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The Presenting Feature and Role of General Practitioners and Non-neurosurgeon Physicians in Recognizing Pediatric Brain Tumors

[Pediyatrik Beyin Tümörü Tanısının Konmasında Pratisyen ve Beyin Cerrahı Olmayan Hekimlerin Rol ve Özellikleri]

SUMMARY

AIM: To determine the presenting features and role of non-neurosurgeon physician in recognizing the signs and symptoms of brain tumours in children.

METHOD: Medical records of 31 pediatric brain tumor patients treated in Department of Neurosurgery, Soetomo General Hospital, Airlangga University Faculty of Medicine, Surabaya from August 2005 to September 2006 were reviewed.

RESULTS: Thirty five percents f parents went to pediatrician as their first contact physician, 25% to general practitioner, 20% neurologist, 20% to neurosurgeon. Neurosurgeon was the second and third contact physician receiving refferal from non-neurosurgeon physician. The most common symptoms were headache (71%), vomiting (61%), motor weakness (48%), visual disturbance (45%), decrease level of consciousness (45%) and seizures (38%), unsteadiness (35%). The most common symptoms that led the parents to find medical help at any time were motor disturbance (48%), vomiting (48%), visual disturbance (45%), unsteadiness (35%), decrease level of consciousness (32%), seizures (32%), headache (32%). All patients had neurological signs at diagnosis; 58% had papilloedema, 48% cranial nerve abnormalities, 35% cerebellar signs, 32% motor disturbance, 29% a reduced level of consciousness, 12% cranial enlargement. Duration of symptoms at admission was 1 months (32%), 2 months (42%), 3-6 months (19%), more than 6 months (7%). A short symptom interval was significantly associated with high grade tumours and patient age 3 years or younger.

CONCLUSION: The symptoms and signs are often nonspecific, mimicking more common diseases. Therefore, the possibility of a brain neoplasm should always be considered, it materializes very rarely. Benign neurologic symptoms such as headache, which last for 2 months or more, should indicate the need of additional studies. Our results higlighted the neurologic impairments which might facilitate early recognition of a brain neoplasm. Neurologic problems as the only symptom of brain tumor may mislead pediatricians and neuropediatricians, so that a low index of suspicion may delay the diagnosis.

ÖZET

AMAÇ: Çalışmanın amacı; beyin cerrahı olmayan doktorların çocukların beyin tümörü belirti ve semptomlarının tanımadaki rollerinin ve çocukların doktora götürülme şikâyetlerinin araştırılmasıdır.

METOD: Surabaya'da bulunan Airlangga Üniversitesi Tıp Fakültesi Soetomo Hastanesinde Ağustos 2005, Eylül 2006 arasında yatan 31 beyin tümörü hastasının kayıtları incelenmiştir.

BULGULAR: Hastaların ilk başvurduğu doktor %35 pediatri uzmanları, %25 pratisyen hekimler, %20 nöroloji uzmanları, %20 beyin cerrahi uzmanları olmuştur. Beyin cerrahisi uzmanı, diğer doktorların tavsiyesi ile ikinci veya üçüncü ziyaret edilen doktor durumundadır. En sık karşılalılan semptolar; %71 başağrısı, %61 bulantı, %48 motor zayıflama, %45 görme bozukluğu, %45 bilinç bulanıklığı, %38 nöbet geçirme, %35 denge bozukluğudur. Ebeveyni en sık doktora başvurmaya sevkeden %48 motor güçsüzlük, %48 bulantı, %45 görme bozukluğu, %35 denge bozukluğu, %32 bilinç bulanuıklığı, %32 nöbety geçirme, %32 başağrısıdır. Tüm hatalar teşhis sırasında nörolojik belirtilere sahiptiler; %58 papilloadenoma, %48 kraniyal sinir anomalileri, %35 beyinciğe ait semptomlar, %32 motor zayıflama, %29 azalmış bilinç seviyesi, %12 kraniyal genişleme görülmüştür. Başvuru sırasında hastaların %32'sinin bir aydır, %42'sinin iki aydır, %19'unun üç-altı aydır, %7'sinin altı aydan uzun zamandır yakınmalarının eevam ettiği tespit edilmiştir.

SONUÇ: Bu hastalarda belirti ve bulgular genelde, pekçok hastalıkta görülebilen nonspesifik belirti ve bulgulardır. Bu açıdan beyin tümörlerini sık rastlanmasa da her zaman akılda bulundurmak gerekmektedir. Başağrısı gibi masum görünen nörolojik semptomlar iki aydan uzun süreden beri devam ediyorlarsa ilave incelemeler yapılmalıdır. Araştırmamızdan elde edilen bulgular, nörolojik bozukluk durumlarının beyin tümörlerinin erken belirtisi olabileceğini göstermektedir. Nörolojik belirtiler beyin tümörlerinde pediatristlerin ve çocuk nöroloğlaronın da gözden kaçırdığı, bu yüzden de teşhisin geçiktiği durumlardır.

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Key Words: Pediatric Brain Tumor, Neurologic Symptoms, Pediatric Neurosurgery, Child Brain Tumor, Non-Neurosurgeon Physician.

Anahtar Kelimeler: Pediatrik Beyin Tümörleri, Nörolojik Semptomlar, Çocuk Beyin Cerrahisi, Cocuk Beyin Tümörleri, Beyin Cerrahı Olmayan Doktor.

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INTRODUCTION

Brain tumors are the second most common solid malignancy of childhood, immediately following leukemia (1,2). The annual incidence of intracranial neoplasms in children is 1 to 3 per 100.000 (1,3). The increasing annual incidence over 20 years in US (1973–1994) is associated with expanding diagnostic measures especially MRI utilization for the diagnosis of CNS conditions in children. Reports describing the

ability of MRI to detect tumors not detectable by CT imaging support this explanation (4).

Despite the availability of sophisticated neurodiagnostic tools, the utilization of those tools depends on the physician to recognize the clinical symptom and choosing the appropriate tools. The difficulties in diagnosis the brain tumor in young children include: a) young children have a limited ability to describe symptoms characteristic of brain tumors, such as blurred vision or headaches; b) some symptoms of brain tumors, such as vomiting or irritability,can occur in a variety of more common childhood illnesses; c) other symptoms, such as behavioral changes, may be very subtle and develop over a long period of time; d) it can be challenging to perform a thorough neurological assessment and diagnostic tests such as CT or MRI scans on children who are frightened, not fully cooperative, or hard to sedate for procedures (5). Therefore, the possibility of a brain neoplasm should always be borne in mind, even if it materializes only rarely.

The symptoms and signs differ with the location of the tumor and, to a certain extent, its histologic nature, these two factors being related. It is also essential to realize that the clinical significance of pathologic grading may be different from its biological value. A benign tumor that is strategically located so as to be impossible to remove without interfering with the essential neural function is malignant for the patient even if histologically benign (6).

Brain tumors also may produce mass effect and bleed, increasing the risk of brain herniation. Early detection may improve patient outcome because late diagnosis of a brain tumor may affect brain tumor resectability, neoplasm stage, and risk of cerebral herniation (6).

With advances in treatment of these tumors over the past 20 years, improvement in survival may not correspond with how well the family's needs are being met. Symptoms associated with aggressive treatment, sequelae of CNS tumors, as well as the diagnosis itself can have a devastating impact on the child and family (7). Early detection of brain tumor plays a very important role as it may have a significant bearing on clinical outcome (8).

MATERIAL AND METHODS

We reviewed medical records of 31 pediatric patients harbouring brain tumor treated in Department of Neurosurgery, Soetomo General Hospital, Surabaya within period of August 2005 to September 2006. The review includes: the age at onset of symptoms, their first contact to physician, the refferer physician, the presenting features, the symptoms that led the parents find their physician, clinical examination, radiologic examination, initial diagnosis, treatment, and clinical outcome. Data that were not recorded well was taken by re-interview the parents. The data that usually missed in medical records include: the first, second, or third attended physician (when available). The symptoms that led the parents find their physician.

Radiologic examination include CT scan and MRI. All patients underwent CT scan examination, but only 14 patients underwent MRI. The reasons why the MRI was not conducted was their clinical condition or died before the imaging.

Histopathology results were available only for the excised or biopsied tumor.

RESULTS

Data of 31 patients consists of 17 boys and 14 girls were analyzed. The boys to girls ratio was 1.2:1 (Table 1). The mean age was 7.1 yers old (range 18 months to 15 years old). According to specific developmental characteristics, patients were divided into age groups as follows: 0- 2 years, 2-5 years, 5-10 years, and 10-16 years, with the most prevalence in our series was between 5-10 years old (table 1). Careful history was taken, and detailed physical and neurologic examinations were performed.

Table 1. Patient characteristics according to age and sex.

Sex	**n**
Male	17 (55%)
Female	14 (45%)
Age	
0–2 years old	3 (9,6%)
2–5 years old	7 (22%)
5–10 years old	16 (53.4%)
10–16 years old	5 (16%)

The diagnosis was confirmed by brain CT scan with and without iodine contrast injection.

The duration of symptoms before the diagnosis of brain tumor was analyzed. The symptoms were classified into 8 categories: headache, vomiting, visual disturbance (papilloedema, blurred vision, diplopia), motor or limb weakness, unsteadiness or ataxia, seizures, decreased level of consciousness, and others (speech disturbance, enlargement of head circumference, and hearing loss).

Table 2. The most common symptoms found in children with brain tumor

Symptoms	Supratentorial	Infratentorial	Total
Headache	9 (29%)	13 (42%)	71%
Vomiting	7 (22.5%)	13 (41.5%)	64%
Motor weakness	6 (19%)	9 (29%)	48%
Visual disturbance	7 (22.5%)	7 (22.5%)	45%
Decreased level of consciousness	9 (29%)	5 (16%)	45%
Seizure	6 (19%)	6 (19%)	38%
Unsteadiness	2 (6%)	9 (29%)	35%
Other			
Speech disturbance	3 (9.6%)	0 (0%)	9.6%
Enlargement of Head circumference	0 (0%)	4 (12%)	12%
Hearing loss	1 (3.2%)	0 (0%)	3.2%

Table 3. Neurological signs at admission

Sign	Percentage
Papiloedema	58%
Cranial nerve abnormalities	48%
Cerebellar sign	35%
Motor disturbance	32%
Decreased level of consciuosness	29%
Cranial enlargement	12%

Table 4. Symptoms that led the parents to physician

Symptom	Percentage
Motor disturbance	48%
Vomiting	48%
Visual disturbance	45%
Unsteadiness	35%
Decreased level of consciousness	32%
Seizures	32%
Headache	32%

Patients with multiple symptoms were classified in each of the respective categories. Methods of descriptive statistics were used for data processing. Headache was the most common symptom in 71% of study children for both supra- (29%) or infatentorial (42%) tumor (Table 2). It showed considerable qualitative variation, from the pattern of early morning headaches associated with vomiting to the type that may be relieved by usual analgesics. Vomiting was present in 64% of children, alone or associated with headache. Unsteadiness or ataxia was present in 35% of children and motor weakness was observed in 48% of children. Visual disturbances were recorded in 45% of children.

Deacreased level of consciousness occurred in 45%, and convulsions in 38% of children. Presenting symptoms before making the diagnosis are shown in Table 2.

The associated signs at presentation were papiloedema (58%), cranial nerve abnormalities (48%), cerebellar sign (35%), motor disturbance (32%), decreased level of consciousness (29%), and cranial circumference enlargement (12%) (Table 3) .

Motor disturbance observed by parents or self-reported and vomiting is the most common symptoms that made the parents brought their children to the physician. Another symptoms were visual disturbance (45%), unsteadiness (35%), decreased level of consciousness (32%), and seizure (32%) (Table 4).

Headache, known as the most common symptom at presentation, was the most ignored symptom by parents and only couriously examined when it persisted for more than 1-2 months or accompanied by other specific symptoms.

Duration of symptoms from the first onset of symptoms until the parents in contact with physician was variable. Forty two percents of parents brought their children that experienced “common symptoms”(headache and vomiting) to physician in 1 month to 2 months (Table 5).

Table 5. Duration of symptoms before the children presented to the physician

Duration	Common symptoms	Specific neurologic symptom
<1 months	32%	80.8%
1–2 months	42%	12.8%
3–6 months	19%	-
>6 months	7%	6.4%

Table 6. List of first, second and third attended physicians

Contact	GP	Pediatrician	Ophtalmol	Neurologist	Neurosurg	Other
1st	25%	35%	-	20%	20%	-
2nd	-	3%	6%	6%	61%	3%
3rd	-	-	-	-	19%	-

GP: General Practitioner, Opthalmol: ophtalmologist, Neurosurg: neurosurgeon

But the children presented the specific neurologic symptoms (motor disturbance, visual disturbance, seizures, decreased level of consciousness, unsteadiness, and other neurologic symptoms) were brought to physician in less than one month (88%) (Table 5).

Neurosurgeons were the second attended physician in 61% of cases after the parents look for help on non-neurosurgeon physician and the third in 19% of cases. Pediatricians were the most common first attended physicians (35%) (Table 6). Another first attended physicians were general practitioner in 25% of children, neurologist 20%, and neurosurgeon itself in 20% of cases. Ophtalmologists were also the second attended physician in 6% of cases (Table 6).

In one case the parents brought their child to the pediatric surgeon due to GI tract disturbance and diagnosed as obstructive ileus. The child had undergone laparatomy, and none was found in the abdominal cavity. The pediatrician made an MRI of the head due to intractable vomiting and ataxia. The MRI showed a cystic mass in cerebellum suspected as the pilocystic astrocytoma causing hydrocephalus. Surgery was done to excised the tumor and planted a VP shunt. The histopathologic result was in accordance with the radiologic interpretation that led to a final diagnosis of Juvenile type Pilocystic Astrocytoma of the cerebellum.

Infratentorial lesion (68%) were more oftenly encountered than the supratentorial (35%) in children. One patients had both infra- and supratentorial lesion. The infratentorial lesions (58%) are also more commonly associated with hydrocephalus than supratentorial lesions (19%). All of the intraventricular infratententorial lesions were associated with hydrocephalus.

Almost all patients (94% of cases) received surgical intervention, emergency or elective surgical procedures. Emergency procedures inlcuded emergency VP shunting in 5 patients (16%) and external ventricular drainage (EVD) in 1 patients . Removal of the tumor was done in 7 cases (7%) and elective shunting procedures with planned excision (2-step-surgery) were done in 14 patients (45%). Two untreated cases were associated with the sudden decreased of level of consciousness down to coma right after the diagnosis was made.

The results of the treatment were not always satisfied. Only 10 (32%) cases that showed improvements for their clinical signs and symptoms. The rests of cases showed no improvements (39%), worsened (13%), or died (16%).

DISCUSSION

Intracranial tumors presenting in the first years of life are uncommon in general pediatric practice. Many of the presenting symptoms are also common to other frequent childhood illnesses. The diagnosis may be especially difficult in the first two years of life. We tried to identify the reasons for the delay in reaching the diagnosis of brain tumor in children, and to propose a way to reduce the time elapsed from the onset of symptoms and signs to the diagnosis. Our results showed a high incidence of headache (47%) as a presenting symptom of brain tumor in children.

The value of neuroimaging in evaluating headache in children is controversial due to low overall incidence of brain tumors among children with headache (9,10). Clinicians may be trapped between ordering imaging studies with some inherent risks (sedation, contrast use, and false-positive rate) and early detection of intracranial tumors. One study suggests dividing children with headache into high-, intermediate- and low risk groups according to clinical predictors of surgical space-occupying lesions. This study suggests that neuroimaging should be reserved for high-risk patients (2).

The low-risk group with baseline pretest probability of 0.01% (1/10 000) consisted of patients with the sole finding of chronic headaches of >6 months duration (11). The most effective strategy was no imaging with close clinical follow-up. The intermediate-risk group with baseline pretest probability of 0.4% (4/1000) consisted of children with migraine headaches and a normal neurologic examination(10,11). CT followed by MRI when positively showed suspected of brain tumor was obtained by CT scan. was a slightly more effective diagnostic strategy than MRI alone. The high-risk

group with baseline prevalence (prior probability) of 4% consisted of children with headache of <6 months duration and at least 1 other clinical predictor suggestive of a surgical brain tumor such as an abnormal neurologic examination. An increase in the number of predictors has been highly correlated with an increased risk of brain tumor (2).

Besides headache signs of intracranial hypertension are vomiting, papilloedema. Headache may be severe and relieved by vomiting.

Only 58% of our patients had the classical triad of headache, vomiting and papilloedema in our series. In very young children, papilloedema is often absent, but an infant may show a behavioral change. Our results are consistent with those of other authors (12,13).

The role of general practitioner, pediatrician, neurologist, and other non-neurosurgeon specialists are very important to recognize the signs and symptoms that in many cases are frequently overlooked. Symptoms such as headache and vomiting, which was present in 58% of the children with brain tumors in our series, are easily attributable to more common illnesses encountered in daily pediatric practice. The physicians in primary care unit have to be equipped with capabilities to list differensial diagnosis of the common symptoms. For example, vomiting due to increased intracranial pressure is usually unremarkable except for its repetition and frequent morning reccurrence.

Since the distribution of neurosurgeon has not meet the optimal ratio in some areas of our country, the capability of general practitioner and other specialists to recognize the common symptoms and signs are unequivocally needed especially in the setting of rural or remote area where neurosurgeons are far from reach. This capability will lead to prompt diagnosis and early treatment which might improve outcome and decrease morbidity and mortality rate.

Although the advance of neuroimaging can increase the detection rate, the use of sophisticated neuroimaging, until now, only preserved for the child who experienced not only “common symptoms” but also “neurological symptoms”. Only 32% of primary brain tumors were diagnosed using neuroimaging modalities within the first month after the onset of the common symptoms. The reason for not to apply neuroimaging modality in the first place is mostly due to financial and cost-benefit considerations.

In contrast, 84% of Wilms' tumors and 80% of cases of acute leukemia were diagnosed within one month of the onset of symptoms (8). The delay might be caused by: a) the physician that put the possibilities of brain tumor in the last row of their differential diagnosis when facing the children with “common symptoms”; b) the parents that didn’t recognize early sign of intracranial pathology that often subtle, and presented as common symptoms; c) The children itself that might not be able to express their complains well enough. In contrast, the specific neurological signs and symptoms will ring the bell of the physicians and parents for the possibilities of brain tumor, which made 80.8% of parents brought their children to the physician. A careful history including questions about visual symptoms and educational or behavioural difficulties, as well as the more widely recognized symptoms of raised intracranial pressure or motor dysfunction, and a thorough examination including visual assessment and appropriate plotting of growth and head size are important in the diagnosis of brain tumours (14).

CONCLUSION

Intracranial tumors present a diagnostic challenge in young children. The symptoms and signs are often nonspecific, mimicking more common diseases. Therefore, the possibility of a brain neoplasm should always be considered, even if it materializes very rarely. Benign neurologic symptoms such as headache, which last for 2 months or more, should indicate the need of additional studies. Our results higlighted the neurologic impairments which might facilitate early recognition of a brain neoplasm. Neurologic problems as the only symptom of brain tumor may mislead pediatricians and neuropediatricians, so that a low index of suspicion may delay the diagnosis. A careful history taking including questions about visual symptoms and educational or behavioural difficulties are absolutely required in investigating the possibilities of early signs or symptoms of brain tumor. An unequivocal role of general practitioner and non-neurosurgeon specialist, to whom most of the parents came to, are very improtant. They should be equipped with capabilities to recognize even the subtle “common symptoms” which might come with brain tumor in children.

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