Case Report

A Case Report of Pentalogy of Fallot with Headache: Brain Imaging Revealed Subarachnoid Hemorrhage

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ABSTRACT

Pentalogy of Fallot (PoF) is a rare form of congenital cyanotic heart disease. In the presence of the components of Tetralogy of Fallot (ToF), there is an additional defect in PoF; either atrial septal defect (ASD) or patent foramen ovale (PFO). Common Neurological complications of PoF include brain abscess and thrombosis. We present here a case of 18-year-old female with history of headache for 20 days. After her admission into the Department of Cardiology under Chattogram Medical College Hospital, Bangladesh, she was diagnosed as a case of POF; the fifth component was ASD. Her had history of illness suggestive of cyanotic spells since childhood. In the absence of any neurological deficit and cranial nerve involvement, fundoscopy revealed papilledema and sub hyaloid hemorrhage. Chest x-ray and echocardiography revealed typical features of ToF, with additional ASD. CT scan of brain showed subarachnoid hemorrhage. Conservative management was given after consultation with the Department of Neurology of the same hospital. Unlike ToF, which has been extensively described, PoF is less represented in the literature. Therefore, we present this case as an academic interest.

Keywords: *Pentalogy of Fallot, Tetralogy of Fallot, subarachnoid hemorrhage*

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INTRODUCTION

Pentalogy of Fallot (PoF) is a rare variant of Tetralogy of Fallot (ToF). ToF complex congenital cyanotic heart disease presenting 5% to 10% of congenital defects, affecting males and females equally and occurring in 3 to 5 of every 10,000 live births.[2-4] The tetralogy of Fallot (ToF) consists of a ventricular septal defect (VSD), over-riding aorta and right ventricular hypertrophy (RVH) due to obstruction of the right ventricular outflow tract (RVOT).[2] In Pentalogy of Fallot (PoF), there is an additional defect: either atrial septal defect (ASD) or patent foramen ovale (PFO).[5]

In Tetralogy of Fallot (ToF), only 1% of patients survive to the age of 40 years without undergoing surgery.[6] The common neurological fatal complications include cerebrovascular accidents (17%), and brain abscesses (13%).[1] Congenital heart disease (CHD) patients has more than 8 times higher risk of developing intracerebral hemorrhage (ICH) and almost an 8× higher risk of developing subarachnoid hemorrhage (SAH) compared with normal population.[7] To our knowledge, this is the first case reporting SAH in a PoF patient at adolescent age in our country. Unlike ToF, which has been extensively described, PoF is less represented in the literature. Therefore, we present this case as an academic interest.

CASE PRESENTATION

A 16-years old female came with complains of insidious onset of headache, accompanied with several episodes of vomiting. Her past history of illness was suggestive of cyanotic spells and recurrent respiratory tract infections since childhood. However, this was her first ever hospital admission. On physical examination, central cyanosis, suffused conjunctiva, digital clubbing involving both upper and lower extremities, SPO_2 80% in room air. Her Glasgow Coma Scale (GCS) was 15/15. With the intact cranial nerve function and absence of signs of meningism, her fundoscopy reveled sub hyaloid hemorrhage. Her laboratory tests revealed that her hemoglobin conc. was 22g/dl, hematocrit was 71%, platelet count was 50,000/mm^3, and prothrombin time was 34s. Her ECG showed typical presentation of Fallot's tetralogy, i.e., right axis deviation with right

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bundle branch block and right ventricular hypertrophy with large R waves in anterior precordial leads and large S wave in the lateral precordial leads. Her chest radiograph showed a normal-size heart silhouette, with an upturned apex and a concave main pulmonary artery segment, commonly known as "boot-shaped" (Fig. 1). Transthoracic echocardio-graphy revealed large non-restrictive VSD, severe infundibular stenosis, overriding of the aorta, pulmonary artery hypoplasia, right atrium (RA) and right ventricle (RV) dilated, ASD (10 mm), and bidirectional flow (Fig. 2). Non-contrast CT (computed tomography) scan of brain showed basal cisterns and sylvian fissures is due to acute bleeding into the subarachnoid space (Fig. 3). According to the Hunt and Hess Scale (HHS), she had Grade I - Asymptomatic or mild headache and slight nuchal rigidity, The Fisher scale (CT scan appearance) was as follows: Group 2 - Diffuse deposition of subarachnoid blood, no clots, and no layers of blood greater than 1 mm. Conservative management was given after consultation with the Department of Neurology and Critical Care team of the same hospital.

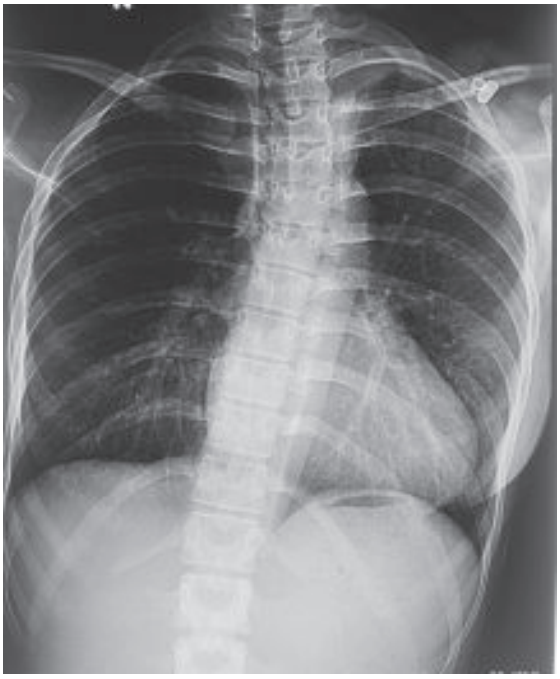

Fig. 1: *Chest x-ray P/A view showing a normal-size "boot-shaped" heart silhouette, with an upturned apex and a concave main pulmonary artery segment.*

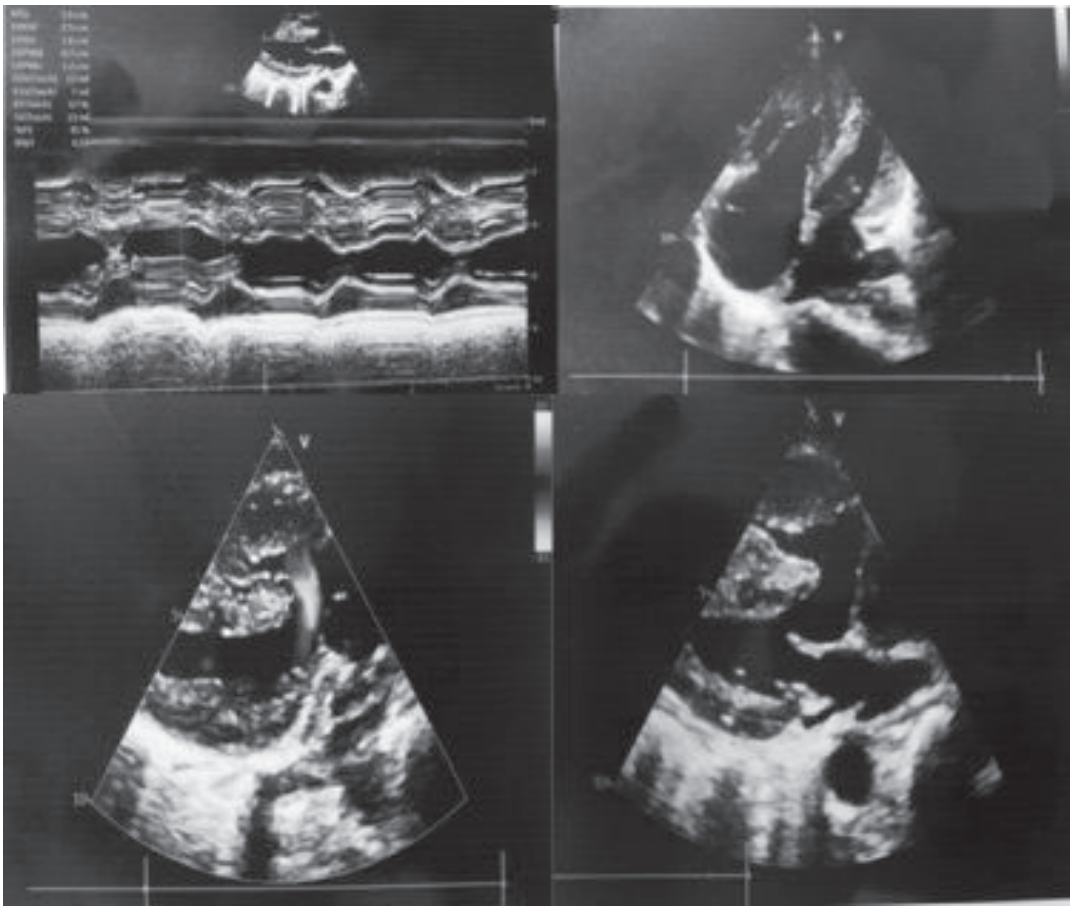

Fig. 2: *Transthoracic echocardiography revealed ToF with ASD.*

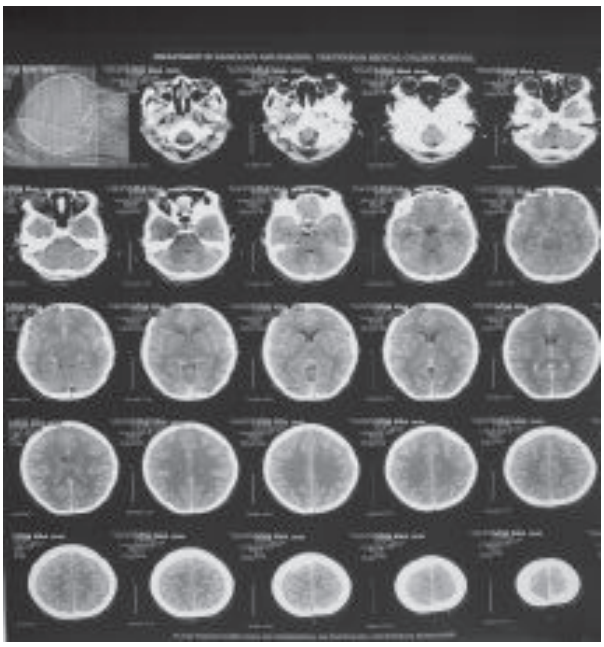

Fig. 3: *Non-contrast CT scan of brain showing subarachnoid hemorrhage.*

DISCUSSION

In association with Tetralogy of Fallot (ToF), the most common type of ASD found with PoF is ostium secundum (OS), followed by the primum ASD.[2,4,5] One of the most important factor that determines the amount of blood that is shunted thorough the VSD in PoF is the ratio of systemic vascular resistance (SVR) to pulmonary vascular resistance (PVR). Greater the SVR and lesser the PVR, lesser will be the shunt. An important factor determining pulmonary blood flow is the presence of infundibular or valvular pulmonary stenosis.[4] Patients with well-balanced shunt and the presence of collateral circuits may initially present at a later stage.[2] However, due to chronic hypoxia, patients with unrepaired ToF are at a high risk for both, thrombosis and hemorrhage. Secondaray polycythaemia in ToF leads to hyperviscosity, which is also a risk factor for both ischemic and hemorrhagic stroke. Mild thrombocytopenia caused by ineffective thrombopoiesis and diminished platelet survival.[4,8] Coagulation defect can be explained by deficient synthesis resulting from systemic hypoxia as well as from sluggishness of the local microcirculation caused by high blood viscosity.[8] Overall, patients with CHD may have an increased risk of ICH because of anatomic abnormalities, such as coarctation of the aorta (CoA) or persisting shunts, or because of multiple surgical and medical interventions including the use of anticoagulants, which have previously been shown to increase the risk of hemorrhagic stroke.[9]

Subarachnoid hemorrhage (SAH) in ToF is extremely rare, so there is paucity of evidence of patho-physiology, onset of symptoms, management plan of SAH in ToF.[9] Non-traumatic non-aneurysmal SAH (NA-SAH) can be devided to perimesencephalic (PM) and non-perimesencephalic (NPM) variety. NPM was defined if the pattern of bleeding was extending to the Sylvian and interhemispheric cistern, and not limited to the PM.cisterns.[10] Compared to aneurysmal SAH, NA-SAH excellent clinical outcome and prognosis, with low risk of complications that include rebleeding, vasospasm, hydrocephalus.[10] The approach to managing noncardiac complications in CHD requires a multidisciplinary team with expertise in subspecialties,[9] therefore, we consulted with the Department of Neurology and Critical Care team of the same hospital.

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