Sydenham chorea in a 5-year-old Saudi patient

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ABSTRACT

بالرغم من التقدم الملحوظ في الرعاية الصحية وارتفاع المستوى المعيشي للسكان في المملكة العربية السعودية، ما تزال الحمى الرثوية تسجل حضورها في العديد من مناطق المملكة وإن كانت بنسب أقل من الماضي. عادة ما تظهر الحمى الرثوية عند الأطفال في سن الدراسة، إلا أنها قد تصيب من هم أصغر من ذلك. يتبدى المرض حين ظهوره في الأعمار الباكرة بإصابة التهاب القلب والمفاصل، أما إصابة الجملة العصبية (رقص سيدنهام) فأقل تواترا. يصف هذا المقال حالة غير مألوفة لطفل يبلغ من العمر 5 أعوام أصيب بالحمى الرثوية تمثلت أعراضها بما يعرف بداء الحركات الرقصية.

Despite improvements in socio-economic status and the standard of health care services, rheumatic fever continuous to occur in Saudi Arabia, although with decreasing frequency. The disease is most commonly observed in school-aged children, but can also occur in a younger age group. Carditis and arthritis are the major clinical symptoms on presentation of acute rheumatic fever in young children. Rheumatic chorea is infrequently reported in young children. Here, a case of Sydenham chorea, in a 5-year-old boy, is presented. Although rare, the diagnosis of Sydenham chorea should always be considered in young children with choreiform movements.

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Acute rheumatic fever (ARF) is most frequently seen in school-aged children. The disease and its neurological manifestation, Sydenham chorea, are considered rare in preschool aged children (<5 years of age). As the most common cause of acquired chorea, Sydenham chorea must be included in the deferential diagnosis of choreiform movements among young children. Failure to recognize this disease may increase the risk of recurrence of ARF as well as rheumatic heart disease. We present a case of Sydenham chorea in a 5-year-old boy. The objective in presenting this particular case is to highlight the need to raise awareness of the possibility of Sydenham chorea in young children.

Case Report. A 5-year-old boy presented to the emergency department with involuntary movements of the extremities, facial grimacing, fluttering eye movements, and slurred speech. The onset of symptoms was approximately 2 weeks prior to presentation, when he began to display unusual hyperactivity and clumsy movements that disappeared while sleeping. There was no history of behavioral change or emotional lability. Frequent attacks of tonsillopharyngitis were reported. The last episode was reported to occur 2 months earlier; at this time he received a 3-day course of Azithromycin. The medical history was otherwise negative. The family history was unremarkable for rheumatic fever, social stress, psychiatric illness, and tic disorders. A normal vaginal delivery following an uneventful pregnancy was reported with a normal post-natal course. All vaccinations were received on schedule and the developmental milestones were appropriate for age. On clinical examination, the temperature was 36.6°C, heart rate 100 per minute, blood pressure 101/58 mm^3 and the respiratory rate 28 per minute. The growth measurements were within normal limits. The ear-nose and throat examination revealed hypertrophic tonsils without signs of inflammation. The cardiovascular system was normal; there were no murmurs detected. The neurological exam showed that he was alert and oriented to time, place, and persons. The thought processes were clear and the child was obviously intelligent. Choreiform movements were apparent on

the face and upper limbs. The cranial nerves, including extra ocular movements, were intact; the sensation exam was normal. There was normal tone and strength on examination. The patient had difficulty with hand grip "milkmaid's sign", and showed sporadic tongue protrusion. His deep tendon reflexes were normal and symmetrical throughout, and the toes were down going. No other stigmata of rheumatic fever were identified. Special investigations revealed a normal complete blood count, as well as normal liver, and renal function. The acute phase reactants included the erythrocyte sedimentation rate, which was 16 mm/hour and the C reactive protein, which was negative. A throat culture was negative for group A streptococcus and the anti-streptolysin O titer was unremarkable. Rheumatic heart involvement was excluded by electrocardiogram and echocardiography. The MRI of the brain was normal. (**Figure 1**). Sydenham chorea was considered as the most likely diagnosis, and long-term Penicillin prophylaxis in the form of monthly Penicillin G benzathine at a dose of 600,000U intra-muscularly was started. Within a week of admission he improved with good control of motor activity, reduced involuntary movements of the extremities and facial grimacing. Spontaneous speech remained reduced but without impairment of communication. He continued to improve throughout his follow up visits and complete resolution of symptoms was observed 5 months from the onset of illness.

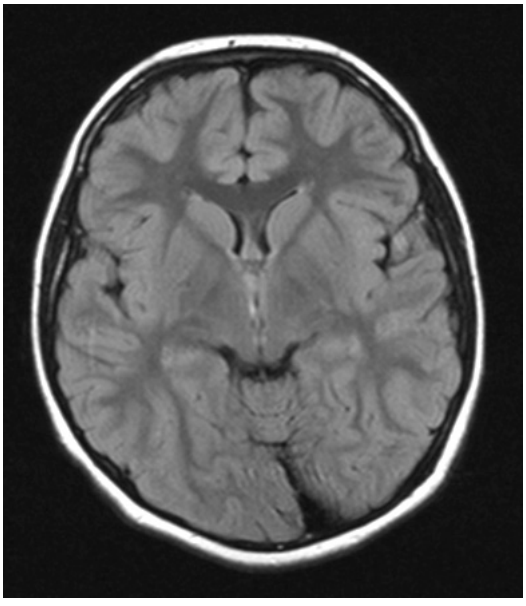

Figure 1 - The T1 brain MRI of a young patient with Sydenham chorea.

Discussion. Recent hospital based studies have shown a dramatic decline in admissions for ARF in Saudi Arabia.[1,2] Rheumatic fever (RF) has been considered a disease of the young (commonly between 5-15 years of age).[3,4] The disease is uncommon in children less than 5 years of age. Published data estimates that ARF occurs in 1-6.8% of children less than 5 years of age.[4,5] Arthritis and carditis are the most common clinical presentation in this age group.[5,6] Although reported in a boy at the age of 2 years and 8 months, Sydenham chorea occurs most often in children before puberty with a female preponderance.[7,8] The incidence of rheumatic chorea in young children varies from almost nonexistent in some series to 11% in another series.[5,6] The patient described in this case report is a young Saudi child diagnosed with Sydenham chorea.[9] The typical neurological signs together with the preceding history of pharyngitis, points towered the diagnosis of Sydenham chorea. The absence of laboratory confirmation of group A streptococcal infection does not rule out the diagnosis of rheumatic chorea.[10] Although rare, the diagnosis of Sydenham chorea should always be considered in young children with choreiform movements. A regimen of continuous penicillin prophylaxis against streptococcal infection reduces the risk of ARF recurrence and the development of subsequent heart disease.

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