Sydenham's chorea

Synonym: St Vitus' dance

What is Sydenham's chorea?

Sydenham's chorea is a neurological manifestation of rheumatic fever (RF). Chorea (from the Greek word meaning 'dance') occurs in 20-40% of patients with RF.[1] Although it is the most common cause of acute chorea worldwide, cases are now rare in the developed world.[2]

An autoimmune process is triggered after infection with group A beta-haemolytic streptococci (GABHS). Molecular mimicry between streptococcal and CNS antigens is thought to underly the pathogenesis of this disease. Cross-reactive anti-basal ganglia antibodies (ABGAs) are formed in genetically predisposed patients and these are thought to cause the neurological insult.[3]

Thomas Sydenham described chorea in 1686 in a work entitled *Schedula monitoria de novae febris ingressu* but the association with RF was not appreciated until 180 years later by Roger in 1866.[4] The Jones Criteria for the diagnosis of RF originated in 1944; however, only on their revision in 1992 was Sydenham's chorea accepted as a diagnostic feature of RF.[5] [6]

Much of the literature concerning Sydenham's chorea is very old. The disease itself appears to have changed over time. It is substantially less common than it was but also symptoms tend to be less severe and relapses less frequent. This may be partially due to penicillin but improved social conditions and a natural reduction in the virulence of the streptococci have also played a role.[1] [7]

How common is Sydenham's chorea? (Epidemiology)

- Typical age of onset is 5-15 years.

- There is a female preponderance reported in most studies.
- A predisposition runs in families.
- The incidence of Sydenham's chorea reflects that of RF. It is rarely seen in developed countries nowadays. In developing countries, the rate is much higher but with much variation and often poor recording.
- Both primary and recurrent episodes are equally common.

Risk factors

Overcrowding and poor hygiene are important.

Clinical features

It often presents with other features typical of RF, at least on the first occasion; however, it can present in isolation, especially if it is a repeat attack. Typically, the first episode occurs 4-8 weeks after an episode of GABHS pharyngitis.

Features of chorea are:

- Involuntary movements.
- Hypotonia.
- Muscular weakness.
- Psychological features.

Movement disorders [8] [9]

- Chorea is an involuntary and random movement such as an arm flying up. This is worse when 'under pressure'. The child may try to convert this into an apparently purposeful movement by pushing back their hair or scratching their nose.
- Hemichorea occurs in about 20% but careful examination will often show milder features on the other side.
- It involves the extremities and the face and they may be accused of 'making faces'. Movements occur at rest and disappear in sleep.

- Chorea interferes with voluntary movements and causes a clumsy gait, dropping and spilling and explosive bursts of dysarthric speech.
- Ask the patient to extend their arms and spread their fingers. Their fingers will move in a way like playing the piano. Muscular weakness leads to inability to maintain a steady grip. Ask the patient to grip your hand or wrist tightly and maintain it. The fluctuating strength of the grip is called 'milkmaid's grip'.
- The 'pronator sign' is hyperpronation of the hands, with the palms facing outward when the arms are held over the head.
- Tone is typically decreased, drastically in some: about 2% of children become bedridden (chorea paralytica).
- Other motor findings include motor impersistence and tics.

Psychological symptoms [10] [11]

Psychological symptoms precede chorea and may be relapsing and remitting. They include:

- Emotional lability.
- Obsessive-compulsive behaviour.
- Cognitive defects.
- Personality change.
- Attention deficit and hyperactivity.

Diagnosis [8]

Many of the laboratory features of RF may be absent, especially if the onset of the disease was a significant time after the streptococcal infection and RF may not have been apparent. Chorea can lag up to six months behind the streptococcal infection. There is no single diagnostic test. Other causes of chorea need to be excluded and often the diagnosis is one of exclusion in a child of the appropriate age.

- By the time that chorea presents it is unusual to be able to isolate GABHS from the pharynx.
- Blood count may be normal and ESR and CRP returned to normal.

- Antistreptolysin O titre and other serological tests described with RF may still be helpful.
- Antineuronal antibodies have been described but they do not form part of routine diagnosis. The antibody titre falls as the disease improves and rises again in relapse. [12]
- MRI studies are often normal but MRI spectroscopy may reveal autoimmune damage to the basal nuclei. [13]

Chorea gravidarum

This is a descriptive rather than an aetiological diagnosis. It simply refers to chorea in pregnancy. In 1900, its incidence was described as high as 1 in 300 deliveries but now it is an extreme rarity in the UK.

- Most cases are a resurgence of Sydenham's chorea in the hormonal milieu of pregnancy but it can also be precipitated by the use of oral contraceptives.
- In the 1930s, at least 35% of patients had a definite history of acute RF (ARF) or Sydenham's chorea and 4% of those with chorea gravidarum had ARF. [14]
- 21% of women experience recurrent chorea with subsequent pregnancies and several cases have been described in which attacks occurred in three, four and even five pregnancies. [15] [16]
- If left untreated, the disease resolves in 30% of patients before delivery but, in the other 70%, it persists until the puerperium. Symptoms frequently fade rapidly in the days after delivery.
- The mortality rate has declined since the 1930s and is now below 1%.

Differential diagnosis

In the 1930s, if a doctor saw a patient with chorea, especially if the patient were a child or young woman, it was a reasonable assumption that the diagnosis was Sydenham's chorea. In Western societies today, such a presentation is **unlikely** to be Sydenham's chorea and considerable thought must be given to the differential diagnosis. The time course of the chorea is useful diagnostically: most previously healthy children with an acute or subacute chorea have an autoimmune aetiology.[17] Additional causes of childhood choreas include:

- Other autoimmune causes, such as seen in systemic lupus erythematosus.
- Genetic causes:
    - 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.[18]
    - 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.[19]
- 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.

Associated diseases

Most patients with Sydenham's chorea will have other manifestations of RF: [20]

- 60-80% have cardiac involvement, particularly of the mitral valve.
- 30% have arthritis and joint involvement.

Sydenham's chorea treatment [8]

- Rest is usually advocated, as traditional teaching is that this reduces the risk of rheumatic heart disease, although this is not evidence-based.
- The most important consideration is secondary prophylaxis, usually with penicillin, to prevent further streptococcal infections.
- A number of drugs may help control the chorea, although many of these can have extrapyramidal effects of their own. Probably the best documented of these is haloperidol but valproate is now most specialists' first-line choice. [20]
- Dopamine receptor-blocking drugs such as pimozide are used for non-responders or those with chorea paralytica (unlicensed use). [21]
- Immune therapies (eg, plasma exchange, intravenous immunoglobulins) may play a role in selected patients. [22]
- Intravenous methylprednisolone followed by a tapering course of oral prednisone have been used to good effect. [23]
- See the separate Rheumatic Fever article regarding the treatment of cardiac complications.
- One case report showed complete resolution of severe Sydenham's chorea with plasmapheresis. [24]

Prognosis

- Commonly, Sydenham's chorea is self-limiting with spontaneous remission. Symptoms generally improve in a week or two and are better by 8-9 months, rarely lasting a year; however, they may occasionally wax and wane for up to 10 years.
- It does not result in any neurological deficit.
- About 20% of patients have a recurrence, usually within two years of the initial attack.[25]

Further reading

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