Lyme Disease and Facial Nerve Palsy in Children

A Management Challenge

A Melrose

Lyme disease can affect all ages and occurs commonly between April and October, with June and July the time when tick nymphs are most active. Children in particular can present with early disseminated Lyme disease, weeks after inoculation with *B. burgdorferi,* demonstrating multiple erythema migrans lesions and unilateral, or bilateral palsy, of cranial nerve VII. Health professionals are failing to recognise the significance of a bite, the onset of Lyme disease(LD) and delay in diagnosis and treatment is resulting in preventable ill health.

(LD) is a tick-borne *spirochaetal* infection with a wide range of neurological and non-neurological manifestations. Its clinical diversity and limitations in serological confirmatory tests make neuro-borreliosis difficult to diagnose on initial presentation. LD is transmitted by the deer tick *Ixodes,* which transfers *Borrelia burgdorferi* to the host. Infecting organisms are passed in tick saliva and bites are not always identi-fied by victim or health professional. Unfed ticks are minute and their bites pain and itch free for many hours after infliction.[1] A tick must be attached for about 36 hours for transmission of. B burgdorferi .If the diagnosis is missed and the infection goes untreat-ed for weeks or months, other symptoms begin to appear. These include fever, headaches and neck stiff-ness (from Lyme meningitis), heart palpitations, dizziness and pain moving from joint to joint and facial palsy. Lyme disease is a frequent cause of facial palsy in children living in an endemic region such as eastern USA. An increased tick population in rural areas of Britain and the spread of deer to urban areas have brought increase in number of LD cases in the UK and the potential for more people and children in particular to suffer neurological complications of the disease.

Aetiology

Recent tick bites often go unrecorded and the cause of a presenting peripheral facial palsy (PFP) in a child may not be initially identifiable. Serological testing and empirical use of antibiotics should however be considered, especially when children present during the summer peak LD season. Confirmatory laboratory serology to document B burgdorferi infection takes a considerable time. The clinician treating a child with facial palsy has to decide whether to initiate empirical therapy with antibiotics, steroids, or both, without availability of Lyme disease test results.

LD facial nerve palsy

Acquired unilateral facial weakness is usually due to paralysis of the peripheral facial nerve and is a rela-tively rare presentation to primary care doctors in UK. Its appearance should ring an alarm bell in the mind of the consulted physician. Central causes, although rare in children, should be considered when a patient presents with sparing of the forehead musculature.[1] Historically, the most common cause of acquired PFP in children was otitis media (often without mastoiditis).[2] However, over the last decade, prevalence of Borrelia burgdorferi infections has increased significantly in Europe ,including Britain, and USA.[3] In US endemic areas. Lyme disease has become a frequent cause, in children.[2,4-7] with the prevalence of facial palsy ranging from 50% to 65%.[6]

High-risk clinical predictors

The following, significantly increase the likelihood of Lyme disease if present:

- facial palsy: presenting during peak Lyme disease season,
- no previous herpetic lesions,
- fever,
- headache.

Erythema and painless cellulitis are also markers for later PFN paralysis.

Treatment

Antibiotic therapy is indicated immediately for Lyme infection facial palsy whereas for Bell's palsy, corticosteroid therapy improves time to resolution of symptoms. Early consideration of tick bite and bor-relosis infection ,timely investigation and appropriate treatment is vital if cases are not to be missed or mistreated.

The following case histories exemplify clinical difficulties in diagnosis and differences in management affecting satisfactory outcome.

Case History 1

A 6-year-old British girl whose parents were on a sab-batical exchange, presented to an A & E department in the eastern USA in July with a history of right ear pain, headache and neck stiffness for one day. During the two previous weeks, she had been lethargic c/o headache and neck stiffness. She also had vomited twice. The family doctor had diagnosed otitis externa 10 days prior and treated her with antibiotic eardrops. There was no history of fever, rash, joint pain, muscle weakness or swelling. Her appetite was poor, the main complaint was tiredness and she was inattentive at school. The patient lived in a wooded urban area known by the parents to be frequented by deer. She had no known tick bites but her older sister had been treated for Lyme disease previously.

On physical examination, the child appeared tired but cooperative. The face was asymmetric with right-sided mouth drooping and she could not close the right eye tightly. The nasolabial fold was decreased, and she could not wrinkle her forehead on the right side. There was neck stiffness and tenderness to posterior palpation behind the ear. Kernig's and Brudzinski's signs were negative. Heart was normal, lungs clear. abdomen normal. There was a faint, erythematous rash on right thigh.

Laboratory studies demonstrated a white blood cell count of 10,400/mm3 with 59% neutrophils, 32% lymphocytes and 3% monocytes; haemoglobin 12.1g/dL; haematocrit 37.3%; and platelets 528,000/mm3. Lumbar puncture revealed glucose 48 mg/dL; protein 62 mg/dL; white blood cell count of 322/mm3 with 27% neutrophils, 57% lymphocytes, 11% monocytes, 2% histiocytes and 1% eosinophils; and red blood cell count of 1/mm3. Gram's stain and culture of the cere-brospinal fluid (CSF) were negative after 72 hours. An electrocardiogram revealed normal sinus rhythm with no PR prolongation.

She was diagnosed as early disseminated Lyme disease with facial nerve palsy, meningitis and secondary erythema migrans (EM). and treated with ceftriaxone for 30 days .Symptoms resolved completely within two weeks of initiating antibiotic therapy. 2-tiered test: enzyme-linked immunosorbent assay screening with Western Blot confirmation was under-taken on her arrival. Serum Lyme immunoglobulin M (IgM) was positive at 7.88 (positive at >1.1); serum Lyme IgG was positive at 2.82 (positive at >1.09); CSF Lyme IgM was positive at 1:2 (normal <1:1); and CSF Lyme IgG was <1:4 (normal <1:4)

Case Presentation 2

A healthy 7 year old English boy who had spent a recent holiday camping in the central Scottish Highlands complained of pain round the right ear, painful neck and an itchy skin below the ear 2 days later, he was feeling tired, had a slight temperature, neck stiffness, a fine red rash demarcated at the edges extending from anterior to the ear, down the neck and he was hypersensitive to touch in this area. Parents were advised by a GP in the group medical practice that he had a skin infection and given one week prescription for erythromycin, as he was allergic to penicillin. 14 days later he complained of twitching of right eye and a right facial palsy appeared. A second doctor in the practice diagnosed Bell's Palsy "resulting from the previous infection". Blood investigation revealed increased white cell count and he was pre-scribed a 10 day course of 50mg prednisone. The right sided palsy had resolved by completion of the medication. A day later, he developed further neck pain, headache, vomiting and was very fatigued. Treated as an emergency in the nearest A&E department, he was given paracetamol and ibuprofen and directed to the family GP 30 miles away.

A third GP in the practice examined him, and referred on to the local paediatrician who saw him 6 days later. The parents suggested the possibility of Lyme disease but GPs, and paediatrician dismissed the possibility of environmental risk from ticks. The family home was in a rural area proximal to wood-land. Deer frequented the garden and a parent and sister had previously suffered tick bites. A serological blood test for antibodies was refused as "there was no bite history and ticks were not a problem in the area", although Scottish Highlands and the home location are on record as high risk regions.

One day after the paediatric visit, a left facial nerve palsy occurred. An MRI scan ordered by the paediatrician was reported as normal. No diagnosis was made or differential diagnosis mentioned. The palsy lasted one week to be replaced by diplopia, (third nerve palsy) when he was seen by the fourth GP in the practice and sent to the optician who provided ordi-nary lens spectacles.

One month later at paediatric follow-up, he was fatigued and exhibited diplopia. No diagnosis or differential diagnosis was offered. and he was referred to a neurologist and ophthalmologist. The following day when he complained of leg cramp, joint pain and dizziness, he was seen by the fifth GP. in the practice who resisted taking serology for Lyme disease, which the parents guided by independent professional advice felt ought to be done, as "this had not been ordered by the paediatrician". As a result of parental pressure one day later venous blood was finally sent for serology.

Ten weeks after the initial presentation he was seen by a neurologist , the possibility of Lyme disease was considered and further investigations undertaken including MRI with contrast medium. Weak positive serology for Lyme Disease was reported and a 2 week course of erythromcycin prescribed. The diplopia and paresis improved and was gone in a further month, but there was intermittent swelling of ankle joints and erythema migrans in the interim. At further neurologist follow-up appointment, six months after the initial infection a diagnosis of Lyme Disease was finally confirmed to the parents.

Discussion

The facial nerve may be paralysed anywhere along its full route to the facial muscles. Causes include acoustic neuroma, Ramsay Hunt syndrome, acute and chronic middle ear infection or tumour, temporal bone fracture, and parotitis. The term Bell's palsy by definition, is a diagnosis of exclusion made only after all other possible causes have been considered. Early recognition and treatment of underlying cause may bring speedy recovery.

Lyme disease should be considered in differential diagnosis of lower motor neurone facial palsy An appropriate environmental history and symptom onset in summer are useful discriminating features. Erythema and painless cellulitis particularly in the ear area are markers preceding the facial palsy related to Lyme disease, which may help clinicians to think of Lyme disease first when sepsis and trauma are not present.[1,2] Early localised evidence of the disease is a characteristic EM rash possibly with fever, malaise, headache, neck stiffness and arthralgia. Untreated the infection may pass to early disseminated disease 3 to5 weeks following the bite with arthralgia, meningitis and cranial nerve palsy. Motor neurone facial palsy should be treated as an emergency. Early treatment may be the important factor in achieving rapid resolu-tion.[5,8] However there is still lack of evidence for treatment recommendations in European disease and no UK guidelines are available

Diagnosis requires a high index of suspicion and knowledge of clinical manifestations, seasonal and geographic features.[2] In areas of endemicity, LD should be prominent in the differential diagnoses for children with possible bite history, expanding circular rash, swollen knees, facial palsy, continuing fatigue and declining school performance. 50% tick bites on adults are below the waist, 60% of bites on children are above the waist.[9] Ticks can attach in the hairline and on the scalp of children and remain undetected for longer than on adults. History of tick bite is often absent but presence of an EM rash is sufficient to make a diagnosis of LD, if associated with characteristic clinical findings. In children, indications of neuro-logical involvement include behaviour changes, learning disabilities, school phobia, hypersensitivities of the skin and headaches. Serology is an adjunct with Lyme Igm and IgG detected by enzyme immune assay or immunofluorescent antibody testing.[6] Antibody titres can rise slowly during the localised stage and a negative result is common in early stage. Lyme IgM peaks 3-6 weeks and IgG weeks to months following tick bite. Despite non-specificity, seropositivity for LD children, with neurological signs, usually signifies active neuro borreliosis. Western blot to detect IgM and IgG bodies against specific B burgdorferi proteins is confirmatory and is a highly specific indication of Borrelia exposure.7The absence of antibodies on Elisa testing does not exclude a LD diagnosis.[10,11]

Conclusion

Deer tick bites are now afflicting rural residents in many parts of Britain. In the second case a series of British doctors failed to consider tick bite infection and possible borreliosis and were reluctant to consider serological testing despite presence of characteristic features of LD. The patient also suffered from the lack of management continuity now a salient feature in NHS primary care. These failures delayed diagnosis and treatment. Facial palsies in children are uncommon in general practice, but when accompanied by skin and other manifestations of spirochaetal infection
LD must be considered in differential diagnosis and facial palsy treated as an emergency with early sero-logical investigation. In an endemic area of the Easten USA a child with a similar case history was serologically investigated early and treated expeditiously on clinical grounds before confirmation of diagnosis by blood test

The Scottish Highlands, uplands and Western Islands are badly affected as are forest areas of England. Deer are now appearing in urban environs Tick bites are common and their infection potential is not always recognised by the afflicted and their doc-tors. Spirochaetal infection can result in Lyme disease with neurological sequelae. Health professionals are failing to recognise the significance of a bite, the onset of Lyme disease and delay in diagnosis and treatment is resulting in preventable ill health.

Allan Melrose is a retired GP.

Resources http://www.lymediseaseaction.org.uk/resources/diy/

References:

1 Markby DP. Lyme disease facial palsy: differentiation from Bell's palsy. *Brit Med J*1989;299:605-6.

2 Mark G Murty GE Markby D Lyme disease facial palsy Brit Med J 1989 299 Corresp.14 Oct 3Bryant K Marshall GS Clinical Manifestations of Tick-Borne Infections in Children, Clin Diagn Lab Immunol. 2000 July; 7(4): 523-527.

3 McGarry JV Travel disease and vector ticks.Trav med & Inf Dis. 2011 9.45-9

4 American Academy of Pediatrics. Lyme disease (Borrelia burgdorferi infection). In Red Book Report on the Committee of Infectious Diseases American Academy of Paediatrics; 2003:407-411.

5 Rassbach C, Jantausch J. Reviewing Lyme disease with facial palsy and meningitis Infectious Diseases in Children 2006 Emerging diseases. Feb1-3.

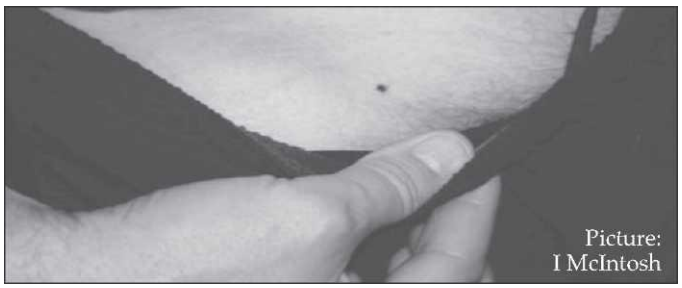
6 Bingham PM, Galetta SL, Athreya B, Sladky J. Neurologic manifestations in children with Lyme disease. Pediatrics. 1995;96:1053-1056.

7 Eppes SC, Nelson DK, Lewis LL, Klein JD. Characterization of Lyme meningitis and comparison with viral meningitis in children. Paediatrics. 1999;103:957-960.

8 Steere AC Sikand VK Presenting manifestations of Lyme disease and treatment outcomes 2003 *New Eng. J Med* 348-2472.

9 Robertson, J N, J S Gray, and P Stewart. 2000. Tick bite and Lyme borreliosis risk at a recreational site in England. *European Journal of Epidemiologypidemiology* 16, no. 7: 647-652.

10 Owen DC Lyme disease -what travellers should know 2008 *Brit. Trav. Health Assoc J*11 3-5.

11 Hoyen D Ticks -Unwelcome travelling companions, 2003 *Brit. Trav. Health Assoc J* 4.13-16.

Tick feeding one day after biting