Acute functional deterioration in a child with cerebral palsy

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SUMMARY

We describe a case of acute functional deterioration in a 13-year-old girl with severe spastic diplegia (GMFCS III) and a new diagnosis of diffuse intrinsic pontine glioma (DIPG). She presented with acute deterioration in mobility and motor function over 1 month, which was associated with dysarthria, dysphagia and behavioural change. Her mother had noticed subtle functional deterioration over the 2 months prior to this. Her physiotherapist who was concerned about her acute functional deterioration referred her for emergency review. Neurological imaging revealed a diffuse pontine lesion consistent with DIPG. She was subsequently referred to oncology. She deteriorated further, clinically, over the next few days and following discussion with the team; her family opted for palliative treatment, given the poor prognosis associated with DIPG.

BACKGROUND

Cerebral palsy (CP) is a relatively common condition. It is a static condition and so deterioration of functional status in patients with CP must be promptly investigated to identify the possible aetiology. In our case, the child presented twice to emergency services with worsening headaches and was diagnosed with migraine. She was discharged with analgesia and, on her second presentation, was referred for an outpatient CT scan. It was the patient's physiotherapist who referred her for emergency review following acute deterioration in function and new onset neurology. It is difficult for the physician assessing a new patient with CP to differentiate between an existing neurological deficit and one, which is new in onset. Therefore, in such a situation, it is important to liaise with the patient's primary team.

CASE PRESENTATION

A 13-year-old girl, former premature infant with severe spastic diplegia (GMFCS III), was referred for emergency review by her physiotherapist following acute deterioration in motor function and mobility over 1 month. She had previously been mobilising with a walking aid but was using a wheelchair at presentation. At her previous physiotherapy appointment, it had been noted that there had been some subtle changes in her speech and behaviour. The physiotherapist arranged for review in 4 weeks time to monitor any further changes. The patient was also referred to the clinical psychologist owing to behavioural changes. The psychologist noted behavioural changes secondary to the onset of dysarthria associated with mood swings and progressive difficulty in mobilising made her socially withdrawn. She had two bad falls in the bathroom and

while getting over to the toilet due to increased tone in her left leg. It was felt that home and school environment was not appropriate for this level of functioning and the clinical psychologist and the physiotherapist recommended further medical assessment.

Over the same period, the patient had been complaining of severe, recurrent headaches and had presented to emergency services on two occasions. There were no symptoms suggestive of increased intracranial pressure, and although there was a history of seizures in the past, there had been none in the few months preceding these presentations. She was diagnosed with migraine and sent home with analgesia initially; however, on her second presentation to A+E, outpatient neuroimaging was requested.

On admission, she was alert and oriented with a Glasgow Coma Scale (GCS) of 15. Her pupils were equal in size and reactive to light. She had right-sided strabismus which was longstanding. She had no papilloedema. Significant dysarthria and dysphagia was noted and associated with a loss of gag reflex. Upper limbs revealed power of 5/5 with increased tone bilaterally. Power in the lower limbs was 3/5 bilaterally and tone was significantly increased. She had clonus bilaterally; however, her plantars were downgoing.

She was started on dexamethasone 4 mg four times a day following the diagnosis. Over the next 5 days, there was significant deterioration in the patient's condition. Her GCS dropped from 15 on admission to 13 and her ability to communicate deteriorated.

INVESTIGATIONS

CT and MRI showed a mass lesion in the brain stem. Figure 1 demonstrates diffuse swelling in the pons with no evidence of obstructive hydrocephalus. MRI brain with and without contrast demonstrated diffuse lesion in the pons suggestive of diffuse intrinsic pontine glioma (figures 2–5). Baseline haematological and biochemical investigations revealed no abnormalities.

DIFFERENTIAL DIAGNOSIS

- Brain neoplasm
- Stroke
- Complex migraine

OUTCOME AND FOLLOW-UP

The patient is currently receiving palliative care.

DISCUSSION

Cerebral palsy describes a group of disorders of the development of movement and posture, causing

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Figure 1 CT brain axial view non-contrast demonstrating diffuse swelling in the brain stem.

activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.¹ It is a static condition. Therefore, although often, there is worsening in function due to musculoskeletal complications; there is rarely an acute deterioration in function or new neurological

Figure 2 MRI postcontrast axial imaging with a non-enhancing diffuse brain stem.

Figure 3 MRI T2-weighted non-contrast axial imaging demonstrating the lesion with low signal around the fourth ventricle.

signs. Thus, when there is a change in the neurological status, prompt further investigation is mandatory.

This patient was diagnosed with diffuse intrinsic pontine glioma (DIPG). This is one condition where no pathological confirmation is necessary for the start of radiotherapy treatment.² The prognosis remains poor irrespective of the treatment, with a mean survival of 9–12 months.³ In this case, the

Figure 4 MRI sagittal non-contrast scan demonstrating the lesion with no evidence of obstructive hydrocephalus.

Figure 5 MTI T2-weighted imaging demonstrating signal change extending from midbrain to medulla.

patient was initially offered radiotherapy but following discussion with the team, her family opted for palliative care due to the poor prognosis.

It can be difficult to identify a change in neurological function in a patient with CP, particularly if one is not familiar with the child. Thus, it is of utmost importance to liaise with the patient's existing multidisciplinary team and, if new onset neurological signs are suspected, prompt neurological imaging should be requested.

It is well known that adolescents often exhibit changes in behaviour. It can be difficult to differentiate between a 'normal change' in behaviour and one that is due to a pathological process, particularly in this age group. Behavioural changes should be taken seriously and a detailed history obtained to ascertain the cause.

Interestingly, a paper published in 1999 showed an increased mortality (standardised mortality rate (SMR)=26) from brain cancer in patients with CP despite the fact that there is no increased incidence in this subgroup.⁴ This may be due to the fact that subtle neurological changes may not be recognised in

individuals with CP. The brain tumours noted in this study were ascertained malignant neoplasms, unspecified to any particular site in the brain (International Classification of Diseases 191.9).⁵ The results from this study also state a possible link between CP and brain cancer, but further studies were recommended to establish this. No association with increased mortality has been reported between DIPG and CP so far in the literature including the article cited above.

In summary, a member of their primary team should assess any child with CP presenting with acute functional deterioration and new neurological onset. If there is a change, neuroimaging should be considered as a priority. It is also important to take a detailed history when presented with behavioural change to avoid missing a pathological cause.

Learning points

- ▶ When presented with a child with cerebral palsy (CP) and neurological symptoms, it is important to liaise with the primary team to ascertain whether there has been a change in baseline neurological and motor function.
- ▶ Change in the neurological picture in a patient with CP must prompt appropriate referral to specialist service and investigations must be carried out, including neuroimaging.
- ▶ A comprehensive history should be taken before attributing behavioural changes to 'being a teenager'.

Competing interests None.

Patient consent Obtained.

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