Case Report

Tetralogy of fallot with right internal jugular vein thrombosis

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Congenital heart diseases are divided into cyanotic and acyanotic.There are several conditions that start acyanotic and become cyanotic with time, eg, Fallot's tetralogy, Ebstein's anomaly and left-to-right shunts developing into the Eisenmenger syndrome. Paediatric stroke is an uncommon entity. The most common cause of stroke in children is probably congenital heart disease. Other risk factors include sickle cell disease, infections and various protrombotic conditions. We present a case report of 14 year old male child who was previously diagnosed with Tetralogy of fallot, presented with altered sensorium Investigations revealed hemorrhagic infarcts in the right parieto-occipital and right cerebellum and thrombosis of right internal jugular vein transverse and sigmoid sinus.

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Key words : **Congenital heart disease,Tetralogy of Fallot, Paediatric Stroke, Hemorrhagic infarcts,Thrombosis.**

The estimated incidence of congenital heart disease (CHD) is 1%, ie, 1 out of 100 children have some form of CHD, either major or minor. In cyanotic heart disease, Tetralogy of Fallot (TOF) remains the most common diagnosis[1]. The estimated incidence of ischemic stroke in children older than 28 days of life is variable, but according to a prospective population study, it averages 13/100,000 for all strokes,7.9/100,000 for ischemic stroke and 5.1/100,000 for hemorrhagic strokes[2]. Systemic arterial circulation can be the source of emboli or it can be paradoxical embolus. Long-standing cyanotic lesions cause polycythemia and anemia, which increase the risk of thromboembolism and cerebral infarction[3]. The diagnosis of cerebral venous thrombosis which has to be differentiated from polycythemia and can be confirmed with Magnetic Resonance Imaging (MRI)[4].

Case Report

A 14 year old boy presented with complaints of fever, headache, altered sensorium for 4 days. The boy was a diagnosed case of Tetralogy of Fallot with history of cyanotic spells and squatting episodes since childhood. There was no history of surgical intervention as per the information given by the mother. On examination patient is in altered sensorium, temperature-99F, tachycardia [110/min], tachypnea [24/min], saturation [54%], central cyanosis, grade 3 clubbing, measured blood pressure is 100/60 mm Hg. Cardiovascular system examination revealed a ejection systolic murmur in pulmonary area. Other system examinations were unremarkable. Complete blood picture revealed Hb – 16.8 gms/dl, WBC – 6700/cmm [Neutrophils – 66%, Lymphocytes – 30%, Eosinophils – 2%], Hematocrit– 55.2%, MCV – 72.1 cubic micron, MCH – 21.9 pg, MCHC – 30.4 gm/dl, plateletcount 1.1lakh / Cmm, Reticulocyte count <0.5%,no hemoparasites. Blood Urea – 26 mg/dl, Serum Creatinine – 1.1 mg/dL, Blood Glucose - 56 mg/dl, Sr. Electrolytes – sodium – 144 meq/l, potassium – 4.9 meq/l, PT Test – 19.6 sec, PT with INR – 1.55, APTT 40.2 sec.Homocysteine levels –13.68umol/L, Lipid profiler evealed total cholesterol – 108mg/dl, HDL – 26 mg/dl, LDL – 69 mg/dl, VLDL -13 mg/dl, Triglycerdies – 67 mg/dl. Blood - ASO titres > 200 I U/ml, CRP - > 6 mg/dl, Widal – Negative, HbSAg & HIV were negative. Arterial blood gas analysis revealed pH – 7.366, PCO_2 – 39 mm Hg, PO_2 – 27 mm Hg, BEecf – (-) 3 mmol, HCO_3 – 22.3 mmol, TCO_2 – 24 mmol/L, SO_2 – 49%, Na – 137 mmol/L, K – 2.1 mmol/L, Ca – 0.87 mmol/L, HCT – 58% PCV, Hb – 19.7gm/dL. Electrocardiogram revealed normal sinus rythm, P pulmonale, biventricular hypertrophy with ischemia with right axis deviation.

2D Echocardiography revealed Tetralogy of Fallot with severe valvular pulmonary stenosis with hypoplastic branch pulmonary artery. CT brain revealed hyperdense lesions in deep cerebellar veins, vein of galen, inferior sagittal and superior sagittal sinuses, dilated right lateral ventricles suggestive of hemorrhage. Magnetic Resonance Imaging of Brain revealed multiple altered signal intensities noted in right high parietal, right parieto–occipital, right cerebellum which is hyperintense on T2WI, FLAIR, isointense with hyperintense rim on T1WI, showing restriction on DWI – suggestive of hemorrhagic infarcts. Altered signal intensity which is iso to heterogenous on all sequences with dilatation of right lateral ventricle suggestive of intraventricular extension of the bleed. Magnetic resonance venogram of brain revealed thrombosis of right internal jugular vein, transverse and sigmoid sinus. Magnetic Resonance Angiography of brain was normal (Figs 1-4).

Discussion

Tetralogy of Fallot was described over 100 years ago by a French physician, Etienne-Louis Arthur Fallot. The original tetralogy described by Fallot in 1888 is: pulmonary stenosis, Ventricular septal defect (VSD), overriding of the aorta, Right ventricle (RV) hypertrophy[1]. The following are additional anomalies or problems commonly associated: Right-sided aortic arch (in 25%), Absent or hypoplastic left pulmonary artery (more common if arch is right sided), Aortic regurgitation caused by large aortic ring plus sub aortic VSD or ASD[5]. When atrial septal defect (ASD) is coexisting

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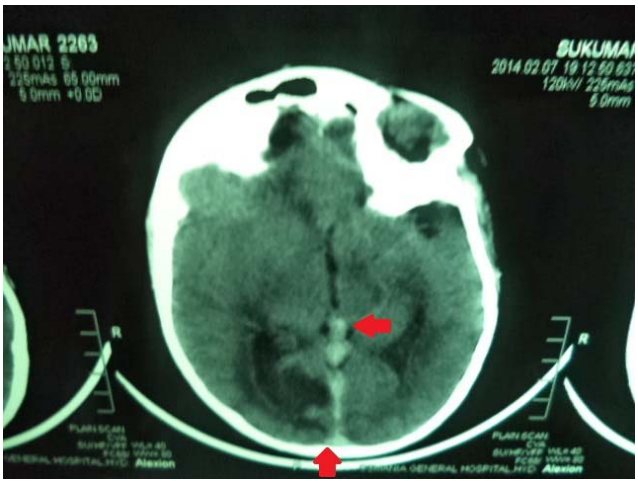

Fig 1 — Illustrating hyperdense deep cerebral veins, vein of Galen, inferior saggital and superior saggital sinuses,hypodensity noted in right occipital lobe involving white matter, dilated right lateral ventricle

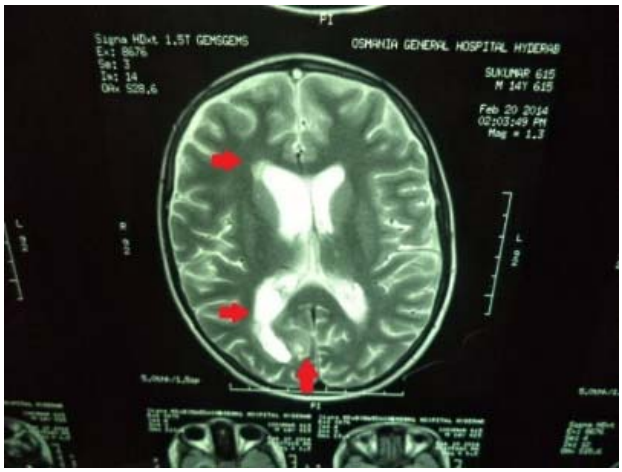

Fig 3 — Illustrating hyperintense areas in right high parietal, right occipital with iso to heterogenous signal intensity on all sequences with dilatation of right lateral ventricle suggestive of intraventricular extension of bleed

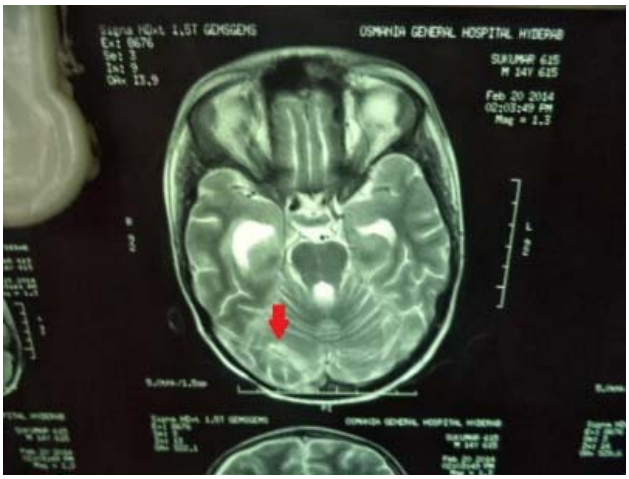

Fig 2 — Illustrating hyperintense area on T2WI in right cerebellum suggestive of hemorrhagic infarcts

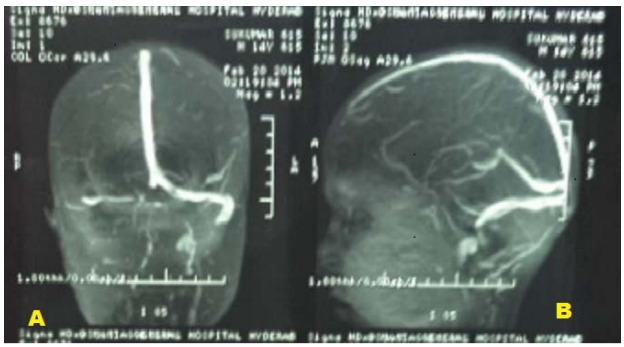

Fig 4 — MRV (A) Coronal, (B) Saggital viewsuggestive of right internal jugular vein, transverse, sigmoid sinus thrombosis

it is called pentalogy of Fallot[6]. The severity of RV outflow obstruction determines the clinical presentation. The severity of hypoplasia of the RV outflow tract varies from mild to complete (pulmonary atresia)[7]. Mild pulmonary stenosis may be associated with the 'acyanotic' child with Fallot's tetralogy[5]. When the RV outflow obstruction is severe, pulmonary blood flow is reduced markedly, and a large volume of desaturated systemic venous blood shunts right-to-left across the VSD. Severe cyanosis and erythrocytosis occur, and symptoms of systemic hypoxemia are prominent. In many infants and children, the obstruction is mild but progressive. Typical clinical presentation is patients are not cyanosed at birth (compare transposition of great arteries). It usually appears at 3–6 months and increases with time. Cyanotic attacks develop often with 'stress', crying or feeding. Increasing cyanosis results in syncope and convulsions. The pulmonary stenotic murmur may disappear during attacks. Cerebral blood flow may be so severely compromised that permanent neurological damage results. Poor growth, delayed milestones are common. Squatting is common in older children once walking starts. In more severe cases, children may squat at rest (knees up to chest and buttocks on the ground). Symptoms of polycythaemia are arterial or venous thromboses, particularly cerebral; children must not be allowed to get dehydrated, which can precipitate this. Complications include infective endocarditis,cerebral abscess (absence of lung filter with right-to-left shunt), paradoxical embolism, gout, acne, kyphoscoliosis, recurrent gingivitis[5]. Stroke can be either due to cerebral arterial thrombosis has been attributed to secondary to erythrocytosis of cyanotic heart disease or due to intracranial dural sinuses or cerebral vein thrombosis usually in association with iron deficiency anemia. Superior saggital sinus, transverse sinus,great vein of Galen and meningeal veins are the common sites of thrombosis in decreasing order of precedence[8].

Medical Management Although the definitive treatment of tetralogy of Fallot is surgical, medical management plays a role before surgery and in the postoperative period. Medical management includes treatment of cyanotic spells with high concentration oxygen, morphine, alpha blockers, prevention of hypoxic spells with propanalol, prevention of dehydration, adequate nutrition, prevention of iron deficiency anemia, phlebotomy for polycythemia, infective endocarditis prophylaxis, good dental hygiene, heparin

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and anticoagulation for stroke[6].

Surgical Management Historically, the approach to tetralogy of Fallot has been either palliation or corrective surgery. Over the years, the age at which corrective surgery can be performed has dropped, so that in most centers primary repair is the procedure of choice at any age. Palliative surgeries inlude modified Blalock-Taussig shunt, potts shunt, Waterston shunt, glenn operation[5]. Corrective surgery includes closure of VSD, preservation of right ventricular formand function, with unobstructed right ventricular outflow tract (RVOT) incorporating a competent pulmonary valve[9]. After total correction there may be further problems such as arrhythmias, RV failure, heart blocks, pulmonary regurgitation, RVOT aneurysm, aortic root dilatation. Repeat cardiac catheterization is sometimes necessary in patients after total correctionn to assess all these factors[5]. Our case is a congenital cyanotic heart disease, Tetralogy of Fallot with hypoplastic pulmonary artery with thrombosis of right internal jugular vein, transverse and sigmoid sinus and hemorrhagic infarcts in right high parietal, right parieto–occipital and right cerebellum.

References

1 Maheshwari S — Congenital Heart disease, API Textbook of Medicine,YP Munjal, 9th edition, Vol 1, 716, 720.

2 Giroud M, Lemesle M, Gouyon JB, Nivelon JL, Mlan C, Dumas R —Cerebrovascular disease in children under 16 years of age in the city of Dijon,France:A Study of incidence and clinical features from 1985 to 1993; *J Clin Epidemiol* 1995; **48:** 1343-8.[PubMed]

3 Carvalho KS, Garg BP — Arterial strokes in children. *Neurol Clin* 2002; **20:** 1079-100.[PubMed]

4 Healy JF, Nichols C — Polycythemia mimicking venous sinus thrombosis. *AJNR Am J Neuroradiol* 2002; **23:** 1402-3. [PubMed]

5 R.H.Swanton and S.Banerjee.Congenital Heart Disease. Chapter 2.Swanton Cardiology: A Concise Guide to Clinical Practice, 6th edition, 2008. 17,53-60.

6 David W Brown, David R Fulton — Congenital Heart Disease in Children and Adolescents , Hurst's The Heart, 13th edition, Fuster, Walsh, Harrington. 1827.

7 Child JS, Aboulhosn J — Congenital Heart Diseases in Adult ,Chapter 19, Harrison's Cardiovascular Medicine, 2e, 2013, 207.

8 Rose SS,Shah AA,Hoover DR,Saidi P — Cyanotic congenital Heart Disease(CCHD) with symptomatic erythrocytosis. *J Gen Intern Med* 2007; **22:** 1775-7.[PMC free article][PubMed]

9 Apitz C, Webb GD, Redington AN — Tetralogy of Fallot. *Lancet* 2009; **374:** 1462-71. doi: 10.1016/S0140-6736(09)60657-7. Epub 2009 Aug 14.1462-71. doi: 10.1016/S0140-6736(09)60657-7. Epub 2009 Aug 14.