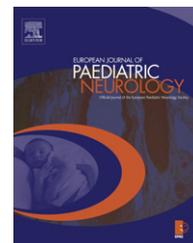




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Original clinical historical contribution

Stuart Green's Vignettes 21, 22 and 23

1. The Girl who wasn't upset on September 11th

A 14-year-old girl on holiday in the States with friends was not enjoying herself as much as her family would have expected. On September 11th 2001 (the day of the terrorist attack on the Twin Towers) she didn't seem "particularly concerned" but carried on her normal activities. The family wondered if she might be "down in the dumps" or depressed but didn't think to make any particular referral. However, when she came back to her parents in the U.K. a few weeks later they noticed a slight change in mood and a general lack of interest in things about her. Over the next few weeks she became more forgetful and occasionally repetitive and this was put down to a possible inappropriate liaison in the States. However, soon afterwards she became weak on one side and showed incontinence.

An MRI scan showed a diffuse asymmetrical leucodystrophic-like picture in the left hemisphere (the radiology suggestive of white matter damage either by metabolic disease or infection or some other process).

The radiological view was that the picture was suggestive of a gliomatosis cerebri, a diffuse slowly growing tumour which infiltrates its way through other cells and doesn't cause problems by pressure but by very slowly damaging the function of the parts of the brain in which it is situated and not uncommonly, although it is a rare condition, starts by affecting the frontal lobes, the so-called quiet areas of the brain (quiet because they don't control movement or speech but control aspects of thought and mood). She had a biopsy which confirmed the condition and then had radiotherapy and made some initial improvement.

When last seen she was coping moderately well but the long term prognosis is unfortunately not at all likely to be good.

2. Never observed it

One of the most distressing metabolic conditions in children is a condition called Lesch–Nyhan syndrome. The condition has progressive abnormal movements (usually starting in the first years of life and getting sadly progressively worse). This only occurs in boys and moreover is associated with abnormal movements, self-destructive biting and aggression

so much so that these children can bite their lips and hands and cause themselves horrendous damage. It is as though they are compelled to do this. Many years ago when I was a young trainee I remember seeing a youngster like this at the National Hospital, Queen Square, London. He was about 7 or 8 at the time and he got progressively worse with years. Drugs seemed to have little effect on his abnormal movements. I can remember very clearly his pathetic figure, a young boy strapped to his wheelchair with violent movements of all four limbs, somewhat modified by the fact that he wore gaiters or tubular supports on his arms to stop him hurting himself. He had bitten his lips so badly that he had to have all his teeth removed. If one removed the tube from his arm his hand immediately went up to his face to hit the side of his head. He seemed perhaps not surprisingly strangely relieved when he was restrained. When we talked to him he appeared to understand although he could hardly speak and could only mouth a few words, but he certainly seemed to understand. He would then certainly lunge forward and jerk you and hiss and spit in a very aggressive way. Afterwards, almost with tears in his eyes his mouth would indicate that he was very sorry. In some way which we don't understand this tragic progressive disease associated with a high level of uric acid in the blood and due to an enzyme deficiency called hypoxanthine guanine phosphoribosyl transferase deficiency uniquely damages certain centres of the brain concerned with posture and emotion. Unfortunately reducing the level of uric acid (which also causes gout) helps the gouty symptoms but doesn't help the brain symptoms. When we asked the great Sir Charles Symons the doyen of British neurology, who at that time was probably about 80 but still making rounds, if he had ever seen this before he said with one of his wonderful quotes "I may have seen it before but I have never noticed it". He did say however he had seen people with episodes of aggression and spitting following the pandemic of encephalitis in 1916 but he didn't know the mechanism. It seemed so tantalisingly close to giving us a clue to an aspect of brain and behaviour where we still unfortunately can't reverse the symptomatology.

Comments

In this Vignette Stuart feels for the child, aches for him, longs for better understanding. He would be encouraged by the immense research efforts underway in his particular

field of metabolic neurology and in Lesch–Nyhan in particular.

3. The Girl who could sing

An 11-year-old girl was admitted to Hospital as an emergency because of a stroke. This was in the days long before MRI scans. A CT scan showed an infarct in the left side of the cortex, compatible with a middle cerebral artery occlusion. The cause was unknown. She had a right hemiparesis and was completely dysphasic. She could neither speak nor understand. However, within a few days she was beginning to understand words although she had great difficulty in speaking. However, after a week she could understand quite well and she could make some noises but no words. However she was able to sing, not only in tune but with the words. She sang the words – I remember it well – to The Wombles of Wimbledon (you can see how old it is!). She sang it perfectly well with good positive and clear words, but when asked to speak the words she could not.

The assumption is that the right side of the brain which controls the expression of music in some way helped here “carry the words” which the left side of the brain was unable to utter.

She went on to make a reasonable recovery.

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