Tetralogy of Fallot with Neurological Manifestations: A Case Report

R.K. Kotokey, Swarup Kar, Arup Sharma, Libe Nyorak, S. Rupshi and Deepanjan Ghosh
Department of Medicine, Assam Medical College, Dibrugarh.

ABSTRACT

Fallot’s Tetralogy is one of the most common cyanotic congenital heart diseases. Most of the patients though die during adulthood (depending upon the severity of pulmonary stenosis) but long term survival has also been recorded in the literature. Though there are various complications of Tetralogy of Fallot; multiple brain abscesses as a sequelae of Fallot’s Tetralogy leading to neurological manifestations is not frequently encountered in this part of North East. A case of Fallot’s Tetralogy presenting with severe neurological deficit due to multiple brain abscesses who was treated conservatively is presented. The patient responded to the conservative management. (J Med Sci 2009;12(1):18-20)

Key words: Tetralogy of Fallot’s (TOF), Brain abscess, Jugular venous pressure (JVP), Intercostal space (ICS), Right ventricle (RV), Ventricular septal defect (VSD), Cardiovascular system (CVS), Right ventricular outflow tract (RVOT).

Introduction

Tetralogy of Fallot (TOF) is a complex of anatomic abnormalities arising from the maldevelopment of right ventricular infundibulum. It was first described by Etienne-Louis Arthur Fallot in 1888. The four components of the TOF are malaligned ventricular septal defect, obstruction of RV outflow, aortic override of the ventricular septal defect, and RV hypertrophy due to the RV ‘seeing’ aortic pressure via the large VSD. TOF may also coexist with AV septal defect. RV outflow tract obstruction is variable. Often a stenotic, bicuspid pulmonary valve with supravalvular hypoplasia exists.

It is the commonest cyanotic congenital heart disease in children above the age of 2 years consisting almost 75% of all blue patients. Associated abnormalities in TOF includes a right aortic arch in about 25% of patients, and abnormalities of the course of coronary arteries in approximately 5%.

The commonest complications of TOF are congestive cardiac failure, infective endocarditis and neurological complications. Out of these complications brain abscess accounts for 5-18.7%. It should be suspected in any cyanotic patient presenting with headache, convulsion, vomiting, with or without fever and neurological deficit. We herewith present a case of TOF who prevented with neurological deficits.

Case Report

A 30 year male was admitted in Assam Medical College, Dibrugarh with the complaints of weakness of the left side of the body for 1- month duration with bowel and bladder incontinence accompanied by high fever, headache and multiple episodes of seizures. The attendant gives history of exertional shortness of breath since one year of life which was gradually increasing. He has history of cyanotic spells which were relieved by squatting. There was a history of bluish discoloration of skin and mucous membrane and swelling of the tip of fingers
which were gradually increasing since one year of his life. The patient did not take advice from any doctor since last 20 years.

On general examination patient was febrile; grade II clubbing was present; cyanosis present; clinical evidence of polycythemia present; JVP raised; oedema present; pulse: 90/min, regular, normal in volume and character, no radio radial or radio femoral delay; BP: 110/80 mm of Hg. On CNS examination patient had dysarthria (cortical in type). His intelligence was average. Motor system examination revealed upper motor neuron type grade 0 power in left upper limb and 3/5 in left lower limb with clasp-knife rigidity and hyper-reflexia. Other systems examination reveals no abnormality.

Cardiovascular system revealed RV type of cardiomegaly, single S1, single S2, and a rough and low pitched ejection systolic murmur (grade 3/6) along the left parasternal area best heard in 2nd and 3rd intercostal space. Other organ systems were normal.

The patient was diagnosed as Tetralogy of Fallot's with left sided hemiplegia due to brain abscesses with moderate congestive heart failure.

**Discussion**

The case presented here is a patient of TOF with multiple brain abscesses on the right side. Patient presented with hemiplegia of the left side with evidence of moderate degree of heart failure.

Natural history of TOF is variable and determined by the degree of RV outflow tract obstruction. Approximately 25% of untreated patients of TOF and RVOT obstruction die within the first year of life, 40% by 4 years, 70% by 10 years and 95% by 40 years. However, cases of
survival of patients into their 80’s have been reported. Incidence of TOF is slightly higher in males than in females.

Neurological complications occur frequently in TOF. Anoxic infarction in the CNS may occur during an anoxic spell and result in hemiplegia. Paradoxical embolism to CNS and venous thrombosis due to sluggish circulation from polycythemia can also result in hemiplegia. Brain abscess is not infrequent. In fact the incidence of brain abscess in patients with cyanotic heart disease has been reported to the range between 5-18.7%, and TOF is the most common cardiac anomaly associated with brain abscess.1

Cyanotic heart disease is the most commonly identified risk factor for development of brain abscess in immunocompetent patients. Most of the brain abscesses are supratentorial in location.1 In patients with cyanotic heart disease, there is a right to left shunt of venous blood in the heart bypassing the pulmonary circulation. Thus, bacteria in the bloodstream are not filtered through the pulmonary circulation where they would normally be removed by phagocytosis. Patients with cyanotic heart disease could have low-perfusion areas in the brain due to chronic severe hypoxemia and metabolic acidosis as well as increased viscosity of blood due to secondary polycythemia. These low-perfusion areas commonly occur in the junction of grey and white matter, and they are prone to seeding by microorganisms that may be present in the bloodstream. The haematogenous mode of spread accounts for the subcortical location as well as multiple numbers of abscesses often on countered in these patients.8

The treatment of choice in these patients is aspiration of the abscess through a burr hole or twist drill craniostomy. But in our patient, we managed him conservatively with broad spectrum antibiotics. He responded well as he recovered completely and could walk normally, took his food himself with a normal intellect and conversation, bladder and bowel habits also recovered completely.

In a developing country like India, where many a times surgical intervention in not possible; a meticulous conservative treatment and care may save the life of many complicated patients.

Acknowledgement

We are thankful to the Principal cum Chief Superintendent, Assam Medical College and Hospital, Dibrugarh for kindly allowing us to publish the hospital records.

References

1. John S Child. Congenital Heart Disease in the Adult; Harrison’s Principles of Internal Medicine, 17th Edition 2008; Chapter 229; p1463.
2. GD Webb, JF Smallhorn, J Therrien, Redington AN. Congenital Heart Disease; in: Braunwald’s Heart Disease; 8th Edition 2007; Chapter 61; p1587.
4. Garry D. Webb, Jeffrey F. Smallhorn, Judith Therrien, Andrew N. Redington. Congenital Heart Disease; Braunwald’s Heart Disease; 8th Edition 2007; Chapter 61; p1587(Int.).