

Interesting Cases

PANDAS: Paediatric Autoimmune Neuropsychiatric Disorder Associated with Group A Streptococci and its Intricacy with Rheumatic Heart Disease - Case Report

Meenakshi Swathi Kumaraswamy¹, Leema Pauline Cornelius², B Shanthi³

¹Junior Resident, Paediatrics, ²Professor and Head, Paediatric Neurology, ³Professor and Head, Paediatric Psychiatry, Madras Medical College (ICH & HC), Tamil Nadu, India.

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Corresponding Author:

Meenakshi Swathi Kumaraswamy, Madras Medical College (ICH & HC), Tamil Nadu, India.

E-mail Id:

meenakshiswathi95@gmail.com

Orcid Id:

<https://orcid.org/0000-0002-4087-1797>

How to cite this article:

Kumaraswamy MS, Cornelius LP, Shanthi B. PANDAS: Paediatric Autoimmune Neuropsychiatric Disorder Associated with Group A Streptococci and its Intricacy with Rheumatic Heart Disease - Case Report. Postgrad J Pediatr Adol Med. 2022;1(2):30-32.

Date of Submission: 2022-07-16

Date of Acceptance: 2022-12-05

A B S T R A C T

Paediatric Autoimmune Neuropsychiatric Disorder Associated with group A Streptococci (PANDAS) is a term used to describe certain neuropsychiatric manifestations which have a tendency to relapse with recurrent Group A streptococcal infection. The molecular mimicry hypothesised as a cause for this infection is the same as that of Rheumatic Heart Disease (RHD) which has not yet been proved. Hence there is a perpetual conflict of interest for the need of continuous antibiotic prophylaxis and their duration for secondary prevention in the paediatric population. Here, we discuss about a 9-year-old boy who presented with choreoathetoid movements and mitral valve involvement. He was treated for neuropsychiatric as well as cardiac complications following streptococcal infection under the suspicion of RHD. Due to poor compliance, the child had relapses of involuntary movements in the intermittent period which is consistent with the criteria of PANDAS. Hence high index of suspicion should be considered for all the children with features of insignificant valvular abnormalities in endemic populations during their subsequent follow-up for signs/symptoms of PANDAS.

Keywords: PANDAS, Rheumatic Heart Disease, Group A Streptococci, OCDs

Introduction

Paediatric Autoimmune Neuropsychiatric Disorder Associated with group A Streptococci (PANDAS) is specifically mentioned for children manifesting psychiatric disturbances post GAS (Group A Streptococcal) infection for which the diagnostic criteria were adapted in 1998 by Swedo SE et al.^{1,2} It has been proposed that these psychiatric symptoms are the result of a cross-reaction between autoimmune antibody response triggered by

GAS infection with that of the brain tissue responsible for manifesting Sydenham chorea. The higher incidence of rheumatic fever and prevalence of streptococcus infection makes the Indian population vulnerable to PANDAS.

Case Report

A 9-year-old developmentally normal male child 1st born of 3rd degree consanguineous, marriage presented with complaints of choreo-athetoid movements of the upper

limb for 2 weeks. His investigations revealed elevated ESR and CRP levels along with high ASO titres - 400 IU/L. Echocardiogram suggested mild AML tip prolapse with minimal MR, hence an initial diagnosis of rheumatic heart disease was considered and the child was started on penicillin prophylaxis, sodium valproate and clonazepam for involuntary movements.

The child was a defaulter who discontinued medications and presented three months later with similar complaints associated with hyperactivity. He was treated with haloperidol and trihexyphenidyl for his aggressive behaviour and then again restarted on penicillin prophylaxis.

Two years later the child presented once again with involuntary movements in the form of shrugging his shoulders, lip smacking, fidgeting and repetitive chewing suggesting OCD, which was again triggered by the discontinuation of medications. Detailed evaluation for involuntary movements and tics ruled out other possible causes like Tourette/OCDs, Wilson's disease, transient tics and systemic autoimmune diseases. MRI findings during this admission showed bilateral basal ganglia calcifications consistent with basal ganglia vasculopathy in PANDAS and his repeat ECHO suggested normal cardiac functions with no valvular involvement. Currently, the child is on sodium valproate, haloperidol, trihexyphenidyl, penicillin prophylaxis, behaviour therapy, and on long-term follow-up.

Discussion

PANDAS is a diagnosis of exclusion and there is no confirmatory biomarker to establish this condition. Several studies have been carried out to analyse their association with the onset and recurrence of streptococcal infection, suggesting that they are not immune-mediated but immune-regulated molecular mimicry responses to GAS infection. Although children in PANDAS do not have any pathological mitral valve regurgitation, they share a similar clinical course with Sydenham chorea.³ This case report emphasises the need for continuous follow-up and observation for OCDs, tics, and eating disturbances in children diagnosed with cardiac valvular involvement associated with streptococcal infection. Even though continuous antibiotic prophylaxis is advised only in patients with rheumatic heart disease, there is no clear-cut evidence suggesting the need for long-term antibiotic prophylaxis in PANDAS. The case discussed above shows a significant onset of symptoms each time during the discontinuation of penicillin prophylaxis along with high titres of ASO, thus making it difficult to distinguish between RHD and PANDAS. Neuropsychiatric manifestations in these children need standard pharmacological, cognitive, and behavioural therapy to prevent and overcome disabilities affecting their scholastic and psychosocial adjustments, thereby stressing that timely interventions will be

helpful.⁴ The potential benefits of treatment with plasma exchange/ IVIG compared with placebo were evaluated in a randomised controlled trial that included 29 children who met PANDAS criteria and were severely affected. Adverse effects, which occurred in approximately two-thirds of patients in the treatment groups, included nausea, vomiting, headache, and dizziness. Immune modulatory therapy with corticosteroids, intravenous immunoglobulin, and plasma exchange has been carried out but there is no proper evidence for a correlation between therapeutic response, their mechanism of therapeutic benefits, and the rate of antibody removal.^{5,6} In one placebo-controlled trial, an equal number of GAS infections occurred in both penicillin and placebo groups; failure to achieve adequate prophylaxis against GAS, prohibited drawing conclusions regarding the efficacy of penicillin in PANDAS prevention.⁷

Conclusion

The lack of awareness of PANDAS makes them unnoticed and under reported in an endemic country like India. Although significant carditis is not seen in PANDAS, trace/mild regurgitation may be present as ECHO findings which shouldn't be masked under the blanket of rheumatic heart disease. Hence co-ordination between paediatric neurologists, psychiatrists, and cardiologists can help in understanding and suspecting PANDAS in children coming from high-index regions of GAS prevalence, thereby making it possible to reduce morbidity associated with this disorder.

Source of Funding: None

Conflict of Interest: None

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