

Brief Report

Challenges in the Identification and Treatment of PANDAS: A Case Series

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Summary

Paediatric autoimmune neuropsychiatric disorder associated with streptococcal infection (PANDAS), is characterized by childhood-onset obsessive-compulsive disorder (OCD) and Tic disorder that has been found to have a post infectious autoimmune-mediated etiology, where the onset and subsequent exacerbations of symptoms is temporally related to group A beta-hemolytic streptococci (GABHS) infection. In addition to the use of anti-tic and antiobsessional agents, the use of Penicillin during the acute phase and for prophylaxis, tonsillectomy, immunomodulatory therapies such as plasma exchange and intravenous immunoglobulin, etc. have all been reported to improve the symptoms. We describe five cases of neuropsychiatric symptoms triggered by streptococcal infection in an Arab population and highlight the challenges faced by clinicians in the identification and management of PANDAS.

Key words: PANDAS, OCD, Tics, neuropsychiatric symptoms, streptococcal infection.

Introduction

A spectrum of neurobehavioral symptoms including Tics and Obsessive Compulsive Behaviors (OCB) occurring in association with streptococcal infection is termed Paediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infection (PANDAS) [1]. In these patients, the symptoms of Tics, are exacerbated abruptly within days or weeks following Group A Beta Haemolytic Streptococcal (GABHS) infections as evidenced by high antibodies, throat culture and sensitivity.

The National Institute of Mental Health's (NIMH) criteria for PANDAS includes: (i) Tic disorder and/or OCD; (ii) Paediatric onset, from 3 years to puberty; (iii) Abrupt onset and a course characterized by dramatic exacerbations; (iv) The onset or exacerbation is temporally related to GABHS infection; (v) Neurological abnormalities; hyperactivity, fidgetiness, restlessness or abnormal movements such as choreiform movements.

While there is no single diagnostic laboratory test available, evidence of GABH streptococcal infection

through throat culture, elevated antistreptolysin O (ASO) titer, etc. can support the diagnosis. ASO titer may be negative during the first attack or when symptomatic, but a rise in titer or if it became positive during exacerbations, that is good evidence for recent streptococcal infection. Antibodies reacting with cytoplasm of subthalamic and caudate nuclei neurons have been described in chorea and acute rheumatic fever. In this regard, elevated antistreptococcal DNAase-B (AntiDNAse-B) titer and presence of anti-basal ganglia antibodies have been reported in PANDAS.

Zabriskie and colleagues [2] found a B-cell marker (later identified as D8/D17), a monoclonal antibody attached to the surface of B cells, as a susceptibility factor for Rheumatic Fever (RF). Parents and siblings of patients with RF were also found to have the same finding, thus suggesting a genetic susceptibility. A subsequent study found that this marker was positive in 85% of patients with PANDAS, 89% of Sydenham's Chorea patients but only in 17% of controls. Thus it was postulated that those with this particular B-cell marker are at greater risk of developing CNS sequelae and neuropsychiatric symptoms after a streptococcal infection. However, subsequent finding by Murphy and colleagues [3], that this marker was positive in a group of all 31 patients with Tourette's syndrome and OCD without any association with streptococcal infection,

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