A Rare Case of Tetralogy Of Fallot with Right Atrial Thrombus presenting with Hemiplegia complicating the clinical course

Authors-

1) Dr. Parag Sharma
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)
2) Dr. Satyajit Samal
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)
3) Dr. Debmalya Saha
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)
4) Dr. S.E.H. Naqvi
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)
5) Dr. Saket Aggarwal
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)
6) Dr. M.A. Geelani
   (Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)

First author- Dr. Parag Sharma

(Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)

Email id- dr.parag.sharma2817@gmail.com

Corresponding author- Dr. Satyajit Samal

(Department of Cardiothoracic Surgery, GB Pant Hospital, New Delhi, India)

Email id- drsatyajitctvs@gmail.com

Abstract

Cerebrovascular Accidents represent a dangerous complication of cyanotic children with tetralogy of fallot with incidence of 8.6%.Tetralogy of Fallot has been associated with raised hematocrit with low arterial Saturation.

Here we describe an 18 months old female child of Tetralogy of Fallot with Cyanotic spells acute onset right sided hemiplegia with Right Atrial thrombus who underwent emergency surgery with intraoperative predicament.

Keywords

Tetralogy of Fallot, Right atrial thrombus, Hemiplegia, Anticoagulation

Introduction

Tetralogy of Fallot (TOF) is the most common congenital heart disease with incidence 9-14 %. TOF remains the most common type of congenital heart lesion seen beyond infancy and childhood with about 5 % of patients surviving to the age of 40 years.[(1)]

Stroke and brain abscess are the most common neurological complications of uncorrected congenital heart disease. Long-standing cyanotic lesions cause polycythemia and anemia, which increase the risk of thromboembolism and cerebral infarction. [(2,3,)] Various causes of stroke have been recognized in children with cyanotic heart disease including thromboembolism, prolonged hypotension, and polycythemia. [(7)] Iron-deficiency anemia and dehydration are other factors that hasten the formation of thrombus in these patients.[(8)]

We here report a rare clinical entity of a patient with tetralogy of Fallot with Right Atrial thrombus with right sided hemiplegia. For reporting of this rare case, approval was taken from the institutional & zonal ethical committee and due informed consent was taken from the parents of the patient in their own language as per institutional proforma.

Case Report

An 18 month-old female presented with a history of cyanosis and recurrent cyanotic spells with acute onset right sided hemiplegia. 2D echocardiogram & TEE demonstrated a large ventricular septal defect (VSD) and severe infundibular and pulmonary valvular stenosis consistent with tetralogy of Fallot (TOF). In addition, there was a (2 × 1)cm right atrial mass originating near the IVC opening and suspected to be a thrombus. A computed tomography angiogram confirmed the presence of the right atrial Thrombus of size (2 ×1)cm. NCCT brain showed presence of left gangliocapsular infarct. A preoperative thrombotic workup revealed a hemoglobin of 24 g/dl, a hematocrit of 72%, a white blood cell count of 5000, a platelet count of 88,000, a bleeding time of 80 seconds, a clotting time of 2 minutes 3 seconds, and an international normalized ratio of 1.4. Keeping on low molecular heparin with and platelet transfusion, the patient underwent urgent repair of the TOF. Patient was operated under standard cardiopulmonary bypass with mild hypothermia with aortic and bicaval cannulation. A thrombus of size 2×1cm found in right atrium was carefully removed taking care of the atrial wall and a small pericardial patch was sutured at the site of thrombus. RVOT muscle bundle excision was done adequately. VSD closure was done with a dacron patch through transatrial approach. Trans annular pericardial patch augmentation of RVOT and main pulmonary artery done. Patient was weaned of CPB uneventfully and had a stable postoperative course in the ICU. Patient was discharged 7 days after the surgery and was advised Aspirin 50 mg tablet OD.

Patient came for follow up at regular intervals and a repeat 2D echo was done 2 and 6 months after surgery which showed no thrombus and normal biventricular function with no flow across the VSD patch.

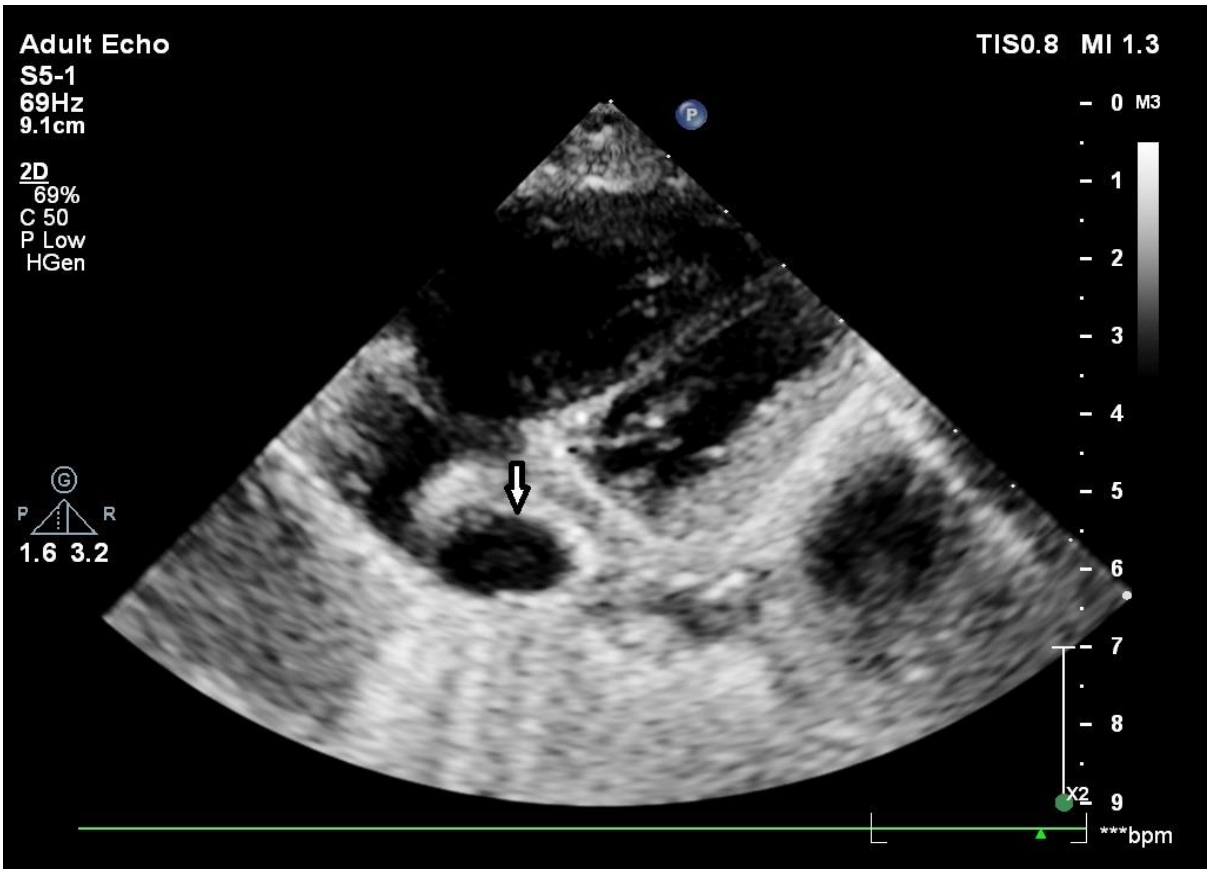

Figure 1. Echocardiography (PSAX view). The white arrow shows right atrial thrombus.

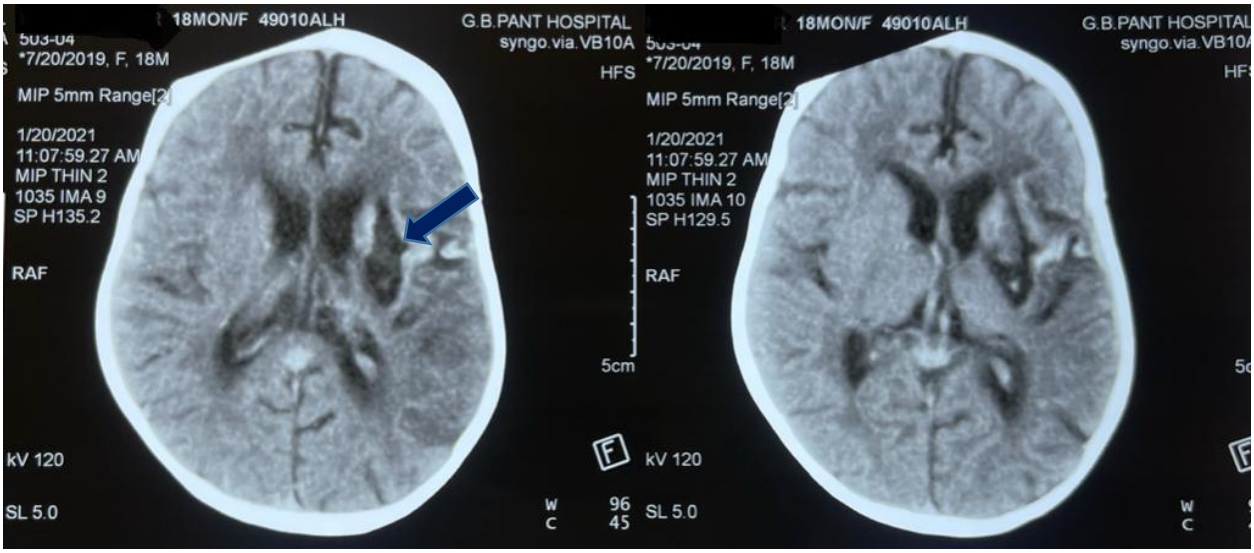

Figure 2. NCCT scan of brain. The blue arrow shows infarct in left gangliocapsular region.

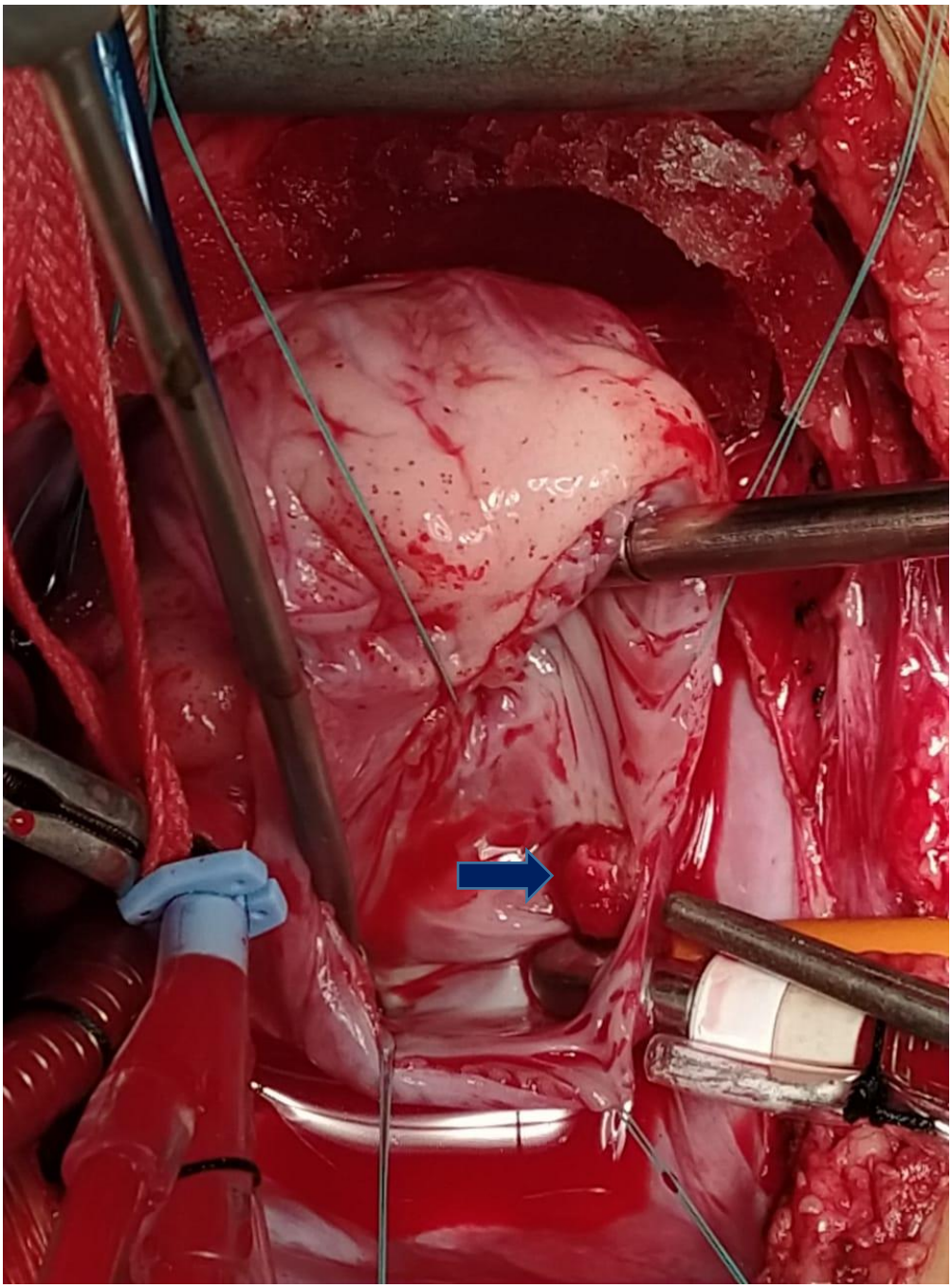

Figure 3. Intraoperative photograph with opened right atrium. The blue arrow shows thrombus in right atrium.

DISCUSSION

Stroke and brain abscess are the most common neurological complications of uncorrected congenital heart disease. Various causes of stroke have been recognized in children with cyanotic heart disease including thromboembolism, prolonged hypotension, and polycythemia. [7] An association between elevated hematocrit and thrombosis has been established in polycythemia vera. However, no association has been established between elevated hematocrit and secondary erythrocytosis in case of cyanotic heart disease.[8] Iron-deficiency anemia and dehydration are other factors that hasten the formation of thrombus in these patients. Congenital heart disease patients with iron deficiency anemia develop infarction due to thrombosis commonly in intracranial dural sinuses like superior sagittal sinus, transverse sinus, great vein of Galen, and meningeal veins. [9] Cerebral emboli originate either in the systemic arterial circulation or in the venous circulation, that is, lower extremity veins or pelvic veins. [10] Corrected as well as uncorrected cases of TOF are associated with increased risk of developing pulmonary thrombosis, venous thrombosis, cerebral embolism, and infective endocarditis. [11] A paradoxical embolism is a type of stroke or arterial thrombosis caused by a thrombus of venous origin through a defect in the heart that creates a potential for right-to-left shunting of blood as in patients with TOF.[11] In our case the child with TOF had infarct in left gangliocapsular region with a thrombus in right atrium without any other systemic cause for thromboembolism. We suspect that the hyperviscosity due to the elevated hematocrit and decreased platelet count was responsible for the right atrial thrombus formation and cardioembolic stroke. To best of our knowledge, this is the first case case to be reported in the literature. Various options for treatment in children with cerebral sinus thrombosis include standard or low molecular weight heparin for 7-10 days, followed by oral anticoagulants for 3-6 months. Though anticoagulation is controversial, use of heparin or low molecular weight heparin may be considered in children with cardiac embolism, arterial dissection, coagulation disorders, and recurrent stroke. Anticoagulant treatment may be indicated in a child with progressive deterioration, or during initial evaluation of a new cerebral infarction. Low-dose aspirin (3-5 mg/kg) is recommended if anticoagulation is not given. [12,13] Tetralogy of Fallot is well known as the cause of brain abscess but this case report draws attention to an interesting association of TOF with cardioembolic stroke with right atrial thrombus formation.

Conflict Of Interest – None

Ethical Approval – Approved by institutional & zonal ethical committee

Informed Consent – Taken from patient parents in their own language

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