"Cat scratch disease presenting as meningomyeloradiculopathy"

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Accuracy of clinical diagnosis in Down's syndrome

Hindle and Medakkar showed that the clinical diagnosis of Down's syndrome is inaccurate in one third of cases. We can imagine how stressful it will be for the parents if they have been told that their child may have Down's syndrome and then subsequently karyotype proves to be normal. We conducted a retrospective study to estimate the accuracy of clinically suspicion in our region and in our hospital in particular.

Using the regional cytogenetic laboratory database, all clinically suspected cases of Down's syndrome born in the West Midlands region during the period June 2000 to December 2002 were identified and karyotype results analysed. All babies identified from Birmingham Women's Hospital were studied in detail by reviewing case notes. Of 233 suspected cases from the whole West Midlands region, 148 cases were positive by karyotype. Hence the accuracy of clinical suspicion was 64%. These figures were similar to results from Hindley and Medakkar,1 which showed this was 68% nationally and 64% in the Manchester region. However, from Birmingham Women's Hospital, of 29 cases identified, 25 had a karyotype of trisomy 21 and so a higher accuracy rate of 86%.

We cross checked the patient data from Birmingham Women's Hospital with the rest of the region and found that there were no missed cases from our hospital. Based on the information given to parents before doing the karyotype, in 22 babies where parents were told the diagnosis of Down's syndrome was felt to be certain, karyotype was positive in all 22. However, in seven cases where they were told a positive diagnosis was possible, four had a normal karyotype. All 25 cases that were confirmed positive were seen by a consultant before test. In 23 of 25 babies, clinical suspicion occurred within the first 2 days of life; in two of the babies who were preterm, it took at least 3–4 weeks for clinical suspicion to develop. When we analysed the four negative cases, two were tested without being seen by a consultant. One case was tested just based on profound hypotonia at 31 weeks but no other clinical features. In the final case, karyotyping was done to reassure the parents because there was positive suspicion given by two independent midwives and a registrar, but the consultant felt the baby was normal.

Our data from Birmingham Women's Hospital showed a favourable accuracy rate compared with the previous study.1 This can be explained by the fact that the tertiary hospital may have more experienced neonatologists compared to the broad cohort of junior and senior paediatricians involved in other parts of the region. We believe that assessment by a senior paediatrician before testing may minimise the risk of negative results. There may be difficulty in diagnosing Down's syndrome in preterm babies who may take some time to manifest classic features. We also agree with Hindley and Medakkar that some sort of scoring system like Fried's index may also be useful in improving the accuracy of clinical diagnosis. However, a large prospective study is needed to evaluate those scoring systems.

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References

Beware CSF pressure measured under general anaesthesia

Wraige et al describe three children suffering from idiopathic intracranial hypertension (IIH) in the absence of papilloedema.1 MRI findings in two cases along with an initial symptomatic improvement following lumbar puncture support the diagnosis. In the third case, MRI scan was normal, and the child's headaches did not respond to lumbar puncture or acetazolamide. In all three cases CSF pressure was measured under general anaesthesia with control of position and of carbon dioxide concentration, presumably by end tidal CO2 monitoring. The anaesthetic technique is not reported.

The message that children with IIH may not have papilloedema is a valuable one. However, we would like to add a note of caution regarding the measurement of CSF pressure under general anaesthesia. We have found unexpectedly raised CSF pressure when performing lumbar punctures under sevoflurane anaesthesia, administered to facilitate MRI scanning, in children with a variety of neurological disorders.

All inhalational anaesthetic agents have a cerebral vasodilating action and will increase cerebral blood volume and hence intracranial pressure (ICP). In addition the spontaneously breathing child will sustain an appreciable increase as deep sedation on paediatric wards becomes less acceptable and increasingly demand driven. We report meningomyeloradiculitis as a presenting picture in a child with cat scratch disease (CSD) and identify the need to include this infection in the differential diagnosis of meningomyeloradiculopathies. We also show the likely benefit of antibiotic therapy.

Case report
An 11 year old girl presented three months prior to hospitalisation with low back pain radiating to the lower limbs with paresthesia, limping, and a progressively decreasing strength in the right lower limb. She had sphincter disturbances with frequency of micturition and dysuria. A month later, she developed a stiff spine with impairment in daily activities. Initial examination (day 0; D0) showed decreased strength in the right lower limb with thigh atrophy and reduced sensations. There was scoliosis (dextroconvexity), MRI (fig 1) revealed enlarged conus medullaris, with hyperintensity on T2. Cerebrospinal fluid (CSF; sub-occipital puncture) showed lymphocytic meningitis.
Serologic immunofluorescence revealed a Bartonella henselae (BH) IgG titre of 1/1024; IgM was 1/28. Interrogation revealed cat exposure. Ofloxacin 30 mg/kg/day and rifampicin 20 mg/kg/day were given for six weeks. Recovery in general state and usual activities started a few days after treatment initiation. Improvement of scoliosis followed. All motor, sphincter, and sensory disturbances gradually recovered over one month.

On D60, full spine x ray, T1 and T2 weighted spinal cord MRI, and CSF normalised. BH serology showed a negative IgM, and IGG was at 1/512. At the last assessment (D90), clinical examination was normal.

Comment

CSD is a self limited infection. Neurological involvement is rare; full recovery in these cases has been reported but may take several months. In these neurological forms, antibiotics were suggested to accelerate resolution.

In our patient, combined antibiotic therapy resulted in dramatic improvement, supporting such a therapeutic approach.

The spectrum of infiltrative or space occupying lesions of the central nervous system (CNS) is wide and includes infectious/parasitic conditions (such as mycobacterial infection, angiostrongylosis, and schistosomiasis), inflammatory, vascular, and metabolic diseases (“pseudotumoral” acute disseminated encephalomyelitis, sarcoidosis, Langerhans histiocytois, GM2 gangliosidosisis, and venous infarcts).1 CSD related CNS infective/inflammatory conditions have therefore to be added to this spectrum of infiltrative pathologies and should be carefully excluded before resorting to an invasive technique such as CNS biopsy.

References


Intra-renal reflux

Intra-renal reflux may accompany high-grade vesico-ureteral reflux (VUR) and represents the severe end of the VUR spectrum. In addition, intra-renal reflux is usually seen in very young patients. Presence of intra-renal reflux is a high risk factor for renal scarring, which is an important cause of chronic renal failure and arterial hypertension in children.1 When Angulo et al investigated VUR, they documented intra-renal reflux in 17/89 kidney units in 61 patients with VUR.2

Voiding cystourethrography remains the gold standard for the diagnosis of VUR3 and is one of the best modalities to demonstrate intra-renal reflux, if present. This is often seen as a wedge or fan shaped flush of contrast starting from the calyces outlining the renal papillae, and may extend to the surface of the kidney (see fig 1).

Early recognition of VUR and prompt management favourably influences the prognosis4 and hence all children at risk should be screened.5 In particular, children with intra-renal reflux should be considered for early intervention to stop reflux (either by endoscopic correction or ureteric implantation) and have regular follow up to monitor renal growth and renal function.

Only wholeness leads to clarity

Authors Lee and Mann argue for law compelling use of cycle helmets by children to prevent road deaths and serious injuries.1 This observer is surprised that the peer reviewers allowed publication of material lacking evidence either that the actual risks faced by child cyclists justify compulsion, or that the real world results of helmet compulsion in other countries justify compulsion in this country. These shortcomings are typical of papers in the medical literature that attempt to address the issue of cyclist safety. I believe that these chronic shortcomings are primarily the consequence of the failure of the peer review process.

In the first place, it is irrational that consideration of helmet laws for children is restricted only to cycling, or even begins with cycling. Although, tragically, around 30 child cyclists have been killed on public roads annually in recent years, typically 110 child pedestrians are killed annually.2 Estimates of death risk per kilometre travelled derived from standard data sources3 do not suggest that child cyclists face greater risks than child pedestrians in most age groups. It is in any case evident that the average child is almost four times more likely to become a casualty while walking rather than cycling. The peer reviewers ought to have insisted on a more general discussion of the risks faced by children in transport. This would have placed the injuries to cyclists in context and enabled priority, surely the basis of any systematic approach to public health interventions.

In the second place the evidence for the effectiveness of cycle helmets is split by an interesting contradiction. The authors cite research based on case-control trials reporting that helmeted cyclists were much less prone to serious head injuries than the barenheaded, at least at the time and in the locality of the research work. However, there is also a substantial body of evidence based on population-level studies of head injuries with increasing helmet use. These studies consistently fail to show material benefit for cyclist populations that took up helmet wearing. This was even true in New Zealand, where cyclists responded willingly to helmet promotion, with voluntary use reaching 60% even before the well obeyed law of January 1994 came into force.4 The famous helmet laws for the states of

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