HeadSmart: are you brain tumour aware?

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Abstract
Brain tumours are the commonest cancer cause of death in children. Early diagnosis is crucial in preventing worse neurological outcome in survivors, however diagnosis is difficult as presenting symptoms can be non-specific. In 2006 the median total diagnostic interval was more than three months in the UK, three times more than in the USA. “HeadSmart: Be Brain Tumour Aware” was launched in 2011 to amplify the impact of the RCPCH diagnostic guidelines published in 2008. By providing high quality guidance on assessment, investigation and referral as well as distributing symptom cards, signposting the website and providing e-learning resources, we have successfully enhanced awareness in health professionals and the public. Moreover, since the guidance, there has been a statistically significant reduction in the UK’s total diagnostic interval from 14.4 weeks in 2006 to one of the shortest in the world at 6.7 weeks. This article reviews the progress that has been made and aims to highlight the advice that has proved useful in reducing the total diagnostic interval.

Keywords awareness campaign; brain tumour; HeadSmart; presentation; signs; symptoms

Case story
A mother initially presented with her 3 month old to the GP as she had concerns that he was leaning his head to one side and had a mild flattening on one side of his head. They were reassured that this would correct with time.

The timeline (Figure 1) shows the journey of this patient to diagnosis and sadly, death.

This is a true story. The child was diagnosed more than 3 months after initial presentation and saw many different health professionals. Unfortunately the initial symptoms of a brain tumour can be non-specific; head tilt is one of the symptoms with low awareness amongst health professionals. The many cases like these in the UK are what prompted the initiation of the HeadSmart campaign; the aim, to accelerate brain tumour diagnosis in children.

Introduction
Early diagnosis of all cancers is currently a national priority in the United Kingdom, as set out by NHS England. We know that childhood brain tumours pose a diagnostic challenge for primary and secondary care; the signs and symptoms that precede diagnosis are non-specific, fluctuate in severity and can mimic other common illnesses. The presentation is very much dependant on the age of the child as well as the tumour location and biology. This cases emphasizes time and the acquisition of symptoms over time. It illustrates that the total diagnostic interval (time from symptom onset to diagnosis) is actually a sum of multiple intervals; this family saw many different health professionals and waited for different opinions along the way, which delayed the diagnosis.

In order to achieve earlier diagnosis for childhood brain tumours we need to address all of the above. The strategy required to reduce the time to diagnosis is to provide education, guidance and raise awareness amongst health professionals and the public and this is what prompted the launch of HeadSmart: Be Brain Tumour Aware www.HeadSmart.org.uk.

What is the size of the problem?
Brain tumours affect 1 in 2400 children under the age of 16 in the UK each year; that is 500 new cases each year. Childhood brain tumours occur at any age and account for a quarter of all childhood cancers. They are the second most frequent malignancy in children after leukaemia, and are now the commonest cancer cause of death. Five year survival rates for brain tumours have improved to over 70% but two-thirds of survivors are left with a mild or moderate disability.

The time from symptom onset to diagnosis, known as the total diagnostic interval (TDI), is one of the longest for all childhood cancers. In a study published in 1986, of 247 children with cancer, 80% of those with leukaemia and 84% of the children with Wilms’ tumour were diagnosed within 4 weeks of symptom onset in comparison to 38% of those with a brain tumour. The international median TDI is reported as 8.5 weeks but a multicentre study in the UK between 2004 and 2006 showed a median TDI of 14 weeks.

Along the pathway of TDI, there is symptom progression. In the early stage, due to the rarity of the disease positive predictive values are null and void. In the later stages there is an accumulation of signs and symptoms which eventually become localizing; by this point their outcome is much poorer. A prolonged TDI in childhood brain tumours is associated with an increased risk of life-threatening neurological complications at presentation and a worse cognitive outcome in survivors. It has also been associated with a reduced likelihood of achieving complete tumour resection which is an important prognostic indicator in some tumour types.

What are the diagnostic difficulties?
The reasons for prolonged TDIs are multi-factorial. Firstly, the rarity of brain tumours means health professionals do not
routinely consider this as a diagnosis when presented with non-specific symptoms.

The presenting symptoms of brain tumours include nausea, vomiting, headache and lethargy which are commonly misdiagnosed as gastroenteritis, migraine or behavioural problems. Furthermore, the majority of children who present with a brain tumour have a completely normal neurological examination contrary to the beliefs of many health professionals. Brain imaging

The child was sent home with palliative care input and died just 16 days after the vomiting started and just a week after diagnosis.

Figure 1 Timeline showed the journey of a 3 month old baby to diagnosis.

Figure 2 A symptom card showing symptoms in different age groups.
in children also contributes to the delay; it often requires general anaesthesia or sedation and is mistakenly considered costly.

In order to diagnose effectively we need to have knowledge of the risk of the disease in children and young people, be aware of the signs and symptoms of brain tumours at all ages and have knowledge of the diagnostic process.

There are a number of pre-disposing factors which are associated with an increased risk of childhood brain tumours. These include a personal or family history of a brain tumour, leukaemia, sarcoma or early onset breast cancer, prior therapeutic CNS irradiation, Neurofibromatosis, Tuberous sclerosis and other familial genetic syndromes. When children with these pre-disposing factors present, it should lower the threshold for investigation or referral.

Early diagnosis is a national priority; reducing the time to diagnosis improves outcomes but also reduces cost for the NHS. The difficulty when heightening awareness of brain tumours is achieving the balance between being aware and raising anxiety or over-investigation. Precision in guidance is essential in order to investigate those who warrant it.

How do brain tumours present in children?

Symptoms of brain tumours can be non-specific; they can occur singularly or in combination. Symptoms frequently fluctuate in severity; resolution and then recurrence does not exclude a brain tumour. More importantly, they present differently in different ages. Figure 2 shows the signs and symptoms according to age.
The Diagnosis of Brain Tumours in Children: A Guideline for Healthcare Professionals

HEADACHES:
- Consider a brain tumour in any child presenting with a new, persistent headache
- Headaches that are persistent at any time
- Children aged younger than 4 years may not be able to complain of a headache—observe behaviour.

COMMON HEADACHE PITFALLS:
- Persistent headaches that make a child feel sick
- Persistent headaches that cause an elevation
- Persistent headaches at any time in a child younger than 4 yrs

COMMON HEADACHE PITFALLS:
- Confusion or disorientation and a headache when the headache character changes

* Persistent = continuous or recurrent headache present for more than 4 weeks

NAUSEA AND VOMITING:
- Consider a brain tumour in any child with persistent nausea and/or vomiting
- A child with persistent nausea and/or vomiting requires specialist assessment within 2 weeks

COMMON NAUSEA AND VOMITING PITFALLS:
- Failing to consider a CNS cause for persistent nausea and vomiting

* Persistent = nausea and/or vomiting present for more than 2 weeks

VISUAL SYMPTOMS AND SIGNS:
- Consider a brain tumour in any child presenting with a persisting visual abnormality
- Behavioural change
- Diabetes insipidus

COMMON VISUAL SYMPTOMS AND SIGNS:
- Consider a brain tumour in any child presenting with visual symptoms

* Persistent = visual abnormality present for more than 2 weeks

ASSESS THESE CHILDREN WITH:
- History:
  - Associated symptoms
  - Any predisposing factors

ASSESSMENT PITFALLS:
- The initial symptoms of a brain tumour frequently mimic those that occur with common childhood conditions
- Symptoms frequently, headaches, vomiting, and recurrent seizures do not exclude a brain tumour
- Neurological examination does not exclude a brain tumour
- Language difficulties
- Interpretation services if necessary

Figure 5 A summary of the guideline, reproduced by kind permission of Archives of Disease in Childhood. “The diagnosis of brain tumours in children: a guideline to assist healthcare professionals in the assessment of children who may have a brain tumour.”
Tumour location is another factor as illustrated in Figure 3. The most common symptoms and signs vary according to the where the tumour is; for example, brainstem tumours present most commonly with abnormal gait and cranial nerve palsies whereas posterior fossa tumours most commonly present with nausea/vomiting and headaches.

The symptom profile will also change over time from “at onset” to “at diagnosis” (Figure 4). Visual and motor symptoms increase the most by 53% and 45% respectively from symptom onset to diagnosis. This is important to consider when reassessing patient symptoms over time; headaches may not change much but looking at the progression of visual, motor or behaviour over multiple consultations should alert you to a possible diagnosis of brain tumour.

Awareness of these nuances of presentation will help health professionals, the public and young people recognize the signs and symptoms of brain tumours earlier.

What guidance exists?
A clinical guideline was developed in 2004 by the Children’s Brain Tumour Research Centre at the University of Nottingham. It aimed to reduce the TDI by providing evidence-based guidance for healthcare professionals in primary and secondary care.

It was reviewed and endorsed by the Royal College of Paediatrics and Child Health and received accreditation by NICE Evidence. It is applicable to all children aged 0–18 years and advises on the following:
The symptoms and signs that may occur in children with a brain tumour.
Assessment of children presenting with these symptoms and signs.
Indications and waiting times for imaging children with these signs and symptoms.

Figure 5 shows a summary of the guidance available. Highlighting acceptable time scales for review and referral and using MRI as the modality of choice for diagnosis are some of the key things not addressed in the NICE guideline. These key things will reduce our TDI by reducing some of the sums of intervals which frequently are down to length of wait for a review or scan, or indeed having the wrong type of scan.

“HeadSmart: Be Brain Tumour Aware” was launched in June 2011 to amplify the impact of the RCPCH guideline. The aim was to raise awareness of the symptoms of brain tumours amongst parents/carers, health professionals and young people themselves. Awareness interventions including a symptom checklist card, a website and e-learning packages were developed to encourage self-referral and provide easy access to the guidance for health professionals. The decision support tool provides specific and easy to follow advice for healthcare professionals to “reassure, review or refer” patients which aims to reduce the time to imaging, rather than increase the number of children who need to be imaged (Figure 6).

The overall project plan was for a targeted campaign with education outreach using objective outcome measures in order to assess success.

What was its impact on national performance?

HeadSmart was the world’s first public and professional awareness campaign aimed at speeding up the diagnosis of childhood brain tumours. Since its launch, the campaign has had a dramatic effect on brain tumour diagnosis — taking the UK from having one of the longest TDIs in the world to one of the shortest at just 6.7 weeks (Figure 7). It has clearly changed referral practice shown by a reduction in diagnostic interval (time from initial presentation to healthcare to diagnosis) from a median of 3.3 weeks—1.4 weeks (p = 0.009). This success has led to the campaign being rolled out across Europe following talks in European parliament.

One of the trickiest decisions facing general paediatricians and GPs is whether or not a patient may need to be referred. HeadSmart offers high-quality, evidence-based guidance and easy-to-use tools to support health professionals’ decision making, to help build their confidence and to enable children with a potentially life-threatening illness to be diagnosed as quickly as possible. Prior to the guidance only 32% of paediatricians and 11% of GPs felt confident in diagnosing a brain tumour in children; this increased to 54% of paediatricians and 12% of GPs after the launch of HeadSmart. Overall, 59% of health professionals were aware of HeadSmart; 26% of these were GPs and 73% paediatricians. Between 30 and 37% had seen the quick reference guide, symptom card or leaflet and 19% had seen or visited the website. Between 70 and 91% of these felt the tools were extremely useful to them.

For the public, the awareness tools have offered increased confidence in self-referrals as well as evidence-based materials to assist health professionals in reassuring parents and patients.

What can you do?

The Cancer Taskforce Strategy published by NHS England in 2015 specifies that all cancers should be diagnosed within 4 weeks by 2020 and has highlighted HeadSmart for evaluation. We encourage you to review your own practice, access our awareness tools and use the e-learning resources to improve upon your knowledge so that we can successfully meet this target together.

FURTHER READING

11 The HeadSmart website: www.headsmart.org.uk.

Practice points

• Initial symptoms of brain tumour frequently mimic those that occur with many common childhood conditions
• Symptoms of brain tumours can fluctuate in severity — resolution and recurrence does not exclude a brain tumour
• Presentation of symptoms and signs depend upon the age of the child
• A normal neurological examination does not exclude a brain tumour
• Use the decision support tools to “reassure, review or scan”. (www.headsmart.org.uk)