a bidirectional shunt. There was a history of sweating and difficulty in continuous sucking during breast-feeding. However, there was no history of cyanosis, cyanotic spells, or recurrent upper or lower respiratory tract infection. All developmental milestones were achieved according to age till date.

At 4 months of age, the patient had multiple episodes of tonic posturing associated with vomiting and irritable cry. He was evaluated and found to have a choroid plexus papilloma with hydrocephalus for which right ventriculoperitoneal shunt was done and the child improved symptomatically.

On examination, the child was alert and active, and his head circumference was 42 cm, anterior fontanelle was lax and sunken, and shunt site was healthy. He had posterior plagiocephaly (positional). There was no focal neurological deficit. Airway examination appeared normal, and there were no other congenital anomalies or dysmorphic facial features. His heart rate was 112 beats/min, and the respiratory rate was 40 breaths/min with the saturation of 98 to 99%. Cardiovascular examination revealed pansystolic murmur in the pulmonary area radiating all over the precordium. Respiratory and abdominal examinations were within normal limits.

His blood investigations were within normal limits with the hemoglobin of 11 g/dL. Magnetic resonance imaging (MRI) of the brain with contrast showed a well-defined large lobulated frond-like mass filling the atrium of right lateral ventricle measuring approximately 4.0 × 3.3 × 1.4 cm with hydrocephalus (►Fig. 1). The mass was hypointense on T1W sequences and hyperintense on T2W sequences with marked
enhancement on contrast administration. Electrocardiogram
(ECG) showed sinus rhythm, right ventricular hypertrophy,
and right axis deviation. Echocardiography revealed TOF with
the components of subaortic ventricular septal defect (VSD, 9
mm) with a bidirectional shunt, severe infundibular valvular
stenosis with doming of pulmonary valve seen, and less than
50% overriding of the aorta with an ejection fraction of 55%.

A multidisciplinary team involving the neurosurgeon,
cardiologist, and neuroanesthesiologist was consulted
regarding the management, and it was decided to proceed
with craniotomy and excision because of the large tumor
size and its enhancement. The TOF correction was planned
after the age of 1 year. An informed consent was taken from
the parents after explaining the risks of perioperative car-
diac morbidity and requirement of massive transfusion and
its associated complications and the need for postoperative
ventilation.

It is our routine practice to achieve intravenous access, a
night before surgery, to prevent dehydration in these cases.
Because the child had difficult intravenous access, it was de-
cided not to start an intravenous line in the ward to avoid
sympathetic stimulation. Hydration was continued by oral
means till 2 hours prior to surgery. Syrup levitiracetam 60
mg and phenytoin 15 mg twice daily were continued preop-
eratively. Because the child had a functioning ventriculoperi-
toneal (VP) shunt, we decided to sedate the child with syrup
trichloryl (80 mg/kg) for 45 minutes prior to arrival to oper-
ating room. The operating room was ready with necessary
drugs, equipment, and warm fluids.

The child was asleep on arrival to the operating room and
was induced with sevoflurane (maximum up to 4% increase)
and oxygen after connecting the pulse oximetry. Once the
child was deep, all standard monitors were connected and
a 24G intravenous line was started. Five milligrams of ket
amine, 15 µg of fentanyl, and 5 mg of propofol were given,
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amine, 15 µg of fentanyl, and 5 mg of propofol were given,
and the concentration of sevoflurane was reduced to 2%.
The child was paralyzed with 4 mg of atracurium, intubat-
ed with 4.5-size uncuffed endotracheal tube (ETT), and was
fixed at 12 cm at the lips. Anesthesia was maintained with
air, oxygen, and sevoflurane (minimum alveolar concentra-
tion [MAC] 0.8–1) and an atracurium infusion (5–10 µg/kg/
min) that was titrated to achieve two twitches on train of
four. The arterial CO₂ (PaCO₂) was maintained between 33
and 35 mm Hg. A 24-g arterial line was inserted into the left
radial artery, and right subclavian 5.5F triple lumen central
venous catheter was placed. Nasopharyngeal temperature,
urine output, and sugar were monitored intraoperatively.

The conditions included hypothermia, hypotension,
and respiratory insufficiency. Blood products were given
intravenously, and the patient’s condition improved. The
child started breathing comfortably with the respiratory rate
of 30 breath/min with good tidal volume and was awake.

Discussion

Tetralogy of Fallot is one of the common cyanotic CHDs,
which accounts for approximately 10% of cyanotic heart
diseases, and 50% infants with severe pulmonary ste-
nosis die of hypoxia and congestive heart failure during

**Fig. 1** (A–C) T1W postcontrast images of MRI of brain showing a large well-defined mass in the right atrium with hydrocephalus. MRI, magnetic resonance imaging.
the first year of life. Children with less severe pulmonary obstruction surviving this period later present with hypoxia, cyanosis, polycythemia, coagulopathies, congestive heart failure, and cyanotic spells. Children with mild pulmonary obstruction even survive to adulthood without any symptoms; they often referred to as “pink tetralogy of Fallot.” TOF has four major components: right ventricular hypertrophy, overriding aorta, membranous ventricular septal defect, and right ventricular outflow tract obstruction. In our case, the child was considered as “pink tet” in which the degree of right ventricular outflow obstruction is minimal, resulting in minimal or no significant right-to-left shunt. Therefore, the child did not have hypoxia, cyanosis, or polycythemia during the preoperative period. Intraoperatively, measures were taken to avoid “tet spell” by maintaining the SVR using noradrenaline and by avoiding factors that can increase the pulmonary vascular resistance (PVR) (hypoxia, hypercarbia, and acidosis). Also, unlike adults (in whom only extreme hypoxia can lead to cerebral vasodilation), in infants even a minor degree of hypoxia can lead to an increase in cerebral blood flow and thereby the intracranial pressure (ICP) that can reduce the cerebral perfusion.

Maintaining the cerebral hemodynamics, while keeping the anesthetic goals of TOF in mind, is a tough challenge for an anesthesiologist, especially while managing a bleeding vascular tumor in a small infant. In our case, because the child had VP shunt and a difficult intravenous access, he was induced using sevoflurane and anesthesia was deepened with ketamine and propofol before intubation.

To provide brain relaxation for adequate surgical exposure and avoid undue pressure on the underlying brain by the surgical retractor, mannitol was given. Mannitol can cause osmotic diuresis, dehydration, and can lead to hypovolemia that can precipitate “tet spell.” Fluids were titrated to keep the pulse pressure variation (PPV) less than 13, and the urine output was replaced adequately with Ringer’s lactate. Blood loss was promptly replaced with red blood cells to avoid hypovolemia. A low dose of noradrenaline infusion was started to avoid acute hypovolemia in times of surgical bleeding and to maintain the SVR. We took measures to prevent intraoperative hypoxia, hypercarbia, acidosis, and hypothermia because these can precipitate a “tet spell.” Scalp block was given before the surgical incision and before extubation to avoid the noxious stimulant-induced catecholamine release and to provide intra- as well as postoperative analgesia.

Conclusion

Thorough understanding of patient’s complex cardiovascular physiology and cerebral pathology, meticulous planning, and administration of titrated anesthetics to maintain both cardiovascular and cerebrovascular homeostasis are important for the successful outcome. A multidisciplinary approach involving the neurosurgeon, neuroanesthesiologist, cardiologist, and the neurointensivist is needed for a successful outcome.

Funding

None.

Conflict of Interest

None declared.

Note

This case report has not been submitted elsewhere for publication or presented at a meeting.

Author’s Contributions

All authors made material contributions to the handling of this case and to the intellectual content of this article.

References