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Laporan Kasus

Komplikasi Perdarahan Intrakranial pada Anak dengan Tetralogy of Fallot: Sebuah Laporan Kasus

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Abstrak

Pendahuluan: *Tetralogy of Fallot* (ToF) adalah salah satu penyakit jantung bawaan (PJB) sianotik yang memiliki prevalensi sekitar 7%-10% dari seluruh kejadian penyakit jantung bawaan, proporsi antara laki-laki dan perempuan yang sama dan terjadi pada 3-5 per 10.000 kelahiran. Komplikasi ini pada ToF jarang terjadi, namun dapat menjadi berbahaya bila tidak segera mendapat penanganan. Laporan ini diharapkan dapat memberi rekomendasi penanganan pada kasus ToF yang memburuk dan menyebabkan komplikasi perdarahan intrakranial

Ilustrasi Kasus: Ditemukan kasus pada anak usia 6 tahun dengan keluhan penurunan kesadaran 4 jam sebelum masuk rumah sakit. Anak juga mengalami keluhan demam, batuk, pilek, muntah menyemprot, dan kejang. Pada pemeriksaan fisik didapatkan kesadaran yang normal, demam dan dispneu. Pada pemeriksaan penunjang didapatkan tanda perdarahan subdural, perdarahan *subarachnoid*, dan peningkatan tekanan intrakranial. Pasien sebelumnya sudah didiagnosis ToF dan rutin kontrol. Dari hasil pemeriksaan tersebut, pasien diterapi untuk menurunkan tekanan intrakranial serta manajemen infeksi. Setelah pasien kontrol, keluhan sudah berkurang dan klinis pasien berangsur membaik

Diskusi: Penyakit jantung bawaan dapat menyebabkan pembentukan trombus di dalam jantung, kemudian menyebabkan emboli yang beredar dalam sirkulasi perifer. Eritrosis sekunder dapat meningkatkan viskositas darah, hipoksemia kronis juga dapat mengaktifkan neutrofil dan sel mononuklear yang menyebabkan cedera endotel. Trombosit dan endotel berinteraksi dan meningkatkan pembentukan trombus intravaskular.

Simpulan: ToF dapat menyebabkan kematian utamanya akibat hipoksia, gangguan serebrovaskular, dan abses otak. Gangguan serebrovaskular termasuk perdarahan intrakranial pada kasus ToF dapat terjadi karena multi-faktor

Kata Kunci: *intrakranial, perdarahan, tetralogy of fallot*

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Intracranial Hemorrhagic Complications in a Child with Tetralogy of Fallot: A Case Report

Abstract

***Background**: Tetralogy of Fallot (ToF) is one of the cyanotic congenital heart diseases, accounting for roughly 7%–10% of all congenital heart-disease cases, with an equal male-to-female ratio and an incidence of 3 – 5 per 10 000 live births. Intracranial hemorrhagic complications in ToF are rare but can be life-threatening if not managed promptly. This report aims to offer practical recommendations for handling deteriorating ToF cases that lead to intracranial bleeding.* ***Case Illustration**: A six-year-old child presented with a four-hour history of reduced consciousness prior to hospital admission. Additional complaints included fever, cough, rhinitis, projectile vomiting, and seizures. Physical examination showed normal consciousness at admission, fever, and dyspnea. Imaging and ancillary tests revealed subdural and subarachnoid hemorrhages accompanied by elevated intracranial pressure. The patient had a known diagnosis of ToF and attended regular follow-ups. Management focused on lowering intracranial pressure and treating the infection. On subsequent follow-up visits, the symptoms subsided and the patient's clinical condition gradually improved.* ***Discussion**: Congenital heart disease can promote intracardiac thrombus formation, giving rise to emboli in the peripheral circulation. Secondary erythrocytosis increases blood viscosity, while chronic hypoxemia activates neutrophils and mononuclear cells, leading to endothelial injury. Platelet–endothelial interactions further enhance intravascular thrombogenesis, collectively predisposing ToF patients to cerebrovascular events.* ***Conclusion**: ToF can cause mortality primarily through hypoxia, cerebrovascular complications, and brain abscesses. Cerebrovascular complications, including intracranial hemorrhage, arise through multifactorial mechanisms.*

Keywords: hemorrhage, intracranial, Tetralogy of Fallot

1. BACKGROUND

Tetralogy of Fallot (ToF) is a cyanotic congenital heart disease characterized by four cardiac anomalies, which are ventricular septal defect, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy and accounts for 7-10 % of all congenital heart disease cases.[1], [2] Intracerebral complications can include brain abscess, stroke, and intracranial hemorrhage.[3]

Intracranial hemorrhage is divided into four broad categories: epidural, subdural, subarachnoid, and intraparenchymal bleeding. The incidence of subdural hemorrhage is estimated at 5 %–25 % in patients with significant head trauma, while subarachnoid hemorrhage represents about 5 % of all strokes.[4], [5]

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This report presents a demonstrative case to emphasize the need for early recognition and timely management of life-threatening complications arising from the deterioration of ToF.

2. CASE ILLUSTRATION

A 6-year-old girl presented to the Emergency Department with a four-hour history of decreased consciousness following a two-day febrile upper respiratory illness. Notably, the episode was preceded by projectile vomiting and transient loss of consciousness, raising concern for acute intracranial pathology.

On arrival, she developed a generalized tonic seizure lasting <5 minutes at a temperature of 38 °C, followed by full post-ictal recovery. A second seizure occurred on hospital day 2 despite a lower temperature (37.7 °C), suggesting a non-simple febrile seizure etiology. Persistent headache, dizziness, and meningeal signs further supported central nervous system involvement.

The patient had a known history of Tetralogy of Fallot (ToF) diagnosed at 9 months of age, representing a significant predisposing factor for cerebrovascular complications.

On examination, she appeared acutely ill with cyanosis and hypoxemia (SpO_2 87% on oxygen). Neurological findings were notable for nuchal rigidity, positive Brudzinski signs, and clonus, indicating meningeal irritation and possible increased intracranial pressure. Digital clubbing was also present, consistent with chronic hypoxia.

Laboratory evaluation demonstrated polycythemia (elevated hemoglobin and hematocrit), leukocytosis, and mild electrolyte imbalance. Coagulation studies showed prolonged prothrombin time and shortened activated partial thromboplastin time, suggesting a potential hemostatic disturbance contributing to hemorrhagic risk.

Cranial CT (Figure 1 and Figure 2) revealed extensive intracranial hemorrhage, including a right convexity subdural hemorrhage with significant mass effect and midline shift, as well as diffuse subarachnoid hemorrhage involving multiple intracranial compartments. Imaging findings confirmed elevated intracranial pressure and explained the patient's acute neurological deterioration.

Echocardiography (Figure 3) confirmed complex ToF with pulmonary atresia, atrial septal defect, patent ductus arteriosus, and a single coronary artery ostium, reinforcing the underlying cyanotic heart disease as a key risk factor.

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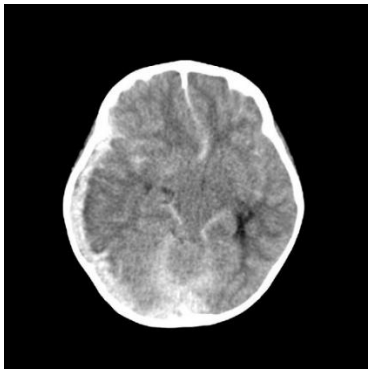

Figure 1. Non-contrast axial head CT scan showing subdural and subarachnoid hemorrhages with signs of elevated intracranial pressure (ICP).

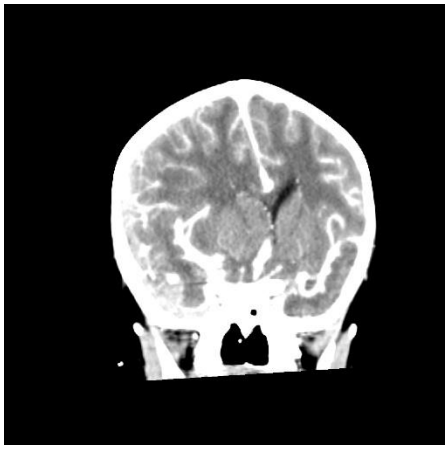

Figure 2. Contrast-enhanced coronal head CT scan showing subdural and subarachnoid hemorrhages with signs of elevated intracranial pressure (ICP).

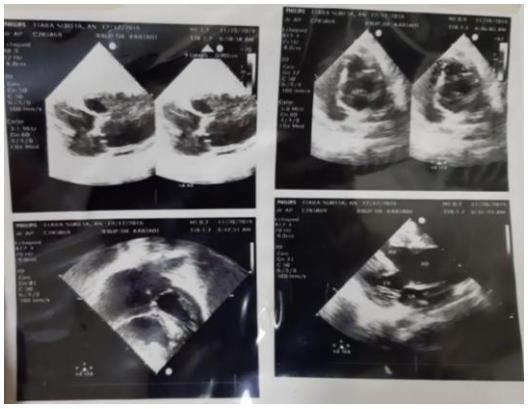

Figure 3. Echocardiographic image demonstrating findings consistent with ToF.

The patient was managed conservatively with osmotherapy (mannitol), seizure control, broad-spectrum antibiotics, corticosteroids, and supportive therapy including oxygen and propranolol. Early clinical improvement was observed within 24 hours, with resolution of fever, improved oxygenation, and no recurrent seizures.

By hospital day 3, neurological status had normalized with resolution of meningeal signs. Follow-up CT on day 5 showed partial hemorrhage resolution and reduced midline shift, indicating decreased intracranial pressure.

The patient was discharged on day 7 in stable condition, with no residual neurological deficits. At two-week follow-up, she remained asymptomatic without recurrence of seizures or cyanotic spells.

3. DISCUSSION

Tetralogy of Fallot (ToF) is a cyanotic congenital heart disease characterized by four structural defects (ventricular septal defect, pulmonary stenosis, overriding aorta, and right-ventricular hypertrophy).[1] ToF is the most common single congenital cardiac anomaly, accounting for roughly 5 % of all congenital heart-disease cases encountered in clinical practice. Reported causes of death in untreated ToF include severe hypoxia (62 %), cerebrovascular disorders (17 %), and brain abscesses (13 %).[5]

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In the present case, the patient had unrepaired ToF with pulmonary atresia and chronic hypoxemia, as evidenced by persistent cyanosis and low oxygen saturation despite oxygen therapy. These conditions likely contributed to the development of cerebrovascular complications, consistent with previous reports indicating a significant proportion of mortality in ToF is related to cerebrovascular disorders.[5]

Although cerebrovascular accidents (CVAs) are uncommon in young patients, they occur more frequently in individuals from rural areas lacking adequate medical facilities. Congenital heart disease predisposes to intracardiac thrombus formation, which may fragment and embolize into the systemic circulation.[6] In this patient, although no intracardiac thrombus was detected on echocardiography, the underlying cyanotic heart disease still represents a major risk factor for cerebrovascular events.

Alioğlu et al. documented intra-cardiac thrombosis in three of nine children with ToF.[7] Several factors heighten thrombotic risk in cyanotic heart disease: chronic acidosis promotes fibrin deposition; secondary erythrocytosis and hypoxia-induced activation of pro-coagulant pathways raise tissue-factor expression and impair fibrinolysis. Elevated blood viscosity from erythrocytosis diminishes cerebral perfusion, favoring in-situ clot formation. Chronic hypoxemia also activates neutrophils and mononuclear cells to release vasoactive and chemotactic mediators that damage the endothelium. Platelet–endothelial interactions then amplify intravascular thrombogenesis and trigger the coagulation cascade. Disruption of fibrinolysis, exemplified by raised plasminogen-activator inhibitor-1, further increases thrombotic propensity, supporting future consideration of anticoagulation in ToF patients who have not yet undergone surgical repair.[8]

In our patient, laboratory findings of elevated hemoglobin, hematocrit, and erythrocyte count are consistent with secondary erythrocytosis and hyperviscosity, which may have impaired cerebral perfusion and contributed to vascular instability. Interestingly, despite the predominance of thrombotic mechanisms described in the literature, this patient developed hemorrhagic complications, suggesting a complex interplay between thrombosis and bleeding tendencies.

Kaplan et al. reported ToF patients presenting with intracranial bleeding and even cerebral abscess; brain abscesses occur in about 13 % of ToF cases, facilitated by pulmonary hypertension and right-to-left shunting.[9] Although the precise pathophysiology remains unclear, intense local inflammation around septic emboli is thought to damage cerebral vessels and the blood–brain barrier, while elevated hematocrit

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and hypoxia adversely affect cardiovascular function and increase bleeding risk.[10] This mechanism may explain the findings in our patient, who presented with extensive subdural and subarachnoid hemorrhages. Chronic hypoxemia and elevated hematocrit likely contributed to vascular fragility and increased susceptibility to intracranial bleeding.

Fiorda-Díaz et al. described a patient with Alagille syndrome (ALGS) and unrepaired ToF who suffered a ruptured intracranial aneurysm with concomitant subarachnoid hemorrhage. ALGS—a genetic disorder involving multisystem dysfunction of the liver, cardiovascular system, and central nervous system—is frequently complicated by cerebrovascular anomalies such as aneurysms and arterial stenoses, and approximately 12 % of ALGS cases coexist with ToF.[11] Intracranial hemorrhage is a leading cause of death in ALGS; therefore, when ALGS co-occurs with ToF, vigilant neurovascular monitoring is essential. Although our patient had no confirmed genetic syndrome such as ALGS, this report highlights that patient with ToF alone may still develop severe intracranial hemorrhage, emphasizing the need for early neurological evaluation even in the absence of known syndromic associations.

Ai et al. documented a 40-year-old patient with ToF who developed intracerebral hemorrhage (ICH).[5] Mild thrombocytopenia—secondary to ineffective thrombopoiesis and reduced platelet survival—contributed to bleeding risk, as did hepatic congestion and clotting-factor abnormalities, reflected by a > 20 % prolongation of prothrombin time (PT) and activated partial thromboplastin time (APTT). Prolonged severe hypoxia further impairs cardiovascular integrity, predisposing cerebral vessels to rupture.[12] Similarly, in our patient, coagulation abnormalities were observed, including prolonged prothrombin time, which may have contributed to the hemorrhagic presentation.

Polycythemia in cyanotic congenital heart disease results from chronic hypoxemia caused by right-to-left shunting. The hypoxic kidney releases erythropoietin, stimulating bone-marrow erythropoiesis to augment oxygen-carrying capacity. Initially beneficial, rising hematocrit eventually produces marked hyperviscosity, reducing tissue perfusion and total oxygen delivery, thereby increasing the risk of veno-occlusive events and the hyperviscosity syndrome. Symptoms typically emerge once hematocrit exceeds 65 % and include headache, arthralgia, chest pain, irritability, anorexia, dyspnea, and exercise intolerance.[13] In the present case, the patient exhibited symptoms such as headache and dizziness, along with laboratory evidence of elevated hematocrit, supporting the presence of hyperviscosity. These factors likely contributed to both neurological symptoms and

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increased risk of intracranial complications.

This report has several inherent limitations. First, it describes the clinical course of a single patient; therefore, the observations cannot be generalized to all children with unrepaired Tetralogy of Fallot who develop intracranial hemorrhage. Second, genetic testing, a full thrombophilia work-up, and advanced neuroimaging (e.g., MRI or catheter angiography) could not be performed because of logistical and financial constraints, leaving potential pre-existing cerebrovascular anomalies or pro-coagulant disorders uncharacterized. Third, serial hematologic and inflammatory markers were obtained only during the acute hospitalization; longer-term laboratory surveillance might have clarified the evolution of coagulation abnormalities and inflammatory status. Fourth, follow-up neuroimaging was limited to a single repeat CT scan five days after presentation; the absence of serial imaging beyond discharge restricts our ability to document the trajectory of hemorrhage resolution and possible late complications such as hydrocephalus. Finally, the short outpatient control (2 weeks) precludes conclusions about long-term neurological, cardiac, and neurodevelopmental outcomes. Future multicenter series or prospective studies are needed to better delineate risk factors, optimal monitoring strategies, and evidence-based management protocols for intracranial hemorrhagic events in children with cyanotic congenital heart disease.

4. CONCLUSION

ToF is a high-risk cyanotic congenital heart disease in which survival depends not only on surgical correction but also on vigilant monitoring for extra-cardiac complications. This case highlights intracranial hemorrhage as a life-threatening manifestation resulting from the interplay of chronic hypoxemia, erythrocytosis, coagulation abnormalities, and endothelial dysfunction. Early recognition of neurological warning signs and prompt multidisciplinary management were essential for a favorable outcome.

We recommend a multidisciplinary follow-up approach, including periodic evaluation of hematologic and coagulation parameters, with neuroimaging when indicated. Preventive strategies such as hematocrit optimization, selective anticoagulation, and caregiver education may help reduce morbidity. Future studies should include prospective designs with larger sample sizes and long-term follow-up to better define risk factors, outcomes, and optimal preventive strategies for cerebrovascular complications in cyanotic congenital heart disease.

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