

Two cases of paediatric autoimmune neuropsychiatric disorder associated with group A beta haemolytic streptococcal infection

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Prominent neurological and/or psychiatric sequelae occur in a small percentage of children with streptococcal infection. Of these, Sydenham chorea (SC) is the best recognized syndrome. Paediatric autoimmune neuropsychiatric disorders associated with group A beta haemolytic streptococcal infection (PANDAS) is also a well-defined syndrome in which motor and/or vocal tics and/or obsessive-compulsive disorder (OCD) are the main clinical features¹.

Case 1

S.A., a 9 year old boy, presented with involuntary jerky movements of head, neck and shoulder regions of 4 months duration. Onset of symptoms was sudden and persisted at the same intensity throughout. There were no involuntary vocal utterances, excessive eye blinking or jerky movements elsewhere in the body. His development was age appropriate. S.A. had a throat infection 4 weeks before onset of symptoms, and had treatment with antibiotics. Neck movements similar to that in the current episode occurred 18 months previously, lasted 2 weeks, and disappeared without treatment. Parents were uncertain whether that episode too was preceded by a throat infection. There was no family history of movement disorders or psychiatric illness. Results of laboratory investigations were as follows: ESR 9 mm/1st hour; blood picture - red blood cells were normocytic normochromic, white blood cell count $7.3 \times 10^9/L$ with neutrophils 49% lymphocytes 41 % eosinophils 2.8 %; ECG - normal rate and rhythm; echocardiogram - small ASD 5 mm in size with a left to right shunt and awaiting review in 1 year; throat swab - beta haemolytic streptococci not isolated; ASOT - initially 800 IU /ml, and 400 IU /ml two weeks later. Treatment was oral penicillin 250 mg 6 hourly for 2 weeks. Tics were treated with

risperidone 1 mg/day with marked reduction of tic frequency at one month follow up.

Case 2

M.K., a 9 year old boy presented with a history of sudden onset jerky involuntary movements of the head and neck of 2 weeks duration. Movements were frequent, present throughout the day with associated pain. There was no facial grimacing, excessive blinking or involuntary vocal utterances. There was no past history of similar abnormal movements. There was a history of recent throat infection but no tonsillitis. No family history of tics or psychiatric disorder. The development was age appropriate. Laboratory investigation results were as follows: ESR 4 mm/1st hour; blood picture - red blood cells were normocytic normochromic, white blood cell count $10.1 \times 10^9/L$ with neutrophils 20% lymphocytes 72 % eosinophils 8 %; ECG - normal rate and rhythm; echocardiogram - no abnormalities; throat swab - beta hemolytic streptococci were not isolated; ASOT - initially 400 IU /ml and remained at 400 IU /ml two weeks later. Treatment was oral penicillin 250 mg 6 hourly for 2 weeks. Tics were treated with risperidone 1 mg/day with marked reduction of tic frequency at one month follow up.

Discussion

Published research guidelines for diagnosis of PANDAS specify the presence of a tic disorder and/or obsessive compulsive disorder; prepubertal age at onset between 3 and 12 years; abrupt onset and/or episodic course of symptom severity; temporal association between symptom exacerbations and streptococcal infection, confirmed by positive culture and/or elevated anti streptococcal titres; and the presence of neurological abnormalities during periods of symptom exacerbation². Both our patients fulfilled the diagnostic criteria except for a positive culture for streptococcal infection, probably as a result of treatment with antibiotics prior to diagnosis. Tics, but not OCD was diagnosed in our patients, though the two may occur together.

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Autoimmune antibodies to basal ganglia are implicated in the aetiology of PANDAS and the onset of symptoms of tics and OCD². It is hypothesized that the pathogenic process is triggered by an abnormal immune response to streptococcal infection in a susceptible host. Imaging studies identify a distinct subgroup of subjects with OCD and/or tics who have enlarged basal ganglia, thereby supporting this hypothesis³.

Close similarities with SC may imply that there is a potential for carditis in PANDAS too. However, carditis or valvular disease has not been demonstrated in PANDAS⁴. Accordingly, the available treatment guidelines specify that prophylactic antibiotics are not indicated^{4,5}. Also, penicillin prophylaxis was found to be ineffective in preventing exacerbation of tics and OCD symptoms in PANDAS⁶. Thus, PANDAS is identified as a clinical entity distinct from SC¹.

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