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TERSON SYNDROME IN A PATIENT WITH UNREPAIRED TETRALOGY OF FALLOT

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ABSTRACT
Tetralogy of Fallot is the most commonly seen cyanotic congenital heart disease. It includes four cardinal features: right ventricular outflow tract obstruction, misaligned ventricular septal defect, overriding aorta, and concentric right ventricular hypertrophy. Intracranial pathologies such as brain abscesses, cerebrovascular accidents, and intracranial haemorrhage all have been reported with Tetralogy of Fallot. Here we present a case who had subarachnoid haemorrhage with vitreous haemorrhage in Tetralogy of Fallot.

KEY WORDS: Tetralogy of Fallot, Ventricular septal defect, Terson syndrome

INTRODUCTION
The most frequent congenital cardiac abnormality seen in clinical practice is tetralogy of Fallot (TOF). It includes the following major features: right ventricular outflow tract obstruction, misaligned ventricular septal defect, overriding aorta, and concentric right ventricular hypertrophy. TOF remains the most common cyanotic congenital heart disease to reach adulthood without surgical repair. Currently in TOF patients, life expectancy has increased, making it more challenging to manage chronic multisystem disorders. Intracranial pathologies such as brain abscesses, cerebrovascular accidents, and intracranial haemorrhage all have been previously documented in TOF patients.[1]

We describe the case of a 20-year-old male, who had significant subarachnoid hemorrhage (SAH), sub conjunctival haemorrhage, vitreous haemorrhage and an unrepaired TOF. Terson syndrome refers to vitreous haemorrhage associated with subarachnoid haemorrhage, occasionally accompanied by retinal haemorrhages. Intraocular haemorrhages occur because of raised intra cranial pressure, causing obstruction of central retinal vein resulting in venous stasis that ultimately leads to vitreous and intra-retinal haemorrhages. It is an independent poor prognostic indicator in patients with subarachnoid haemorrhage.

CASE PRESENTATION
A 20-year-old male patient, previously diagnosed case of tetralogy of Fallot at 14 years of age without receiving treatment, presented in Emergency Room (ER) of Pakistan Institute of Medical Sciences with complains of headache that was sudden onset, very severe in intensity, with blurring of vision, neck pain and vomiting for three days. There was no history of fever, seizures or altered sensorium. At presentation in ER, his GCS was 15/15, blood pressure 150/100 mm Hg, pulse 88 beats/min, and SpO2 76 percent at room air. General physical examination revealed abnormal bluish skin color i.e. cyanosis on his lips, tips of fingers and toes and tip of nose due to his cyanotic congenital heart disease i.e. Tetralogy of Fallot. Grade 4 clubbing was in both finger and toe nails. He had pink lips, protruding eyes with obvious bilateral sub conjunctival hemorrhages. He had MRC grade 5 motor power in both upper and lower limbs, neck was supple in all four planes, plantars were bilateral down-going. On chest auscultation, there was bilateral equal air entry with harsh vesicular breathing. Cardiac auscultation revealed normal first and second heart sounds. Table 1 below shows the laboratory investigations of patient including hematology, serum chemistries and immunology investigations.

Table 1: Baseline laboratory investigations of patient

Blood complete picture:	**Reference Range:**
Hb 21.5	13.0 - 17.0 g/dL
HCT 70	40 - 55%
MCV 84.9	80 to 100 femtoliter
MCH 26.2	27 to 31 picograms/cell
TLC 5360	5,000-10,000/mL
Platelets 33000	150,000 - 400,000/mL
Liver function tests:	
Bilirubin total 0.392	0.1 to 1.2 mg/dL
ALT 12.7	4 to 36 U/L
ALP 108	44 to 147 (IU/L)
Renal function tests:	
Urea 55.5	5 to 20 mg/dl
Serum creatinine 2.45	0.74 to 1.35 mg/dL
CPK 42.5	10 to 120 (mcg/L)
Serum electrolytes:	
Serum sodium 114.6	135 to 145 (mEq/L)
Serum potassium 5.71	3.7 to 5.2 (mEq/L)
Coagulation profile:	
PT 40	11 to 13.5 seconds
APTT 13	30-40 seconds
INR 1	1.1 or below is considered normal
HbsAg Negative	
Anti HCV Negative	

Echocardiography done several months ago had showed levocardia, situs solitus with normal venous connections, atrio ventricular concordance, ventricular arterial concordance, intact inter atrial symptoms, large peri-membranous VSD with overriding of aorta, shunting right to left alongside good sized left ventricle, severe right ventricular outflow obstruction , good size pulmonary vasculature, descending aorta 10.8 mm ,MPA 10 mm , RPA 10.2 mm , LPA 9.4 mm , no PDA , normal left aortic arch; all features conclusive of tetralogy of Fallot. Repeated echocardiography done on this presentation, showing ventricular septal defect measuring 23 mm with overriding of aorta, concentric left ventricular hypertrophy and right ventricular hypertrophy, with ejection fraction of 60 percent.

In neuroimaging, a CT scan brain (shown in figure - 1) showed hyperdense material filling the subarachnoid space and more apparent around circle of Willis.

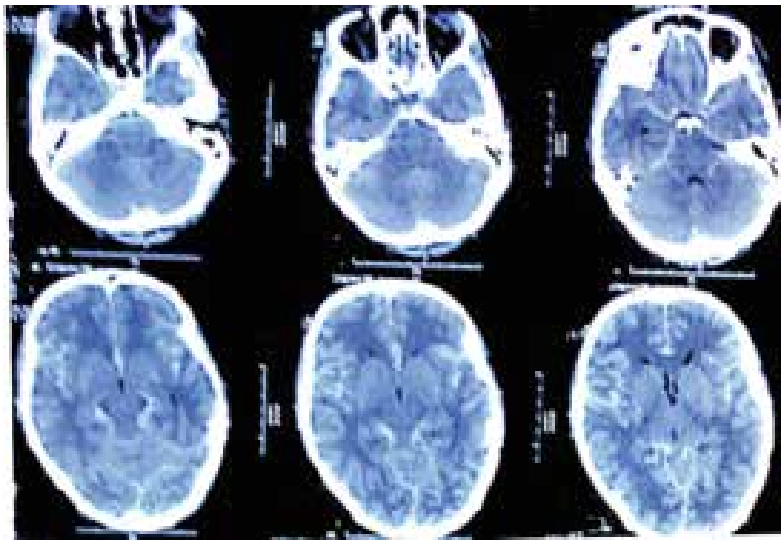

Figure 1: CT Brain plain showed hyperdense areas (sub arachnoid haemorrhage) more prominent around circle of Willis.

CT angiography of brain was done which did not show any aneurysm.

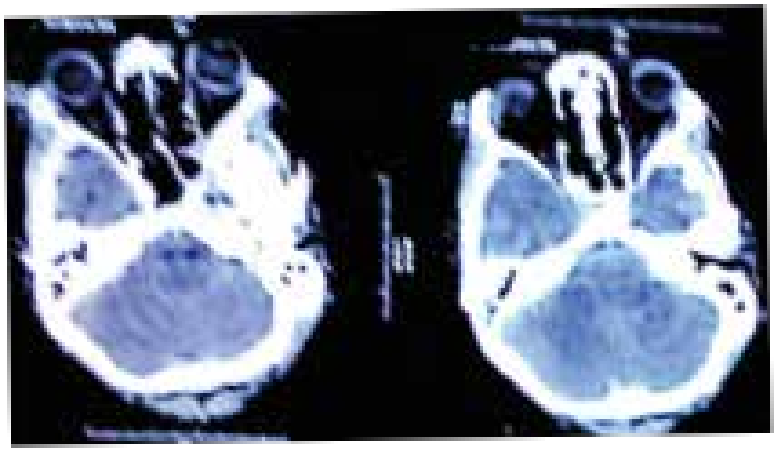

Figure 2: CT Brain plain showed hyperdense lesion in vitreous body

Detailed ophthalmologic examination of patient was in plan but he became very sick quickly and it was deferred. Based on his CT brain plain and CT angiography findings, a diagnosis of non-aneurysmal subarachnoid haemorrhage with vitreous haemorrhage, more obvious in left orbital region was made. His Hunt and Hess grade was 2. Patient was given oxygen support, IV fluids, IV mannitol 150 ml 6 hourly, tab nimodipine 60 mg started every 4 hourly, pain relief coverage added, empirical antibiotics added for prophylaxis due to immunocompromised state of patient. Platelets were transfused due to thrombocytopenia accompanied by overt bleeding. Critical care and neurosurgical consult were taken regarding his management, and neurosurgery advised that any surgical procedure would be very high risk due to untreated cyanotic heart disease and if needed an external ventricular drain (EVD) could be placed. After discussion with patients' guardian it was decided to take the conservative management route. Unfortunately, the patient did not respond to any treatment and his neurological status deteriorated and he rapidly became comatose and died after 24 hours.

DISCUSSION

Tetralogy of Fallot accounts for approximately around 6-7percent of congenital heart diseases and is one of the most common congenital heart diseases.[1,2] The

prevalence of TOF is approximately 4-5 /10000 live births. Unrepaired TOF is associated with poor survival, with one half of individuals dying in first decade of life and only few living till third decade of life. Management of intracerebral haemorrhage and cerebrovascular accidents are more challenging in patients with congenital heart diseases.[1] Previously, there were very few cases reported regarding association of Terson syndrome with subarachnoid haemorrhages. Terson syndrome results in higher Hunt and Hess grade and is associated with significantly higher risk of mortality. It has been reported in 13-50% of patients with subarachnoid haemorrhage. Terson syndrome has also been reported in patients with spontaneous and traumatic SAH, including several cases of cerebral venous sinus thrombosis.[3]

In our case, we report Terson syndrome in patient with unrepaired TOF. Multiple factors were involved in critical and deteriorating condition of our patient which included severe hypoxia, decreased platelets, subconjunctival and vitreous haemorrhages and unrepaired TOF; all these factors exacerbating his compromised cardiopulmonary condition and thus higher risk of mortality, although his mental and motor condition improved to some extent initially with supportive care. Headache, deteriorating GCS and blurring of vision are major predictors of Terson syndrome in SAH patients and were present in our patient.[4] Chronic hypoxemia is predisposing factor for thickening of vessel wall and early atherosclerosis and that in turn causes a higher risk of haemorrhage.[1] Adults with unrepaired congenital heart diseases are at greater risk of intracerebral haemorrhage, making it more challenging to manage these patients. Early corrections of these defects should result in better outcomes.

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Zeeshan Munawar; case management, data analysis, manuscript revision
Ifrah Ayaz; case management, data analysis, manuscript revision
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All the authors have approved the final version of the article, and agree to be accountable for all aspects of the work.