Case Report

Post-Pump Chorea: Choreoathetosis Following Pulmonary Thromboendarterectomy

Safa Savadi-Oskouei\textsuperscript{a,b}, Daryoush Savadi-Oskouei\textsuperscript{a}, Aidin Baghbani-Oskouei\textsuperscript{a,*}

\textsuperscript{a}Department of Neurology, Neuroscience Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.
\textsuperscript{b}Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.

Introduction

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 [1]. It is mostly observed in children or adolescents and rarely in adults [2-4] and considered as a rare complication of open heart surgery with cardiopulmonary bypass [5, 6]. The incidence of PPC in patients undergoing hypothermic cardiac surgery is determined from 1 to 11\% [2, 7]; Medlock et al. in a study during a period of 10 years, reported an incidence of 1.2\% [8].

PPC has since been observed mainly by pediatric cardiovascular services as a complication of surgery for congenital cardiac defects [9-12]; thus, the aim of this study is to highlight this uncommon neurological disorder in an adult patient. Herein, we report a 40-year old man who developed chorea after cardiopulmonary bypass and how we approached and managed him.

Case Presentation

A 40-year old man suffering from progressive functional dyspnea since a year, had been diagnosed by pulmonary embolism and was on warfarin therapy for 6 months till the sudden onset of palpitation, cyanosis and severe dyspnea occurred in August 2018. Accordingly, he was scheduled for pulmonary artery thromboendarterectomy procedure for diagnosed massive pulmonary thromboembolism and pulmonary artery hypertension.

Anesthesia was induced with midazolam, ketamine, sufentanil, and cisatracurium with propofol as a maintenance dose. The surgery was performed successfully and no noticeable incidence was recognized throughout the procedure or in the early post-operative period. Total cardiopulmonary bypass/aortic clamping time was 58 minutes and cooling temperature was set at 20oC. He received sedative drugs while being under mechanical ventilation for 10 days after surgical treatment. Within a few days after extubation and achieving consciousness, he developed severe agitation, dysarthric speech, and cognitive disorder following irregular, unpredictable and purposeless movements involving all four extremities predominantly in the right side. No involuntary movement was present in the face or trunk. Awareness and stress exaggerated the movements while

Abstract

Post-pump chorea syndrome is a rare choreo-athetoid movement disorder following cardiopulmonary bypass. Herein, we report a rare case of post-pump chorea in a 40 year-old gentleman who was candidate for pulmonary artery thromboendarterectomy procedure. Within a few days after surgery and achieving consciousness, he developed severe agitation, dysarthric speech, and cognitive disorder following irregular, unpredictable and purposeless movements involving all four extremities predominantly in the right side. Based on the relevant medical history, imaging and laboratory findings, the diagnosis of post-pump chorea was confirmed. The symptoms improved after one month treatment with haloperidol, tetrabenazine, and clonazepam.

Keywords: Choreoathetosis, Post pump, Cardiopulmonary bypass, Hypothermia.

©2020 Swedish Science Pioneers, All rights reserved.
from chorea rarely experience feeding and walking disabilities as in approximately 1/3 to 1/2 of patients [8] typically occurs within two weeks after surgery and is transient without a rhythmic pattern. 

Muscles which flows from one part of the body to another together for days. It is characterized by irregular, unpredictable dancing mania among large numbers of people were dancing for choral dancing and was initially applied for epidemics of which induces inappropriate disinhibition of thalamic projections to the premotor and motor cortex are also considered to provoke chorea [3]. 

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 Wilson diseases and other possible risk factors for PPC evolution. He had no previous exposition to anticonvulsant, central nervous system stimulants, benzodiazepines, neuroleptic drugs or dopamine receptor blocking agents, which can cause tardive dyskinesia. Moreover, metabolic, post infectious, endocrine and immune-mediated chorea were evaluated. Laboratory findings including liver and thyroid function tests, lupus anticoagulant, CH50, C3, C4, anti-nuclear antibody, anti-neutrophil cytoplasmic antibodies, and biochemistry tests were all normal. Brain magnetic resonance imaging (MRI) showed no pathological finding in the basal ganglia and Gadolinium enhancement was not detected. Moreover, electroencephalography showed slow activity without focal abnormalities and the analysis of cerebral fluid was negative. 

Considering cardiac problems, electrocardiography showed a huge right ventricle and atrium with abnormal paradoxic septal motion indicating severe pulmonary hypertension. 

Based on the relevant medical history, imaging and laboratory findings, the diagnosis of PPC was retained by a neurologist. After one month receiving totally 10mg/day of haloperidol, 75mg/day of tetrabenazine and 1mg/day of clonazepam treatment, the choreo-athetoid movements gradually decreased till vanished completely one month later. Thus, the drugs were tapered within the next two months and the clinical follow-up examinations were completely normal. 

**Discussion**

In this report, we described a case of PPC in a 40-year-old male, in which all other relevant probable causes of choreic syndrome were excluded. The term chorea is derived from Latin word for choral dancing and was initially applied for epidemics of dancing mania among large numbers of people were dancing together for days. It is characterized by irregular, unpredictable and purposeless movements involving both proximal and distal muscles which flows from one part of the body to another without a rhythmic pattern. 

The incidence of PPC in patients undergoing hypothermic cardiac surgery is reported to be 1.2%. This phenomenon typically occurs within two weeks after surgery and is transient in approximately 1/3 to 1/2 of patients [8]. Patients suffering from chorea rarely experience feeding and walking disabilities as well as dysphagia, due to bulbar chorea and orofacial dyskinesia. Furthermore, supranuclear ophthalmoplegia and oculomotor apraxia may be developed [13]. 

Along with the benefits of deep hypothermia during cardiac surgeries, the increased risk for neurological complications such as seizure, strokes, motor disorders, peripheral neuropathy, mental status impairment and also abnormal cognitive function was observed in different studies [7, 12].

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 [12]. 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 [3].

Taken together, based on the findings of previous studies, PPC seems to associate with prolonged time on pump [14], circulatory arrest time [2, 8], deep hypothermic induced hypoxia due to impaired cerebral blood flow [12], and faster rate of rewarming after hypothermia [6]. Although our patient did not suffer a circulatory arrest, he spent a considerable amount of time on the pump at temperature of 20°C or less. 

Regarding the results of imaging modalities, some studies found increased ventricular size [8] and mild cortical atrophy with no focal parenchymal lesions or changes in basal ganglia perfusion [12]. Some others illustrated bilateral signal hyperintensity at the globus pallidus on brain MRI. In the study conducted by Passarin et al. the positron emission tomography using fluorodeoxyglucose (FDGC-PET) showed a bilateral basal ganglia hypofunction, which became normal 6 months after haloperidol administration [3]. In our patient, ventricular size was normal and no cortical atrophy or other pathological finding was observed on brain MRI. 

The only available treatment for PPC is symptomatic therapy with neuroleptic agents and the improvement varies from days to months according to different studies [8, 13]. We used haloperidol, tetrabenazine, and clonazepam, which showed favorable response. 

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. 

**Conflict of Interest**

The authors declare that there is no conflict of interest. 

**Ethical approval**

Informed consent has been obtained from the patient to publish
References: