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(Accepted 30 August 2000)

Lesson of the week

A painful hip as a presentation of Guillain-Barré syndrome in children

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Pain of the lower limb in children is a symptom of many conditions and can lead to diagnostic difficulties; the clinician may search for a musculoskeletal rather than a neurological cause. Guillain-Barré syndrome must be considered when a child presents with pain of the lower limb. Misdiagnosis is more common in the paediatric population because the child's history and cooperation in the neurological examination are often limited. Failure to diagnose Guillain-Barré syndrome and to initiate prompt treatment is potentially life threatening as the disease can quickly lead to respiratory failure and death from muscle paralysis. We describe a child with Guillain-Barré syndrome who initially presented with a painful hip.

Case report

A two year old boy presented with pain in his right leg. Three weeks earlier he had had tonsillitis, fever, and diarrhoea. His doctor had treated him with amoxicillin, and his symptoms subsequently settled. The pain in his leg started two days before admission. He limped and was reluctant to walk. The pain was precipitated by walking and was vaguely localised to the hip. He had no other joint symptoms or relevant history. On examination he was apyrexial but irritable. He had difficulty sitting and refused to stand or weight bear. Neurological examination revealed intact cranial nerves, although the gag reflex was depressed, which was partly attributed to his uncooperative behaviour. However, there was no indication that he had been choking.

Examination of the upper limbs revealed normal tone and power, but reflexes of the biceps, triceps, and supinator were diminished. The patient was distressed on flexion, extension, and internal rotation of the right hip. Otherwise the joints had full range of movement with no swelling or tenderness, although the patient preferred to lie with his hips flexed and mildly rotated externally. The patella and ankle reflexes were diminished bilaterally. The plantar responses were extensor.

Ultrasonography of the hips showed a symmetrical appearance with no synovial thickening or effusions. A radiograph of the hips showed a slightly irregular epiphysis on the right side but otherwise gave normal results. A bone scan was later performed, but no definitive abnormality was found.

Tests for blood count, erythrocyte sedimentation rate, and C-reactive protein concentration gave normal results. An orthopaedic opinion was sought, and an irritable right hip was considered likely. The patient was initially given paracetamol for analgesia, but because of increasing hip pain and irritability he was given Oramorph (Boehringer Ingelheim) and his right leg was placed on skin traction.

On day 4 after admission he continued to deteriorate; he mentioned pain in his neck, elbows, and wrist. He became weak, lethargic, and disinterested in feeds. He was unable to sit up independently. A musculoskeletal cause was considered, and he was referred to a rheumatologist at a tertiary centre. On examination only generalised limb weakness and hypotonia were found, and a neurological opinion was subsequently sought. By this time he had started to cough and was

Guillain-Barré syndrome should be part of the differential diagnosis of children with painful lower limbs

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BMJ 2001;322:149-50

aspirating fluids. His voice was now much softer, and he exhibited a weak high pitched cry. He had a weak gag reflex, and antigravity movements in his upper and lower limbs were minimal. Tone was symmetrically reduced, and he was now areflexic.

On day 8 after admission, he underwent electromyography and nerve conduction studies. They showed a patchy demyelinating polyneuropathy, with all nerves involved. Guillain-Barré syndrome was diagnosed. He was treated with a high dose of immunoglobulins. He did not require respiratory support and quickly improved. The pain subsided gradually, and he received speech therapy and physiotherapy. At outpatients' follow up four months later he had made a complete recovery.

Discussion

Pain of the lower limb is an important symptom to recognise and diagnose in childhood. It is the presenting complaint in many conditions. The commonest causes of pain are transient synovitis, avascular necrosis of the femoral head (Legg-Calvé-Perthes disease), osteomyelitis, septic arthritis, and trauma. Guillain-Barré syndrome is a relatively uncommon condition, estimated at 0.8 per 100 000 person years in people under 17 years.¹ It is now, however, the commonest cause of acute general paralysis in developed countries.² The literature shows a wide spectrum of clinical presentation for Guillain-Barré syndrome but classically it presents as a progressive ascending symmetrical weakness with areflexia. On a background of progressive motor paralysis, there are sensory symptoms such as paraesthesia and pain. These symptoms are often overlooked and underrated in clinical practice, especially in children, and are probably undertreated. They provide clues, however, that are vital to the early diagnosis of Guillain-Barré syndrome.

Pain as a primary complaint in paediatric Guillain-Barré syndrome, commonly preceding onset of motor paralysis, has been reported.³⁻⁶ The pain may be severe enough to cause children to become irritable and withdrawn, and this may lead to misdiagnosis. Many children who have presented with pain as an early symptom of Guillain-Barré syndrome have been initially diagnosed with an encephalopathy or a musculoskeletal disease such as osteomyelitis.⁷ Nguyen et al retrospectively reported on a series of children under six years old with Guillain-Barré syndrome.⁸ They found that pain was a symptom in all the children at some time during their stay in hospital and was present on admission in 23 of 29 (79%). Pain was often the most important symptom and led to misdiagnosis in 20 patients (69%). Back pain and lower limb pain aggravated by elevation of a straightened leg were the most common pain syndromes, which were present in 24 patients (83%). Moulin et al's prospective longitudinal study found that 89% of patients (mean age 50.6 years) with Guillain-Barré syndrome also complained of pain during the course of their illness.³ Moreover, pain intensity on admission was found to correlate with presenting neurological deficits, but it was not a good predictor of prognosis. Differentiating the reluctance to move because of limb pain from neurological weakness at the outset of Guillain-Barré syndrome is difficult because children are generally poor historians, and their cooperation during neurological assessment

is often limited. For example, a child may seem to have an ataxic or antalgic gait, which masks the underlying neurological deficit. Younger children especially find it difficult to express their discomfort, which can also mislead the clinician.

Pain of the lower limb is often the predominant symptom in Guillain-Barré syndrome, which usually precedes the onset of the ascending symmetrical weakness and hyporeflexia. Clinicians should be aware of this and the myriad clinical features of the syndrome. We recommend close observation and follow up. Although the Guillain-Barré syndrome is a relatively uncommon condition, the consequences of a missed diagnosis and delayed treatment can be catastrophic.

We thank Dr Sue Thompson, consultant paediatrician, West Suffolk Hospital, Bury St Edmunds and Dr Robert Tasker, honorary consultant paediatrician, Dr Joanna Chikwe, senior house officer (surgery), and Dr Chris Allen, clinical dean and consultant neurologist, Addenbrooke's Hospital, for their help, ideas, and suggestions.

Contributors: TT initiated this case report, reviewed the literature, drafted the manuscript, and coordinated discussions and subsequent drafts. CN-J contributed core ideas, edited the paper, and looked after the patient jointly with Dr Sue Thompson on the ward. Both authors act as guarantors for the paper.

Funding: None.

Competing interests: None declared.

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(Accepted 5 April 2000)

Corrections and clarifications

Should treatment recommendations for lipid lowering drugs be based on absolute coronary risk or risk reduction?

A risk reduction formula in this article by S Ramachandran and colleagues (11 March, pp 677-9) contained an error. The final sentence of the second paragraph should read: "The reduction in risk was calculated as the absolute five year risk \times age factor (the age factor = $(0.02357(a)^2 - 3.179(a) + 165.3) \div 100$, where a = the patient's age). The formula is calculated from reference 2."

ABC of colorectal cancer: Innovative treatment for colon cancer

In the box titled "Vaccination with autologous tumour cells" in this article by G A Chung-Faye and D J Kerr (2 December, pp 1397-9) some results from Vermorken et al (1999) were wrongly reported. The second bullet point should read: "In those receiving vaccination, there was a significant reduction in recurrence (44%, 95% confidence interval 7% to 66%) and an increase [not reduction] in overall survival, although this did not reach significance." Also, on page 1397, the graph showing relative risk of colorectal cancer with use of non-steroidal anti-inflammatory drugs somehow lost its vertical line at x = 1 indicating treatment equivalence.