Neuroimaging to diagnose central nervous system tumours in children

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Abstract

Question Headache, vomiting, lethargy, and seizures are common symptoms in healthy children with benign viral illnesses, but they are also signs that could represent a central nervous system (CNS) tumour. Primary care providers and guardians are hesitant to expose children to radiation associated with computed tomography scans or take on risks associated with the sedation frequently needed for magnetic resonance imaging. When should primary care providers order radiologic head imaging for children with common symptoms to identify those with a CNS tumour?

Answer Central nervous system tumours have no pathognomonic features, which often results in delays in diagnosis. Owing to the high prevalence of infratentorial tumours, children commonly present with symptoms of increased intracranial pressure, making a detailed history and a comprehensive physical examination, including ophthalmoscopy for papilledema, especially important. Magnetic resonance imaging is the criterion standard test but it may take time to access, and young children may need sedation. Hence, computed tomography may be a preferable first option.

The HeadSmart initiative in the United Kingdom provides guidance to obtain brain imaging within 4 weeks of onset of persistent symptoms that are associated with CNS tumours. We advocate applying the same criteria in Canada in order to reduce delay in diagnosis of CNS tumours in children.

La neuro-imagerie pour diagnostiquer des tumeurs au système nerveux central chez l'enfant

Résumé

Question Les céphalées, les vomissements, la léthargie et les convulsions sont des symptômes communs chez des enfants en santé atteints d'une maladie virale bénigne, mais ils sont aussi des signes qui pourraient évoquer une tumeur au système nerveux central (SNC). Les professionnels des soins primaires et les parents hésitent à exposer des enfants au rayonnement associé aux tomodensitométries ou à prendre les risques liés à la sédation souvent nécessaire pour l'imagerie par résonnance magnétique. Dans quelles circonstances les médecins de soins primaires devraient-ils prescrire une imagerie radiologique de la tête chez des enfants présentant des symptômes courants dans le but d'identifier ceux qui sont porteurs d'une tumeur au SNC?

Réponse Les tumeurs au SNC n'ont pas de caractéristiques pathognomoniques, ce qui entraîne souvent des retards dans le diagnostic. En raison de la prévalence élevée des tumeurs infratentorielles, les enfants présentent habituellement des symptômes d'une pression intracrânienne accrue, ce qui accentue davantage l'importance d'une anamnèse détaillée et d'un examen physique exhaustif, y compris une ophtalmoscopie pour détecter un œdème papillaire. L'imagerie par résonnance magnétique est l'examen critère standard, mais il peut falloir du temps avant de pouvoir y accéder, et les enfants peuvent nécessiter une sédation. Par conséquent, une tomodensitométrie peut être une première option à privilégier.

L'initiative HeadSmart, au Royaume-Uni, offre des conseils pour obtenir une imagerie cérébrale dans les 4 semaines suivant l'apparition de symptômes persistants. Nous préconisons l'application des mêmes critères au Canada afin de réduire les retards dans le diagnostic des tumeurs au SNC chez les enfants.

Central nervous system (CNS) tumours are the second most common cancer and the most common solid tumour in childhood. Despite considerable improvement in overall survival for children with CNS tumours,[1] it continues to be a devastating diagnosis for their families and care providers. Early detection and multimodal therapy provided by a specialized multidisciplinary team have led to substantial improvement in the survival and quality of life of children with CNS tumours over the years.[2] However, delay in diagnosis continues to be reported, and misdiagnosis in favour of more common pediatric conditions, such as migraine headaches, viral gastroenteritis, or psychological problems, has been well documented.[3]

Delay in diagnosis

The process leading to diagnosis of CNS tumours usually starts with parents noticing symptoms and seeking medical advice from a primary care physician. Thereafter, the physician may raise the alert if there is concerning medical history or a noticeable deficit upon neurologic physical examination, resulting in referral to a specialist. Following this, imaging is usually undertaken to support a definitive diagnosis of CNS tumour. Shortening these prediagnostic symptomatic intervals ([PSIs] 1, 2, and 3, respectively) is critical to shortening time to diagnosis and treatment.

Factors associated with delay in diagnosis have been documented in the literature and include older age (usually older than 4 years), benign tumours (owing to their slow progression), and location of tumours in the supratentorial region (low-grade glioma, for example).

While diagnostic delay may not impact cure rates,[4] it is likely to affect morbidity[5] and to increase the stress on families.[6]

Tumour symptoms in CNS

Symptoms of CNS tumours are similar to those of other common conditions and include headache, vomiting, lethargy, and seizures, and there are no pathognomonic features, making the diagnosis in children difficult.[3] In a review of 163 children in British Columbia with CNS tumours between 2000 and 2016, the most common symptoms were headache, vomiting, seizures, and ataxia. Forty percent of children presented with hydrocephalus and 17.6% presented with clinically detectable papilledema.[7] Other symptoms that have often been associated with delay in diagnosis include vision changes, weakness, pain, behavioural symptoms, and endocrinopathies.[8,9] A single sign or symptom, unless it is a prominent focal abnormality on neurologic examination, is uncommon at the time of diagnosis.[10] Psychological changes have also been noted to be frequent among children with CNS tumours. Some physical symptoms may be considered to be psychosomatic in origin.[11]

However, most children with a CNS tumour will have notable signs on a full physical examination.[3] In an analysis of 3276 pediatric patients, less than 3% of those with headache and a brain tumour had no findings of abnormality on careful neurologic examination,[12] and among 72 children with CNS tumours whose symptoms started as headache, abnormalities on physical examination had developed in 85% within 2 months of headache onset.[13] Thus, a comprehensive physical examination, including ophthalmoscopy, would help care providers identify those children who require prompt imaging studies.[3]

When to image

In recent years, Canada has seen better access to advanced imaging, including computed tomography (CT) and magnetic resonance imaging (MRI). However, risks from exposure to radiation from CT and the need for sedation in young children for MRI, with its own inherent risks, necessitates judicious use of each of those tests. The imaging chosen as the first step will likely depend on patient- and health system–related factors. Obtaining a CT scan of the brain as the first step may be faster in most centres in Canada.

The United Kingdom's National Collaborating Centre for Primary Care developed referral guidelines for suspected cancer, including specific guidance for children and young people.[14] Based on its systematic review of the literature and Delphi consensus process,[15] it concluded that MRI is the modality of choice for making a diagnosis, when possible, and recommended that patients selected for nonemergency imaging have that imaging performed within 2 weeks of presentation, with results given to the family within 1 week afterward.

Preventing delays

After finding considerable delays in CNS tumour diagnosis in a national survey in the United Kingdom,[8] the Royal College of Paediatrics and Child Health launched the HeadSmart initiative in 2011[16,17]—a practitioner-targeted awareness quality improvement strategy, followed by a public awareness campaign. This initiative used a systematic review and meta-analysis to identify age-related differences in symptoms. Awareness materials with an age-stratified symptom checklist and algorithms for urgent referrals were distributed to health care professionals, health organizations, professional bodies, and the public (schools, child care facilities, hospital waiting rooms, local authorities, charities, and commercial networks). The result was a meaningful decrease in median PSI from 14 to 6.7 weeks.[18] The authors recommend obtaining brain imaging within 4 weeks if a child presents with symptoms and signs that may be due to a CNS tumour. We advocate applying the same criteria in Canada in order to reduce PSI.

In a survey of practising Canadian family physicians,[19] participants described barriers to diagnosis in Canada, including long wait times for consultation and imaging investigations, the geography of Canada (distance to tertiary care centres), access to resources (specialist information and current guidelines on investigations and management), lack of experience given the rarity of diagnosis, patient-related factors (young age, need for sedation), and health system barriers. More than 80% of those surveyed were not aware of any available resources or guidelines to help manage a patient or navigate a referral, and dissemination of an online resource like HeadSmart may provide helpful general educational materials for both care providers and patients.

Conclusion

Central nervous system tumours in children pose a diagnostic challenge with considerable variability in time from symptom onset to diagnosis. Most children with

a CNS tumour will have notable signs on a full physical examination within months of presentation and this should prompt consideration of head imaging including CT, MRI, or both within 4 weeks, as well as subspecialty referral. Although time to diagnosis may not necessarily affect survival, the perceived delay can be devastating for both families and care providers.

Competing interests
None declared

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