



Case Report

Successful Intravenous Thrombolytic Therapy in a Patient with Tetralogy of Fallot Presenting with Stroke

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Abstract

Congenital heart disease patients would have an elevated stroke risk compared to the general population. The prevalence of adults living with this condition is increasing, and knowledge of their risks and management should be of common knowledge to adult clinicians. The mechanisms of stroke implicate right to left shunt allowing paradoxical embolization. Anticoagulation of patients with complex congenital heart disease is preferred with vitamin K antagonist. We describe a successful thrombolysis case of a patient with Tetralogy of Fallot who presented at the emergency room with an ischemic stroke in time for intravascular thrombolysis treatment and had favorable outcomes.

Keywords: Congenital heart disease; Cyanotic congenital heart disease; Tetralogy of Fallot; Ischemic Stroke; Intravascular thrombolysis

Objectives

Stroke is a significant cause of morbidity and mortality. In Mexico, congenital heart disease has been reported to be approximately 0.8 to 1.4%; the prevalence is higher in premature patients, being as high as 43 per 1000 live births [1]. We report the case of a woman with a history of cyanotic heart disease who presented to the emergency room with motor aphasia and right pyramidal tract syndrome after one hour of evolution. The main objective of this case presentation is to be aware of the clinical

complications and management of people living with congenital heart disease.

Congenital heart disease, which arises from the abnormal or incomplete formation of the heart, valves, and blood vessels, is the most common congenital disability worldwide. Significant variability in its presentation results in heterogeneous morbidity and mortality in the life span [2]. Cyanotic congenital heart disease has reduced flow through the pulmonary circulation with a right-to-left shunting of blood, resulting in cyanosis and compensatory erythrocytosis. Tetralogy of Fallot and Eisenmenger syndrome are the most common cyanotic congenital heart disease associated with stroke. Cyanotic congenital heart disease predisposes to ischemic stroke by allowing paradoxical embolization and triggering

hyperviscosity. In normal circumstances, the pulmonary capillary bed acts as a filter, preventing venous embolic material from reaching the arterial circulation. Right to left shunts allow emboli to cross into the arterial circulation; this phenomenon is one of the most common mechanisms of stroke in patients with congenital heart disease. The hyperviscosity associated with cyanotic congenital heart disease is determined by polycythemia and compensatory erythrocytosis with hematocrit at times over 70% leading to reduced oxygen delivery and increased thrombotic events [3].

Methods

Case Presentation

A right-handed woman in her forties with a history of cyanotic congenital heart disease, with no previous medical, follow-up, nor use of any drugs, was admitted to the emergency room complaining of trouble speaking and weakness of her right side of her body, she presented after an hour evolution of sudden motor aphasia, supranuclear right facial palsy, superior and inferior right limb weakness and right extensor plantar reflex. A general physical examination revealed a holosystolic heart murmur at the left sternal border and digital clubbing. When she arrived, her evaluation of the National Institute of Health Stroke Scale was 9 points for a partial facial (lower) paralysis (+2), right arm (+2) and leg (+2) motor drift, mild-moderate loss of sensation; less sharp/duller (+1), mild-moderate aphasia (+1) and mild-moderate dysarthria (+1).

Finger stick blood glucose was 89 mg/dL, blood pressure 136/89 mmHg and oxygen saturation of 70%. An urgent brain computed tomography scan showed no early ischemic changes or bleeding (Figure 1). Therefore, the patient had no contraindications for intravascular thrombolysis treatment. After a signed authorization was obtained thrombolysis with Alteplase 0.9mg/Kg was administered, with a 10% (4.5mg) of the total dose intravenous (IV) bolus over 1 minute, followed by the remaining 90% (40.5mg) as a continuous infusion over 60 minutes. Patient was admitted to the Intensive Care Unit for close surveillance.

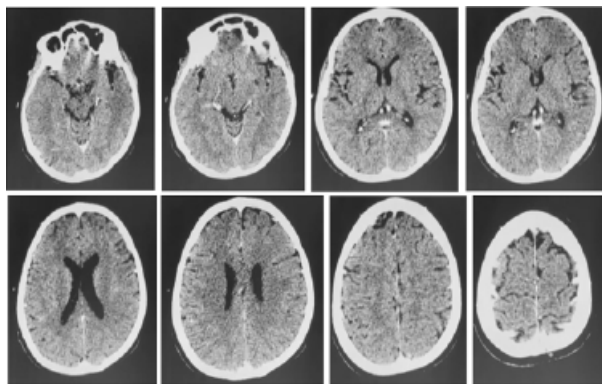


Figure 1: Computer tomography scan of the head was obtained within 25 minutes from the patient's arrival to the ER room, with no early changes seen at the ASPECTS regions.

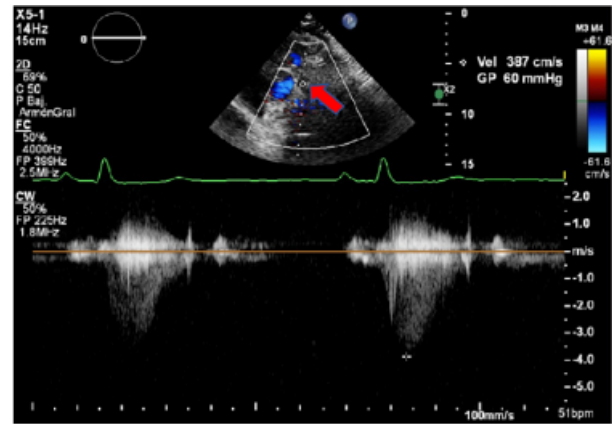


Figure 2: Continuous Doppler of pulmonary valve level showing severe sub-valvular stenosis with a peak gradient of 60 mmHg.

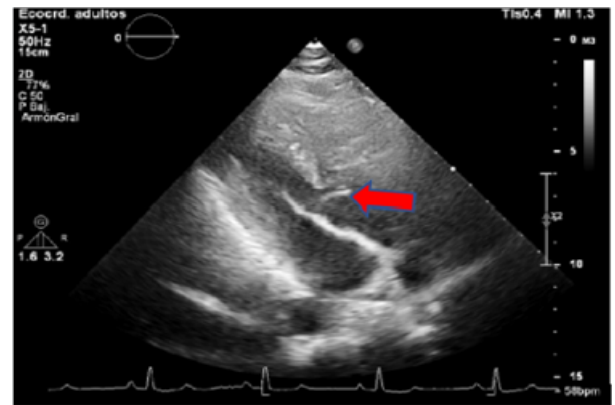


Figure 3: Aorta with a degree of override of 50% misplaced at the ventricular septal defect.

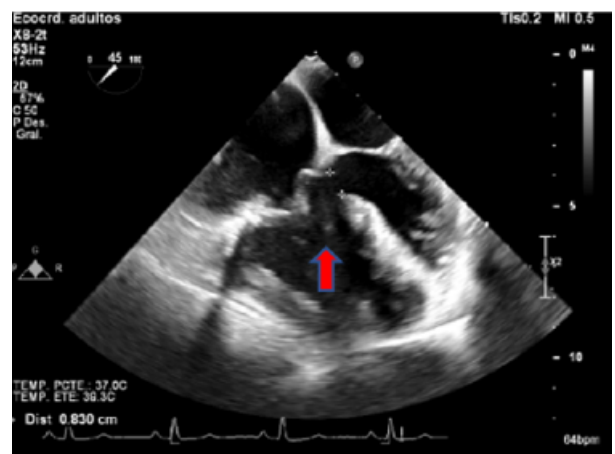


Figure 4: Ventricular septum defect of 15 mm.

Results

Thrombolysis was considered effective, finding a complete resolution of the neurological deficits, regaining total motor and language function. No intracranial hemorrhages or ischemic lesions were shown at the 24 hours control computed tomography scan. Remarkable biochemistry parameters found were hemoglobin of 24.2 g/dL, hematocrit of 72%, mean corpuscular volumen of 95 fl and mean corpuscular hemoglobin concentration 32 pg, no other abnormal results were found. Electrocardiogram at the follow up evaluation showed normal sinus rhythm and showed no dysrhythmia. A transesophageal echocardiogram was performed with a conclusion of Tetralogy of Fallot with bidirectional shunt and a normal left ventricular ejection fraction. Anticoagulation with a vitamin K antagonist was initiated achieving an International Normalized Ratio of 2. The patient was referred to the cardiology and rehabilitation departments and patient was discharged home. She remained well at follow-up 3 months later, being able to return to all of her previous activities, with a Modified Rankin Score of 0.

Discussion

Patients with congenital heart disease have a higher risk of stroke with an estimated prevalence in patients with Tetralogy of Fallot of 2.4% being this condition a major contributor to morbidity in this population despite absence of classical cardiovascular risk factors. It is believed that the incidence to be seen three times higher in cyanotic patients [4]. Prevention of Stroke in adults with congenital heart disease demands a multidisciplinary approach. Current scores to stratify risk of stroke in patients with atrial fibrillation may not apply to this population because this form of complex congenital heart disease coexists with other pathophysiology mechanisms besides dysrhythmia as a source of stroke even in the absence of atrial fibrillation [5].

Treatment of paradoxical emboli should consider both clot prevention and shunt correction [6]. It is well described in the literature that vitamin K antagonists would be preferred in cyanotic heart disease as there is insufficient safety and efficacy data to support routine non-vitamin K antagonist oral anticoagulant [7,8]. Treatment with intravenous recombinant tissue plasminogen (tPA) should also be performed in suitable patients who present later (6-12 hours). It is now known that everyone's penumbra is different because of different collaterals, cerebral blood flow reserve and tolerance to ischemia; the advent of new Magnetic Resonance Imaging technology has allowed performing rapid multimodal imaging studies to evaluate acute ischaemic stroke beyond the acute phase of presentation. Magnetic Resonance Imaging diffusion-weighted imaging/fluid attenuation inversion response (DWI/FLAIR) mismatch guides recombinant tissue plasminogen therapy for these patients [9]. The possibility of symptomatic cerebral haemorrhage as the primary adverse event should always be monitored. However, there appears to be no difference in this complication when the time of recombinant tissue plasminogen administration is "off-level" [10]. Multisystemic genetic syndromes are outside the scope of this case.

Conclusions

Congenital Heart Disease is the most common congenital disability, affecting 8 of 1000 live births. Tetralogy of Fallot is a congenital cyanotic heart disease characterized by pulmonary stenosis, ventricular septal defect, right ventricular hypertrophy, and aorta overriding the ventricular septal defect that predisposes to an increased risk of ischemic stroke due to increased red blood cell mass (hyperviscosity), activation of pro-coagulant pathways, impaired fibrinolysis and by allowing paradoxical embolization. Thrombolysis must be considered in all patients without contraindications within the first 4.5 hours of an ischemic stroke, even more time in well-selected cases. Scores to stratify the risk of stroke may not be enforced for this specific population because other mechanisms besides dysrhythmia are implicated. Vitamin K antagonists are preferred for anticoagulation therapy without sufficient data for non-vitamin K antagonists. Complex Congenital Heart Disease patients are at risk for stroke due to conventional vascular risk and their interaction with underlying cardiac disease. These patients should be evaluated early with a multidisciplinary team including Cardiologist and Neurologist.

Acknowledgments

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