PANDAS (Paediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infection)

There is gradually accumulating evidence that there are some children who experience sudden onset of a neuropsychiatric disorder (usually obsessive-compulsive disorder (OCD) or tics) following a Group A beta-haemolytic streptococcal infection (GABHS). The acronym PANDAS was first cited in 1998 to describe this group of patients. However, neurological sequelae of streptococcal infection have been well recognised (eg, Sydenham’s chorea described by William Osler in 1894).

Doubt remains about the aetiology of the condition and whether it can be considered an independent disease entity.

More recently the term PANS (paediatric acute-onset neuropsychiatric syndrome) has been suggested, as it captures both the sudden onset and uncertainty about the aetiology.

Epidemiology

PANDAS is an uncommon condition and there is ongoing debate about its link with Sydenham’s chorea and rheumatic fever. It is likely to have a similar autoimmune aetiology and a classification which labels it as one of the acute neuropsychiatric disorders associated with streptococcal infection has been suggested.

Presentation

This is usually dramatic, with the sudden development of psychiatric/behavioural problems such as emotional lability, anxiety, night-time fears, hyperactivity and oppositional behaviour with some cognitive deficits. There may be dyskinesias - eg, mild facial or vocal tics.

The condition follows a relapsing and remitting course. For the diagnosis to be made there should be a temporal relationship between onset/exacerbation (worsening of tics or choreiform movements) and GABHS infection (throat culture or elevated anti-GABHS antibody titres). Using the stringently described criteria of Swedo et al, the diagnosis of PANDAS in a single individual requires longitudinal follow-up of the patient. The proposed working criteria which need to be met to make a diagnosis of PANDAS are:

- Presence of OCD and/or tic disorder according to DSM-5 criteria.
- Onset occurring between 3 years of age and puberty.
- Episodic course.
- Temporal association of exacerbation of symptoms with GABHS infections.
- Presence of abnormal results on neurological examination in absence of chorea.

These children tend to have more widespread neuropsychiatric difficulties than other children with OCD, including enuresis, impulsivity and deterioration in handwriting.

If overtly choreiform movements develop, the child should be considered to have developed Sydenham’s chorea and these children require antibiotic prophylaxis against subsequent GABHS infection.
Management

This is mainly supportive. The case has been made for:

- Looking for active streptococcal infection (ie take throat swabs if children develop a sore throat with pyrexia).[1]
- Subsequent treatment with antibiotics.

Some have even recommended immunomodulatory therapy.[8,9] However, results have been variable.[6]

The current recommendation is to limit the use of immunomodulatory treatments to large clinical trials in a highly selected subgroup of patients with tics or OCD.[10] Despite empirical community use of antibiotics and immunomodulatory therapies in patients with PANDAS we still lack conclusive evidence-based data about their effectiveness.

Some advocate tonsillectomy.[10]

There is some evidence and general professional opinion that conventional treatment for tics (eg, with neuroleptic agents) and OCD (eg, with behavioural therapy or selective serotonin reuptake inhibitors) is effective.[6]

Further reading & references

2. Osler W; On Chorea and Choreiform Affections. 1894.

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