Sydenham's Chorea – A Case Report and Literature Review

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Case Presentation

An 8-year-old girl with a short history of involuntary movements was diagnosed with Sydenham's chorea (SC) by Paediatric Neurology. Aetiology was suspected to be a soft tissue injury and skin infection to her knee sustained a few months before the diagnosis. She was initiated on Penicillin V prophylaxis, followed by Sodium Valproate, which did not improve her symptoms. Sodium Valproate was stopped, and Haloperidol was initiated, which also failed to improve her symptoms. Eventually, with a *combination* of Sodium Valproate and Haloperidol, symptoms of SC improved; however, with extrapyramidal side effects due to Haloperidol.

Referral to Paediatric Neuropsychiatry was made at age 12. Assessment, which included history, mental state & physical examination, revealed a range of co-morbid neuropsychiatric conditions, including anxiety with panic attacks. Some residual movements were distinct from the SC and likely subsequent onset facial motor tics in addition to some more functional tic-like behaviours. Finally, we also recognised symptoms of undiagnosed Autism Spectrum disorder. Treatment included stopping Sodium Valproate, swapping Haloperidol with a low dose of Aripiprazole and initiation of Sertraline for anxiety, combined with psychoeducation for the patient, their family and school. Self-reported symptom improvement of Anxiety: 70%, Tics: 50% and other movements (FTLBs/chorea): 70%, resulting in improved overall functioning.

Background

Sydenham's chorea (SC) is a neuropsychiatric disorder a major manifestation of acute rheumatic fever. It is characterised by hyperkinesis hallmarked with irregular, jerky movements affecting the face, limbs and trunk, associated with emotional lability, hypotonia and other neuropsychiatric symptoms.(1) The condition largely affects children and adolescents. (2) It is an autoimmune disorder of the central nervous system, associated with post - Group A Beta-hemolytic Streptococcal (GAS) infection. (3) However, symptoms can often start several months after the causative infection. (1) its pathogenesis is thought to involve dopamine receptor autoantibodies in the basal ganglia of the brain. (4) The estimated prevalence of new SC cases is 0.16 per 100 000 children aged 0–16 per year in the UK. (5) We are reporting a case assessed, diagnosed and managed at Birmingham Children's Hospital following a referral from the Department of Paediatric Neurology to the Department of Paediatric Neuropsychiatry with a focus on neuropsychiatric co-morbidities and the outcomes.

Methods

We conducted a review of literature published up until 05/03/2024 on bibliographic databases, including Pubmed, Embase, Medline, Psychinfo and EMCare. After the removal of duplicates and irrelevant studies, our search yielded 18 papers, which were a mixture of case reports, RCTs, cohort studies, expert opinions, and systematic reviews. All studies were read to give authors an up-to-date overview of all aspects of SC, including presentation, assessment, diagnosis, and management.

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Results
Assessment & Diagnosis

SC is a clinical diagnosis, with no specific diagnostic tests for a definitive confirmation. (1)

Key features

According to Jones criteria, (6,7) SC is defined as purposeless, involuntary, non-stereotypical movements of the trunk or extremities, often associated with muscle weakness and emotional lability.

Recommended investigations

Measure of inflammation (7)

- Erythrocyte Sedimentation Rate (ESR)(>60mm/hr)
- C-reactive Protein (CRP)(>3.0 mg/dL)

Evidence of (GAS) infection (7)

- Antistrptolysin-O titers (ASO, associated with GAS pharyngitis) (>450 IU/ml) (8)
- Anti-DNAse B titers (ABD, associated with GAS skin infections) (>400 U/ml) (8)

Others (9)

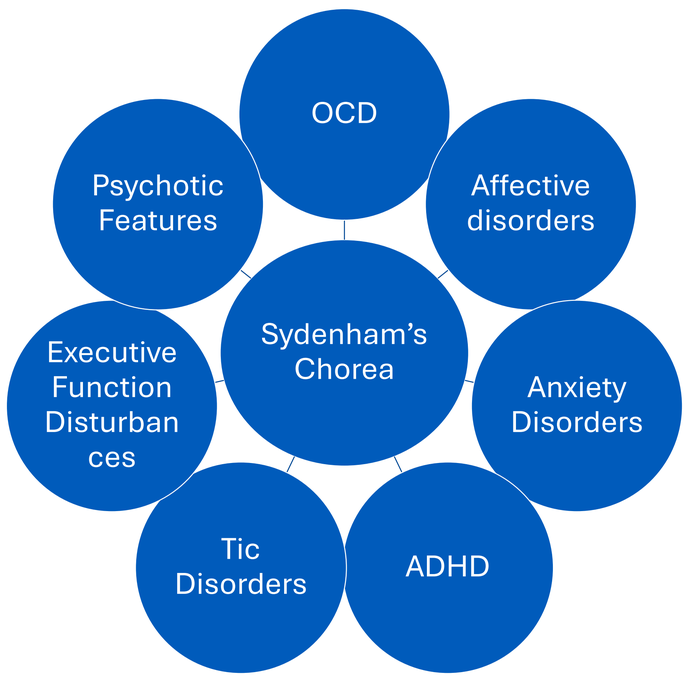
- Electroencephalogram (EEG) (to rule out cardiac involvement)
- Neuroimaging (should be normal)

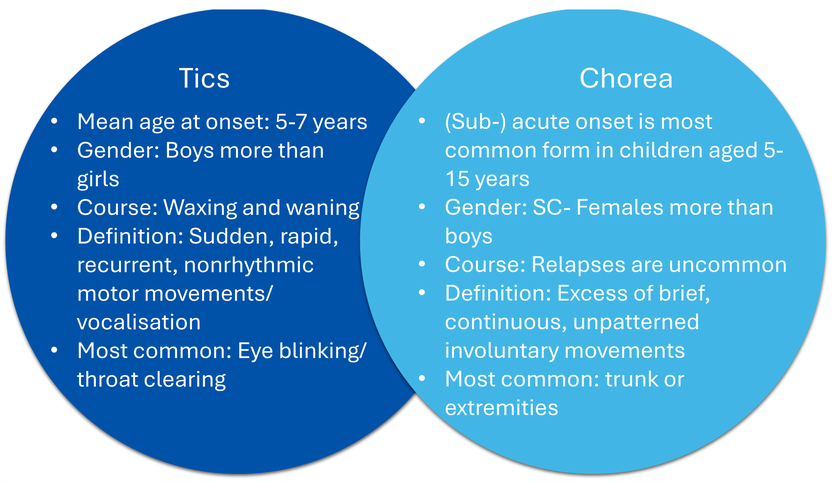
*Titers supportive evidence of SC, but absence does not preclude clinical confirmation (1)

*Tic Disorder must be excluded to confirm the diagnosis of SC (1)

Neuropsychiatric Co-morbidities

SC commonly co-occurs with other neuropsychiatric symptoms or conditions. The best evidence for OCD, ADHD, and affective disorders. Also associations with Tic disorders, anxiety disorders and psychotic symptoms. (10)





Revised Jones Criteria for Acute Rheumatic Fever(6,7)

All patients need evidence of preceding Group A streptococcal infection and... 2 Major Criteria or.... 1 Major and 2 Minor criteria

Major Criteria	Minor Criteria
- Carditis	- Arthralgia
- Arthritis	- Fever (>38.5°C)
- Chorea	- ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL
- Erythema marginatum	- Prolonged PR interval(unless carditis is a major criterion)
- Subcutaneous nodules	

Treatment

A recent systematic review of the treatment of SC made the following key conclusions.

1st Line Dopamine Agonists

- Haloperidol has the best evidence base, but higher rates of side effects. (11)
- Pimozide has fewer side effects but may act slower than haloperidol. (11)

2nd Line Antiepileptics

- Sodium Valproate has the best evidence base, and may act very quickly (within 24-48hrs). (11)

Others

- The role of immunomodulators (steroids, IVIGs) warrants further investigation for conclusive recommendations. (11)
- Penicillin can be used in the acute phase, and as prophylaxis, which appears to reduce the likelihood of further cardiac complications and the recurrence rate of chorea. (1)

Conclusion

Although SC is a rare condition, it remains prevalent in the UK, highlighting the need for collaborative work between Paediatric Neurologists and Paediatric Neuropsychiatrists with early assessment of neuropsychiatric co-morbidities. Further research is necessary to develop a consensus on the assessment and management of SC and its comorbidities.