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Case Report

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Post Pump Chorea: An Untold Mystery

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ABSTRACT

Post pump chorea (PPC) is characterized by choreoathetoid movements which sometimes appear in children following cardiopulmonary bypass surgery, usually within 2 weeks of surgery. Approximately 1% of children who have cardiac surgery develop this syndrome. The mean age of affected individuals is 8–34 months. Risk factors include deep hypothermia and circulatory arrest. This syndrome is often later associated with developmental delay and neurological deficits ranging from mild learning disabilities to progressive hypotonia with obtundation. Chorea can be transient or persistent. Neuroimaging usually reveals brain atrophy without focal abnormalities. The mechanism for this syndrome is not entirely clear, but the literature suggests it may be the result of microembolic phenomena of air, fat, shards of polyvinylchloride tubing, antifoaming agents, and/or platelet fibrin aggregates accumulated during surgery. Here we present a 16 years old boy who presented with choreoathetoid movements sixteen days following aortic balloon valvuloplasty due to congenital bicuspid aortic valve. He was successfully treated with deutetrabenazine and clonazepam for 3 months.

Keywords: Choreoathetoid, Hypothermia, Aortic Balloon Valvuloplasty, Deutetrabenazine

Introduction

Chorea is a movement disorder that causes sudden, involuntary movements. Chorea occurs due to excessive dopamine activity in the brain. Dopamine is a chemical that helps control movement. PPC is a neurologic condition that can occur as a complication of major heart surgery and cardiopulmonary bypass [1,2]. The bypass machine and the dance-like movements of chorea give PPC its name. Another phrase for having had a cardiopulmonary bypass is being "on the pump." And the term "chorea" is Greek for "dance-like." pump, is known as post-pump chorea (PPC)1,2. Besides generalized chorea, comorbid encephalopathy and behavioral changes have been reported [1,2]. The first case series describing PPC was published in the 1960s by Bjork and Hultquist, about children experiencing extrapyramidal symptoms and ultimately death after congenital heart surgeries with deep hypothermia and circulatory arrest [3]. PPC is now well-characterized in the pediatric population, with an estimated incidence of 1.1-1.2% [2,4]. Conversely, PPC was not recognized in adults until the 2000s, as a rare complication with an incidence of 0.046% (2 out of 4,345 cases) in a single-center study [1,5].

Case Report

A 14-year-old school boy from rural Bangladesh, born at term by C-section, has a medical history of moderate-severe mitral aortic stenosis due to congenital bicuspid aortic valve for that he underwent cardiac surgery (aortic balloon valvuloplasty). About sixteen days after the surgery, the patient presented with hyposthenia of all for limbs associated with slower speech, mild dysarthria, and sialorrhea. These movements were involuntary, unpredictable, non-suppressible, non-distractible, and absent in sleep. Neurological evaluation revealed repeated, involuntary, non-finalistic, choreo-athetotic-like distal movements involving the right and left hand and foot that sporadically became wider until they involved the ipsilateral arm and leg too. Light hyposthenia of the both upper and lower limb and a right-hand grasp deficit were reported too. Motor, sensory, and cerebellar functions were intact, specifically no oculomotor disturbance, loss of tone, dysarthria or dysphagia. Examination of other systems is unremarkable, including absence of constitutional symptoms or rheumatological stigmata. There was no movement disorder running in the family. A careful history of the patient ruled out rheumatic fever predisposes and family history of degenerative disease or autosomal dominant/recessive inheritance i.e. Huntington and wilsons diseases and other possible risk factors for PPC evolution. He had no previous exposition to anticonvulsant, central nervous system stimulants,

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benzodiazepines, neuroleptic drugs or dopamine receptor blocking agents, which can cause tardive dyskinesia.

Investigation revealed normal complete blood count, ESR, CRP. Liver function tests, renal function tests, ANA, Urine for spot copper and MRI of brain revealed no abnormalities. Due to financial constraints and unavailability, MR angiography of brain, CSF study, EEG and other metabolic screen could not be performed.

After one month receiving 50mg/day of tetrabenazine and 1mg/day of clonazepam treatment, the choreo-athetoid movements gradually decreased till vanished completely two months later. Thus, the drugs were tapered within the next two months and the clinical follow-up examinations were completely normal.

Discussion

Post-pump chorea (PPC) syndrome is a casual movement disorder with presentation of pure chorea or choreic syndrome in association with other neurological or cognitive disorders [3]. An insidious onset in children younger than 5 years old is more linked to genetic causes. Conversely, symptoms usually begin abruptly in acquired forms after 5 years of age. In the pediatric population, Sydenham's chorea (SC) represents the most common cause of acute chorea. Autoimmune disorders, such as autoimmune encephalitis and systemic lupus erythematosus syndrome (associated with antiphospholipid syndrome), seem to have a large impact on the onset of chorea in children, and females are often the most affected. As well as autoimmune cases, viral encephalitis also plays a role in the genesis of this neurological abnormality. Herpes simplex virus, mumps, varicella parvovirus B19, and measles are typically responsible for neuronal involvement and the possible subsequent onset of chorea. In addition, the presence of other agents such as Borrelia and Toxoplasma should be excluded because of their neurotropism. Drug toxicity should be considered in the diagnostic approach of choreic disorders. A careful medical history is required for the potential intake of dopamine-receptor-blocking agents or antiparkinsonian or antiepileptic drugs. Several inborn metabolic diseases may also present with chorea, notably some specific organic acid disorders, such as glutaric acidemia type 1 and methylmalonic acidemia. When acute hemichorea is reported, it is important to evaluate common complications of type 1 and 2 diabetes. Another peculiar etiology of unilateral chorea, though less frequent in children, is vascular stroke. Most cardioembolic strokes in pediatric patients occur in the setting of cardiac surgery, as survival after this procedure has increased.

PPC has since been observed mainly by pediatric cardiovascular services as a complication of surgery for congenital cardiac defects [6]. This phenomenon typically occurs within two weeks after surgery and is transient in approximately 1/3 to 1/2 of patients [2]. Patients suffering from chorea rarely experience feeding and walking disabilities as well as dysphagia, due to bulbar chorea and orofacial dyskinesia.

The clinical features and disease course of PPC are highly variable, with few consistent characteristics identified [1,2,4,7]. Abnormal movements, in a majority of cases generalized chorea, usually appear within 14 days post-surgery (3-12 days in children, 1-14 days in adults) after a latency period of 1-7 days [1,2,7]. Common risk factors for PPC include deep hypothermia (below 25°C), rapid rewarming (above 0.18°C/minutes), long bypass time (over one hour), and extended total arrest (more than 45 min), the patient's age (with increased vulnerability under one year of age up to 5-6 years) and weight, and the presence of cyanotic heart disease [1,2,7].

Various mechanisms for explaining the causation of chorea have been previously described. The combination of extended exposure to profound hypothermia and alpha stat pH management strategy were shown to develop PPC [8]. Furthermore, the roles of increased blood viscosity with resultant thrombi formation due to extra-corporeal circulation and hypocapnia-induced cerebral vasoconstriction during the rewarming period have been suggested to contribute in neuron injury. Ischemic injuries to basal ganglia due to hypoperfusion of subcortical vascular watershed regions and altering striatal neurotransmitter systems as well as impaired function of the indirect pathway from the caudate and putamen to the internal globus pallidus, which induces inappropriate disinhibition of thalamic projections to the premotor and motor cortex are also considered to provoke chorea [9].

PPC likely represents a heterogeneous condition, and the proposed inflammatory mechanism, along with other previously noted pathogenic processes, provides a more comprehensive understanding [1,2,4,10-13]. For example, deep hypothermia and circulatory arrest, both known risk factors for PPC, have been linked to heightened neuroinflammation, manifested by increased proinflammatory cytokines in the blood and CSF [14-16]. The neuroinflammatory mechanism may also elucidate some of the distinctions between PPC in children and adults [1,2,4,5]. Children, having limited antigen exposure a priori, are believed to have less mature immune systems and more prone to autoimmunity and inflammation [17,18]. The proposed mechanism may also provide justification for the highly-variable course, particularly the late-onset and/or persistent cases, considering that the bovine tissue can continue being an antigen-presenting source long after surgery [1,2,4,7,10,11]. Furthermore, cerebral inflammation can lead to vascular injury or plasma hyperviscosity, both recognized risks for small vessel occlusion, potentially serving as a reciprocal feed-forward mechanism in the persistent PPC case [19,20].

PPC lacks definite biomarkers and hence is a diagnosis by history and exclusion, except that at times (estimated around 38.8% in one study) magnetic resonance imaging (MRI) may reveal symmetrical caudate putamen T2 hyperintensity, increased ventricular size2 and mild cortical atrophy with no focal parenchymal lesions or changes in basal ganglia perfusion [1,2,6,8]. Regarding the disease course, approximately half of the cases resolve completely, while the remainder become persistent and irreversible [1,2,6]. MRI changes may be associated with a higher likelihood of persistent diseases; poor outcome with neurocognitive developmental disorder can be seen in children, but age by itself is not an outcome predictor [1,2,6, 21-26].

Treatment primarily focuses on symptom control, traditionally with antipsychotics, anti-seizure medications, and benzodiazepines and more recently with vesicular monoamine transporter 2 (VMAT2) inhibitors [1,2,6]. Out of the available VMAT2 inhibitors, tetrabenazine have demonstrated some

efficacy, providing partial relief in two adult PPC cases [21,25]. For medically refractory individuals, deep brain stimulation targeting the globus pallidus internus may be considered [27,28]. The improvement varies from days to months according to different studies [2,29]. We used deutetrabenazine, and clonazepam, which showed favorable response.

Typically, the symptoms resolve within 2-4 weeks, although cases have been described with long-term neurological sequelae, ranging from memory, language, and motor function deficits to more severe clinical presentations such as long-term motor and cognitive disabilities [30,31].

Conclusion

To conclude, PPC syndrome is a rare condition which needs careful history and skilled neurological and psychiatric assessments for proper diagnosis. This report raised awareness of such condition, indicating its exclusive pathophysiology, risk factors and specific investigations to identify the syndrome which may easily missed. This case highlights the significance of a thorough differential diagnosis in pediatric chorea, emphasizing the importance of considering even fewer common diagnoses based on a patient's medical history and clinical presentation.

Conflict of Interest: None declared

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