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Case Report

Bilateral Congenital Anophthalmia: An Uncommon Clinical Entity with Anesthetic Considerations

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ABSTRACT

Rare, unusual, and common congenital abnormalities are among those scheduled for eye surgery. Numerous rare ophthalmopathies have significant anesthetic consequences and are linked to clinical disorders. Knowing which syndrome we are dealing with is important, but so is knowing which systems—from the cardiovascular, endocrine, metabolic, neuromuscular, genitourinary, and cerebrovascular systems to the airway—are involved, how much of them are involved, and whether there are any potential anesthetic complications. In uncommon clinical situations, it becomes even more crucial to comprehend these elements since they aid in case planning, predicting, and managing difficulties. We report a rare case of bilateral congenital anophthalmia intended for excision of right ocular edema because congenital anophthalmia is one of the rare disorders with an incidence of less than 3/1000, and microphthalmia has been documented in up to 11% of blind children.

Keywords
Ophthalmopathies; Congenital anophthalmia; Microophthalmia; Ocular surgery.

INTRODUCTION

The lack of ocular tissue inside the orbits is known as anophthalmia. Microphthalmia is a phenotypic continuum characterized by primitive or hypoplastic eyes. On imaging, clinical anophthalmia frequently turns out to be severe microphthalmia. It is one of the uncommon congenital defects that can manifest as a single finding or as a component of the syndrome, with an estimated incidence of 3 per 10,000 live births.1, 2 Anophthalmos has been attributed to both environmental and genetic factors. Low birth weight, advanced mother age, and diseases such as parvovirus, influenza virus, cytomegalovirus, varicella virus, rubella, and toxoplasmosis have all been linked.3.

Anophthalmos may manifest as a unilateral or bilateral lesion and may be linked to further systemic abnormalities, especially those that affect the central nervous system (CNS), musculoskeletal system, and heart. Atrial and ventricular septal defects (ASD and VSD), tetralogy of fallot, hypoplastic left ventricle, pulmonary valve atresia, and bicuspid aortic valve are examples of cardiac correlations.4. Septo-optic dysplasia, corpus callosum dysgenesis, anterior pituitary absence, corpus callosum agenesis, lack of septum pellucidum, dilatation of the ventricles, and polymicrogyria are among the many CNS problems that are frequently linked to bilateral anophthalmia. The growth of the orbital cavity, intraorbital soft tissue, maxilla, maxillary sinus, and mandible is also impacted by the absence of the normal globe, resulting in facial asymmetry, particularly in bilateral cases where sunken orbit and midfacial hypoplasia are common.5-7Ocular imaging, such as computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound, is performed to confirm a clinical diagnosis. Despite a clinical diagnosis, imaging or surgical exploration may reveal that anophthalmos is actually microphthalmos.

CASE REPORT

Over the course of four months, a 33-year-old man who had a history of absent eyes at birth developed a subtle, painless, soft globular bulge in his right eye. Since his mother was deceased, there was no information available about his maternal history, including maternal age and any drug, illness, or environmental toxin exposures during pregnancy. Upon local inspection, the mass

was soft, roughly 4.5 by 4.5 cm, and had a cystic consistency. No clinically noteworthy abnormalities were found during the general physical examination. The BMI was 24.5 kg/m2, the pulse rate was constant at 90 beats per minute, and the blood pressure was 124/84 mm-hg. When the heart was auscultated, there were no murmurs and the sounds were normal. When the heart was auscultated, there were no murmurs and the sounds were normal.

Upon airway assessment, the patient had a beard, Mallampatti grade 3, and the neck's typical range of flexion and extension. Every standard biochemical and hematological test came back within normal bounds. The ECG and chest radiograph showed nothing unusual. According to an external echocardiogram report, the patient had mild tricuspid regurgitation, a right ventricular pressure of 18/7 mmHg, a perimembranous VSD of 2.7 mm, and an ejection fraction of 40–45%.The right orbit's intrazonal and extraconal compartments had a massive, well-defined, lobulated cystic lesion measuring 6.1x4.2x4.1 cm, according to the MRI.

This lesion caused the bony orbit to expand and the medial and lateral rectus muscles to shift. On the top of the right maxillary sinus, the lesion was observed to induce an indentation and downward displacement. Medial to the aforementioned lesion, an oval soft tissue enhancement was observed, indicating calcification presumably of the rudimentary ocular globe with the hypoplastic right optic nerve. Similar little lesions were observed in the left orbit's anteroinferior extraconal region. Every intracranial structure was in normal condition.The patient was brought into the surgical room after written informed consent was obtained. On the right hand, a 20 G intravenous cannula was fastened. Every common anesthetic monitor was connected. Preoxygenation was performed for three minutes using a rendell-baker-soucek (RBS) mask to make sure that no external pressure was placed on the ocular edema because it was expected that the beard would make bag-mask ventilation difficult (Figure 1 and 2). Fentanyl (2 μg/kg), propofol (2 mg/kg), and vecuronium (0.1 mg/kg) were used for induction.

After making sure there was sufficient bilateral chest expansion and square wave end-tidal carbon dioxide, the airway was sealed with a pro-seal laryngeal mask airway no 4. After securing the device, the patient was switched to the pressure control ventilation mode. Nitrous oxide (N2O), isoflurane, and oxygen were used to maintain anesthesia. As part of multimodal analgesia, diclofenac and paracetamol were administered intravenously. When the medial rectus muscle was dissected, the patient experienced a bradycardia episode with an HR of 45 beats per minute. The surgeon was notified and the traction on the medial rectus was then released. After 15 seconds, the surgeons were instructed to continue after administering two drops of the local anesthetic proparacaine into the operative field. The procaine drops were once more injected into the surgical region upon the lateral rectus muscle becoming exposed. The patient stayed stable and experienced no hemodynamic problems after that. Ondansetron 4 mg was given at the end of anesthesia. Glycopyrrolate and neostigmine were used to turn the patient around.

Figure 1. *Pressure on Swelling with Face Mask*

Figure 2. *Preoxygenation with RBS Mask without Pressure on Swelling*

DISCUSSION

Any type of elective or urgent surgical operation can be scheduled for an adult or juvenile anophthalmic patient. A thorough birth history, records of any noteworthy perinatal incidents, and a family history of a related illness can all provide crucial hints about the aetiology. In children, it is necessary to rule out infections caused by Toxoplasmosis, Other Agents, Rubella or German Measles, CMV (Cytomegalovirus), and TORCH (Herpes Simplex) and any associated conditions. Anophthalmia has been linked to a number of syndromes and abnormalities, including heterotaxy, Fraser syndrome, Matthew-Wood syndrome, CHARGE syndrome, oculo-facial-cardio-dental-syndome, and anophthalmia-oesophageal-genital syndrome. A thorough clinical examination should be carried out, paying particular attention to the architecture of the face, heart, and central nervous system. Patients with facial asymmetry may be expected to have a challenging airway. To rule out any heart, central nervous system, or abdominal disease, patients may need pre-operative echocardiography, brain magnetic resonance imaging, or abdominal ultrasonography, depending on the clinical findings. Taking into account the related malformations and organ involvement, this may also assist in adjusting the anesthetic plan and selecting the proper airway management equipment, induction, and maintenance drugs.Our patient

was scheduled to have the eye tumor removed. An intracranial expansion of the tumor and related brain abnormalities were ruled out by pre-operative MRI. A pre-operative ECHO revealed minimal tricuspid regurgitation with an ejection fraction (EF) of 40–45% and a VSD of 2.7 mm. We expected that the beard would make bag-mask ventilation challenging and that wearing a facemask with a tight seal might put additional strain on the ocular mass, perhaps resulting in bleeding or pressure consequences (increased intraocular pressure, which can cause bradycardia). An RBS mask (Figure 2) was utilized for pre-oxygenation to prevent any external pressure on the ocular edema.As in our situation, VSD might manifest as a single flaw or as a part of a cluster of abnormalities. Since the majority of individuals with significant problems present early in childhood, only minor or moderately sized anomalies are considered asymptomatic in maturity. Under anesthesia, maintaining a balance between pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) is crucial for lowering the shunt fraction and, consequently, the likelihood of shunt reversal and subsequent problems. when a left-to-right shunt, VSD tends to grow when SVR rises and PVR falls, which causes pulmonary hypertension, increased pulmonary blood flow, and shunt reversal. Therefore, maintaining appropriate hypotensive anesthesia, as we did in this instance, should lower SVR in order to prevent these problems. This was also the rationale behind choosing LMA ProSealTM over the endotracheal tube in order to prevent the rise in systemic pressures linked to intubation and laryngoscopy.We expected the oculocardiac reflex to occur during surgery because the MRI indicated that the medial and lateral rectus muscles were involved. This was effectively controlled by careful observation and prompt pro-paracaine instillation.8. For fear of atropine-induced tachycardia exacerbating tricuspid regurgitation, neither preventive nor intraoperative atropine was used. Despite our patient's clinical diagnostic of anophthalmia, intraoperative diagnosis of microphthalmia was made because imaging and surgical exploration revealed the presence of a primitive eyeball.

The patient was successfully reversed at the conclusion of the treatment.The dangers of dysrhythmias, thromboembolic events, and occasionally deterioration of the shunt are the primary concerns in the post-operative period for such cases. As a result, patients must be nursed under close supervision, with a focus on reducing discomfort and preserving hemodynamic stability. The patient spent two days in the high dependency unit (HDU) during the post-operative phase in order to accomplish these objectives. The patient was discharged on the third post-operative day in a stable state compared to their pre-operative condition and underwent routine follow-up as planned.

CONCLUSION

Multisystem abnormalities that may or may not be clinically apparent may be linked to congenital ophthalmopathy. A thorough patient pre-operative evaluation aids in predicting, comprehending, and managing any intraoperative issues that might develop after surgery. Therefore, even in cases that appear straightforward, it is crucial to understand the clinical correlations of congenital abnormalities and their implications for anesthesia. The kind and method of anesthesia must be customized by anesthesiologists. In all such cases with pre-existing cardiac abnormalities, the primary concerns of hemodynamic stability and normocarbia should be addressed in conjunction with careful post-operative monitoring.

CONSENT

The patient has provided written permission for publication of the case details.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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