A case study on Sydenham's chorea

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***Abstract*- Sydenham's chorea also known as St. Vitus dance is a post streptococcal rare auto immune disorder which is characterised by development of involuntary, jerky movements of the face, limbs, and trunk. Here this is a case of 19-year-old female, who was a known case of rheumatic chorea since 2016 secondary to rheumatic fever which was progressed to mitral valve prolapse (MVP) in 2019, taken prophylactic injection Benzathine penicillin (Inj. Penidure) every 21 days till 19 years of age. Left medication since 2 months and she was brought to the hospital with 7 days history of abnormal or shaking movements of right upper limb and lower limb with slurred speech. 2D ECHO cardiogram shows rheumatic heart disease, moderate eccentric MR (mitral valve regurgitation). Physicians followed the WHO recommended guidelines for the treatment to prevent future streptococcal infections. The duration of the treatment is depended on the severity of cardiac involvement.**

***Key words*- Sydenham's chorea, Rheumatic chorea, St. Vitus dance, Autoimmune disease, Benzathine penicillin, Streptococcal infection, Rheumatic fever.**

1. Introduction [1,2]

Sydenham's chorea (Rheumatic chorea) also called as St. Vitus dance is a post streptococcal rare auto immune disorder which is characterised by development of involuntary, jerky movements of the face, limbs, and trunk.[1] It primarily affects children and adolescents, with peak incidence between 5 and 15 years of age. The condition is considered an autoimmune response, where the body's immune system attacks its own tissues, leading to inflammation and dysfunction in the basal ganglia of the brain.[2]

2. Aim of the case study

The purpose of this case study is to present a comprehensive analysis of patient diagnosed with Sydenham's chorea describing the clinical presentation, diagnostic challenges and management strategies employed. This study also includes epidemiology, aetiology, pathophysiology, risk factors, diagnostic tests, complications and treatment of Sydenham's chorea.

3. Epidemiology [1,2,3,4]

The incidence of rheumatic fever and Sydenham's chorea in the USA and western Europe has decreased since World War II as a result of improved health care and increased antibiotic use. In the top end of the northern territory in Australia, an area predominantly occupied by indigenous people. The point prevalence of rheumatic fever was 9.6 per 1000 people aged 5-14 years in 1995.[3] This Sydenham's chorea affects individual of all races and ethnicities. 25% of individuals with rheumatic fever develop Sydenham's chorea. [1] It is estimated that there are 50,000 cases of acute rheumatic fever per year in India.[4]

The multicentric survey which was conducted by the Indian council of medical research in school going children shows that rheumatic heart disease and rheumatic fever continues to major public health problem.[2]

Table – 01 OCCURRENCE OF SYDENHAM'S CHOREA IN DIFFERENT COUNTRY

Country	Prevalence	Age Group	Gender	Streptococcal Infection Rates
United States	Rare	Children/Adolescents	Slight female preponderance	Varies, typically lower in recent years
United Kingdom	Rare	Children/Adolescents	Slight female preponderance	Varies, typically lower in recent years
India	Relatively more common	Children/Adolescents	Slight female preponderance	Higher due to increased burden of streptococcal infections
Australia	Rare	Children/Adolescents	Slight female preponderance	Varies, typically lower in recent years
South Africa	Relatively common	Children/Adolescents	Slight female preponderance	Higher due to increased burden of streptococcal infections
Brazil	Relatively common	Children/Adolescents	Slight female preponderance	Higher due to increased burden of streptococcal infections
China	Rare	Children/Adolescents	Slight female preponderance	Varies, typically lower in recent years

Nigeria	Relatively common	Children/Adolescents	Slight female preponderance	Higher due to increased burden of streptococcal infections

4. Etiology [5,6]

- Sydenham's chorea is believed to be autoimmune disorder. It is delayed 6 to 8 weeks after streptococcal pharyngitis. The main cause is group A beta hemolytic streptococcal (GABHS) infection.[5]
- Other autoimmune causes, such as seen in systemic lupus erythematosus. [6]
- Genetic causes: [6]
- Benign hereditary chorea starts in childhood and is a non-progressive chorea.
- Inheritance is usually autosomal dominant, although rare cases of autosomal-recessive and X-linked inheritance have been reported.
- Wilson's disease is an autosomal-recessive disorder of copper metabolism.
- Ataxia telangiectasia and other related conditions.
- Huntington's disease presents most often between the ages of 35 years and 45 years but it can be younger, especially if inherited from the paternal line. There is usually but not invariably, a family history. A juvenile form exists that should be seen as a variation of the normal form and not a distinct entity.
- Athetoid cerebral palsy.
- Drug-induced causes - metoclopramide, phenothiazines and haloperidol are the most important.
- Primary and metastatic brain tumours affecting the basal ganglia.
- Metabolic - bilirubin encephalopathy and toxins, especially carbon monoxide, manganese and organophosphate poisoning.

5. Pathogenesis [1,7,8]

Sydenham's chorea is a major manifestation of acute rheumatic fever which is caused by GABHS plays the major pathogenic role inducing heart valve and articular damage.[7]

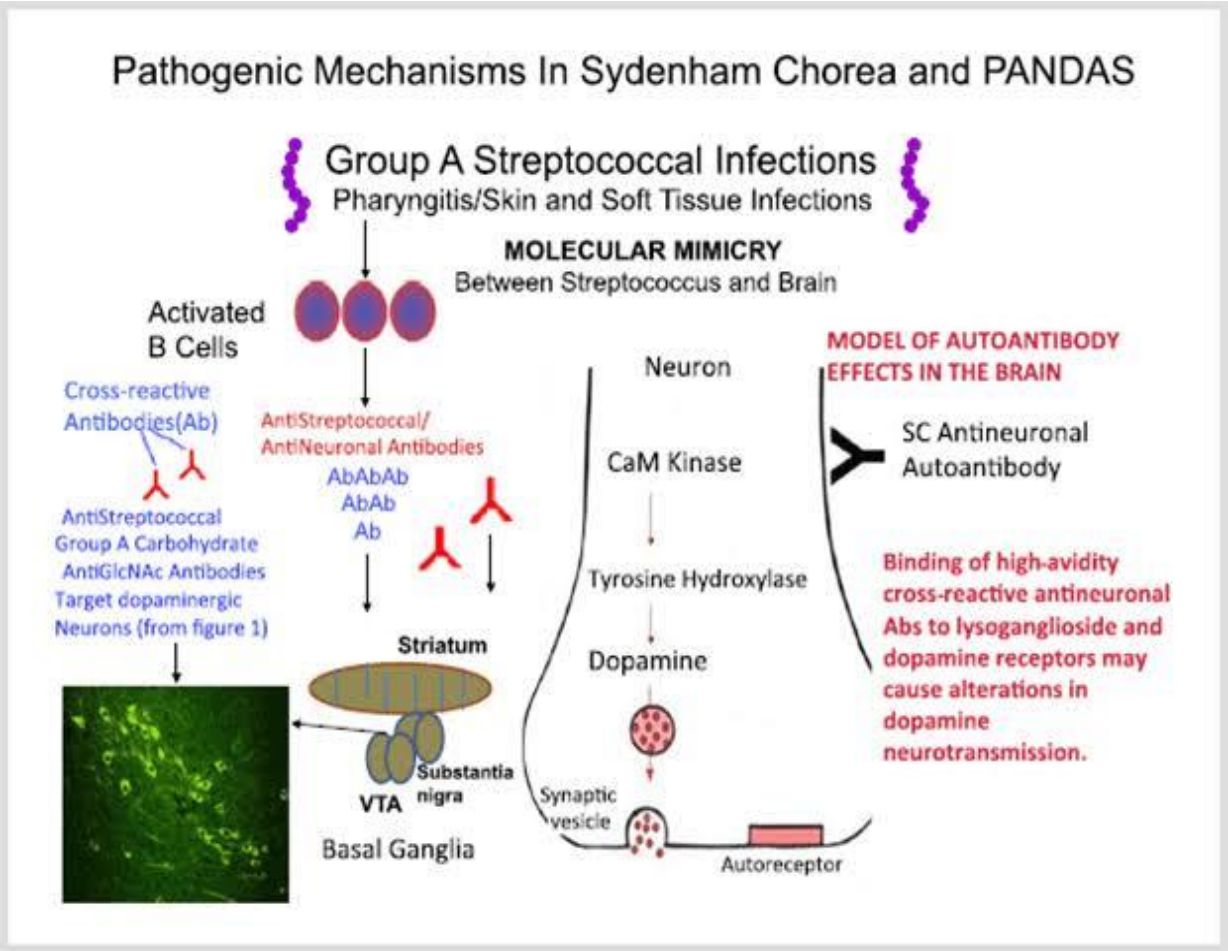
The exact mechanism that causes SC is poorly understood. Researchers believed that antigens (substances that are capable of stimulating an immune system response) on streptococcal bacterial cells are similar to antigen found on brain cells of human body.[1]

When the immune system creates antibodies to fight streptococcal infection, the antibodies also, in genetically predisposed individuals, mistakenly bind to healthy cells.[1]

When they bind to brain cells in basal ganglia epitopes, disrupting cortico basal ganglionic thalamic circuits (CBGTC), leading to motor, behavioural and cognitive symptoms.[1]

Neurotransmitters such as gama-amino butyric acid (GABA), dopamine, noradrenaline and serotonin play a major role in motor movements, attention and emotions.[8]

Motor movements is particularly controlled by dopamine. Excess of dopamine release results in jerky and hyperactivity movements.[8]



6. Risk factors [9]

This Sydenham chorea occurs by the infection Of GABHS so the risk factors include:

- Streptococcal Pharyngitis
- Rheumatic fever
- Rheumatic heart disease
- Over crowding
- Poor hygiene
- Autoimmune disease
- Huntington's disease
- Genetics

7. Table - 02 clinical manifestations [10,6]

Physical symptoms [10]	Clumsiness Involuntary movements Muscular weakness Hand wringing Restlessness Slight grimacing Stumbling &falling
Speech issues [10]	Slurred speech Vocal outbursts
Cognitive and emotional symptoms [10]	Anxiety Difficulty in concentrating
Psychological symptoms [6]	Emotional lability Obsessive-compulsive behaviour Cognitive defects Personality change Attention deficit & hyperactivity

8. Diagnostic tests [11,12]

1. Antibodies to streptococcus to group A

a. ASO (antistreptolysin O) positive > 200 Todd units. a rising titer is more significant

b. Others: anti-deoxyribonuclease B, anti-hyaluronidase, anti-streptokinase
2. Rapid antigen detection test: latex agglutination or enzyme immune assay on throat swab
3. ESR & CRP (raised)
4. ECG: prolonged PR (first degree heart block), second degree or complete heart block
5. 2-D echo [11]
6. **Jones criteria** [12]

Table - 03

Population	Criteria	
	Major	Minor
Low risk	Carditis (clinical or subclinical)	Polyarthralgia
	Arthritis (polyarthritis only)	Fever (>38.5C)
	Chorea	ESR > 60 mm/hr or CRP >3.0 mg/dL (or both)
	Erythema marginatum	Prolonged PR interval (unless carditis is major criterion)
	Subcutaneous nodules	
Moderate and high risk	Carditis (clinical or subclinical)	Monoartharalgia
	Arthritis (monoarthritis, polyarthritis, polyarthralgia)	Fever (>38C)
	Chorea	ESR > 30 mm/hr or CRP >3.0 mg/dL (or both)
	Erythema marginatum	Prolonged PR interval (unless carditis is major criterion)
	Subcutaneous nodules	

9. Treatment [12]

A. General measures

The patient should be kept at strict bed rest until the temperature returns to normal (without the use of antipyretic medication) and the ESR, plus the resting pulse rate, and the ECG have all returned the base line.

B. Medical measures

1. Salicylates

The salicylates markedly reduce fever and swelling they have no effect on natural course of the disease. Adults may require large doses of aspirin, 0.6-0.9g every 4 hours; children are treated with lower doses.

2. Penicillin

Penicillin (benzathine penicillin, 1.2 million minutes intra muscularly once, or procaine penicillin, 6 lakh units intramuscularly daily for 10 days) is used to eradicate streptococcal infection if present. Erythromycin may be substituted (40 mg/kg/day)

3. Corticosteroids

There is no proof that cardiac damage is prevented or minimized by corticosteroids. A short course of corticosteroids (prednisolone, 40-60 mg orally daily, with tapering over 2 weeks) usually causes rapid improvement of the joint symptoms and is indicated when response to salicylates has been inadequate.

4. Dopamine D2 receptor blockers

Drugs like Tetrabenazine and Pipamperon can be used to treat chorea.

5. Anticonvulsants

Drugs like Valproate, carbamazepine can be used in case of convulsions.

6. Neuroleptics (Antipsychotics)

Drugs like Risperidone, olanzapine is another option to treat chorea

10. Complication [10]

- Behavioural changes and inattention
- Obsessive compulsive features
- Tics
- Arthritis
- Endocarditis
- Heart valve disease

11. Case report

A 19-year-old female patient was admitted in Vijayanagara institute of medical science, Ballari (Karnataka) with chief complaints of abnormal or shaking movements of right upper limb and lower limb with slurred speech since 1 week.

Her past history reveals that she was diagnosed as rheumatic chorea in 2016 secondary to rheumatic fever by antistreptolysin O test. Which was progressed to mitral valve prolapse (MVP) in 2019, taken prophylactic injection Benzathine penicillin (Inj. Penidure) every 21 days till 19 years of age. Left medication since 2 months.

No history of convulsion, fever, vomiting and diarrhoea.

On examination her BP was found to be 110/70 mm of Hg, Pulse rate was found to be 80 bpm.
On systemic examination all were found to be normal.

Table – 04 Admission laboratory results

CBC	Hb: 8.5 g/dL (reduced) WBC: 8460 cells/cumm (normal) RBC: 4.61 million/cumm (normal) Platelets: 3.64 lakh/cumm (normal)
Liver function test	Globulin: 3.5 g/dL (increased) A/G ratio: 1.0 (reduced)
Renal function test	Serum creatinine: 0.6 mg/dL
2D Echo cardiogram	• Rheumatic heart disease (RHD) • Moderate eccentric MR (mitral valve regurgitation)
MRI	No significant abnormalities

Treatment

The patient was initially managed with intramuscular Benzathine penicillin as a stat dose to treat rheumatic chorea, inj. Pantoprazole 40 mg OD for 4 days to treat GI irritation, inj Neurobion forte 1 ampule in 100ml NS for 2 days to treat anaemic condition, tab Aspirin 150 mg OD for 4 days, tab Atorvastatin 20 mg OD for 4 days, tab Tetrabenazine (Revocon) 25 mg BD for 3 days from day 3 to treat abnormal jerky movements, tab Prednisolone 20 mg started on day 3 which was slowly tapered to 10 mg.

Table – 05 Discharge medication

Name of the medication	Dose	Route	Frequency	Days
Benzathine penicillin	1.2 MIU	IM	1.2 MIU	Every 21 days
Tetrabenazine	25 mg	PO	½- 0- ½	7 days
Pantoprazole	40 mg	PO	1-0-0	7 days
Prednisolone	5 mg	PO	1-0-0	3 days

12. Discussion

Sydenham's chorea is a rare neurological auto immune disorder. Which is caused by GABHS infections. This condition primarily affects the children of 5-15 years of age where females develop this type of chorea more commonly than males which is characterised by sudden onset of involuntary movements, non-rhythmic and non-repetitive twitching of limbs.

In this case, the female patient of 19 years was admitted with a complaint of abnormal/shaking movements of right upper limb and lower limb with slurred speech since 1 week. And her past history reveals that she was a known case of Sydenham's chorea with mitral valve prolapse, initially the treatment was started with prophylactic injection Benzathine penicillin (Inj. Penidure) every 21 days where she discontinued this medication 2 months ago. So based on her past medical history and present complaints the physicians has advised for CBC, Liver function test, Renal function test, 2D Echo cardiogram and MRI to rule out any abnormalities. In which 2D echo shows Rheumatic heart disease (RHD) with Moderate eccentric MR (mitral valve regurgitation). The treatment was initiated with antibiotic as a prophylactic, proton pump inhibitor, corticosteroid to treat inflammation, antiplatelet agents and antihyperlipidemic to prevent cardiac complications, tetrabenazine is given to treat chorea, neurobion-forte is given to treat anaemic conditions and vitamin deficiency.

13. Conclusion

Diagnosis and management of Sydenham's chorea is very difficult and best done with inter-professional health care team.

In generally when Sydenham's chorea is present in the patient, that person should be referred to cardiologist to rule out the valvular heart disease.

Physicians followed the WHO recommended guidelines for the treatment to prevent future streptococcal infections. The duration of the treatment is dependent on the severity of cardiac involvement.

Prevention of Sydenham's chorea is properly based on timely, Rheumatic fever treatment may lower the risk of condition and the long-term focus of treatment is to prevent permanent damage to heart valves due to heart disease.

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