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ORIGINAL PAPER

Posterior fossa tumors in children: how long does it take to establish the diagnosis?

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Abstract
Objective The aim of this study is to evaluate, for our patient population, the time interval from the first chart-documented symptom to the radiological diagnosis in children and infants with posterior fossa tumors.
Materials and methods We retrospectively analyzed 50 consecutive children (36 men, 14 women) with posterior fossa tumor treated at our department between January 1999 and December 2003. The mean age at time of diagnosis was 98 months (6 months–16 years). The mean follow up was 27 months (6–61 months). The diagnoses included astrocytoma (n=17), medulloblastoma (n=15), ependymoma (n=6), and other tumors (n=12).
Results The mean time interval between onset of symptoms and radiographic diagnosis was 142 days (5–535 days), the median was 59 days. The mean time for Grade I and II tumors was 238 days (n=19) and for tumors Grade III and IV 117 days (n=31). The most common presenting symptoms were headache, nausea, vomiting, ataxia, and oculomotor deficits. Approximately half of the patients were initially diagnosed and treated for other diseases (gastrointestinal infection, appendicitis, psychological behavioral problems, cervical spine strains, different ophthalmologic entities). Specialists (ophthalmologists, orthopedics) tended to diagnose and treat their specific diagnoses (e.g., strabism, torticollis). Parents play a significant role in the process of establishing the correct diagnosis.
Conclusion We conclude that further information and education regarding symptomatology and diagnosis of posterior fossa tumors in children is necessary. Communication has to be improved between parents and referring physicians of all specialties and neurosurgeons.

Keywords Posterior fossa tumor · Patient management · Diagnostic delay · Diagnostic interval · Pediatric brain tumor

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Introduction

Intracranial tumors are the most common solid neoplasm in children occurring in 2.4 to 4 per 100,000 [1–5]. Posterior fossa tumors present approximately 60–70% of all brain tumors in children. Clinical symptoms include headaches, nausea and vomiting, reduced level of consciousness, ataxia, or cranial nerve deficits. Pediatricians and general practitioners have significant responsibility in the process of identification of clinical signs that could point toward a posterior fossa tumor. Among our patients, there were several with long lasting symptoms before the diagnosis was confirmed. Therefore, we wanted to evaluate the diagnostic process in our patients retrospectively.

Issues that we wanted to present and discuss in this paper are: How long does it take in a highly developed medical system to establish the diagnosis? What are the main presenting symptoms in our patient population? What are the most misleading symptoms? What are the main referring specialties? What are the differences in regard of diagnostic workup and referral pattern among the specialties? What are the differences regarding the diagnostic

interval between low-grade and high-grade tumors? What is the parent's role?

Materials and methods

We performed a retrospective chart review and included 50 children with posterior fossa tumors treated in our department between January 1999 and December 2003.

Patients' data are summarized in Tables 1 and 2.

Tumor localization was cerebellar hemispheres (n=23); fourth ventricle (n=11); midbrain, pons or medulla oblongata (n=12), and cerebellopontine angle (n=4).

We evaluated the patient charts for the following information: first chart-documented symptom (including outpatient charts and letters from referring physicians), time interval from first symptom to established radiological diagnosis, presenting symptoms, specialties involved in diagnostic evaluation, specialty referring the patient to our department, misleading diagnoses and treatment plans before established diagnosis of a posterior fossa tumor, and histological diagnosis.

Results

Our study group is of similar epidemiological consistency as comparable groups in the literature: a mean age of 98 months, a dominance of male patients with a ratio of 36:14 to female patients, and the histopathological differentiation is similar to previously studied patient collectives [3, 4].

The mean time interval from the first chart-documented symptom to the day of radiological diagnosis was 142 days, the median was 59 days.

The mean time interval for tumors Grade I and II was 238 days (n=19 patients) and for tumors Grade III and IV 117 days (n=31 patients).

Seventeen patients were diagnosed within the first month; seven patients were diagnosed within 2 months, six patients within 3 months, 13 patients within 4–9 months, and in 7 patients, diagnosis took longer than 9 months (Fig. 1).

Most frequent symptoms were headache, nausea and vomiting, reduced level of consciousness, ataxia, cranial nerve deficits, and torticollis. In 78% of the patients (39/50), hydrocephalus was diagnosed.

Before being transferred to our department, the patients were seen, on average, by two physicians of different specialties. The children were mainly transferred to our department from pediatric hospitals, pediatricians in practice, general practitioners, neurologists, ophthalmologists, and orthopedic surgeons (Table 3).

Table 1 Epidemiological data concerning our patient group of pediatric patients with posterior fossa tumors seen in our hospital (1999–2003)

Patient's data	
Number of patients	50
Gender (*male/female*)	36/14
Mean age (*months*)	98 (6 months–16 years)
Mean follow-up (*months*)	27 (6–61 months)

Table 2 Histopathology of the posterior fossa tumors in pediatric patients seen in our hospital (1999–2003)

Pathology	Number of patients
Astrocytoma	17
Medulloblastoma	15
Ependymoma	6
Other	12
	50

Case examples

Case 1

A 6-year-old girl developed ataxia shortly after starting her first year in school. The school physician diagnosed a "developmental delay" and prescribed physical therapy. Symptoms improved, and no further workup was done at this point.

Two months later, ataxia got worse and was interpreted as psychological in nature, caused by the school environment and stress. The following 3 weeks, dysarthria developed. No further workup was done. The patient was

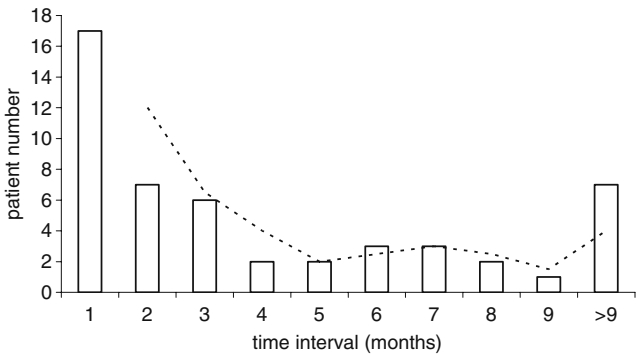

Fig. 1 Number of patients diagnosed within a certain time period (months) after onset of symptoms

Table 3 Specialties being involved in the diagnostic regimen before the admission to our hospital with the diagnosis of a posterior fossa tumor

Referring specialty	Number of patients
Pediatrics (hospital)	23
Pediatrics (praxis)	9
ENT	2
General practitioner	2
Ophthalmologist	3
Neurologist	4
Orthopedics	2
Others	5
	50

admitted by the parents through the pediatric emergency room with nausea, vomiting, and lethargy. Magnetic resonance imaging (MRI) revealed a tumor of the pons. The diagnostic time interval was 3 months and 3 weeks. Histology revealed an anaplastic astrocytoma (WHO III).

Case 2

The mother of a 5-month-old girl noticed a torsion and tilting of the head to the right. The pediatrician prescribed chiropractic therapy. The following weeks, symptoms improved. The torticollis reappeared 7 months after the first symptoms. The parents presented the girl to the pediatric emergency room after the baby started vomiting and appeared less conscious. MRI revealed a tumor of the fourth ventricle with infiltration of the brain stem and the cerebellopontine angle on the left side. The diagnostic time interval was 8 months and 10 days. Histology was ependymoma.

Discussion

How long does it take?

In our patient series, it took 142 days, on average, to establish the diagnosis of a posterior fossa tumor.

What are the possible explanations?

Despite the fact that posterior fossa tumors are among the most significant solid tumors in childhood age, they are still relatively rare and tend to be misdiagnosed for more common differential diagnoses. Patients in our study have been diagnosed and treated for gastrointestinal infection (n=9), appendicitis (n=1), psychological behavioral problems (n=4), cervical spine strains (n=9), and different ophthalmologic entities (n=5).

The awareness of childhood brain tumors within the population is low; the knowledge among physicians is limited. The presentation is often unspecific, especially in infants and younger children [6]. Posterior fossa tumors do not cause seizures as occasionally seen with supratentorial tumors.

What are the most misinterpreted symptoms?

Torticollis has a wide range of differential diagnoses: spasmodic torticollis, hemorrhage into the sternocleidomastoid muscle with subsequent contracture, cervical infection/adenitis, syringomyelia, and cerebellar tumor [7–9]. Torticollis may also present as a secondary correction to reduce diplopia.

A number of children (n=8) already had a torticollis for some time until signs of intracranial pressure occurred and finally the correct diagnosis was made. Retrospectively, they often had mild neurological deficits from the beginning but were missed by the pediatricians and orthopedics probably because of their focused view on orthopedic causes of the symptom.

Diplopia and other oculomotor abnormalities were not always thoroughly evaluated by obtaining an imaging study of the brain. Some patients were treated for strabism conservatively for several weeks.

Who refers early and who refers late?

Most patients were seen and referred by pediatricians and neuropediatricians either in a hospital setting or in private praxis and by general practitioners.

There were delays in establishing the diagnosis, on the one hand, when pediatricians did not interpret the symptoms correctly and, on the other hand, when specialties like orthopedics and ophthalmologists focused on their specific group of differential diagnoses.

Pediatricians and general physicians as the group of physicians seeing pediatric patients on a regular base and following their physical development should be sensitized for the problems of the posterior fossa tumors.

What are the differences regarding the diagnostic interval between low-grade and high-grade tumors?

The differences seen between benign and malignant tumors were expected and are easily understandable. Malignant tumors have a faster growth pattern. This way, they cause a faster developing hydrocephalus or space-occupying effect, which the brain is not able to compensate resulting in early signs of intracranial pressure or neurological symptoms. There were some cases of malignant tumors with a longer

time interval. We assume they were primarily of a lower grade followed by a secondary malignant transformation.

Is a secondary hydrocephalus predictive for an early diagnosis of the tumors?

The majority of tumors have caused a secondary hydrocephalus at the time of diagnosis. Many tumors with a long history of symptoms finally resulted in a hydrocephalus. The symptoms of a raised intracranial pressure led faster to the diagnosis, but were not predictive for a short symptomatic period in general.

What is the parent's role?

Overlooking our data, we had several cases in which only the parent's persistence led finally to the correct diagnosis of a posterior fossa tumor. Thus confirming the belief that parents are the best in interpreting their children's health.

Conclusion

Physicians seeing pediatric patients need to be further sensitized for the symptoms of posterior fossa tumors. The indication for cranial radiological diagnostics especially MRI should be seen wider as in the adult patient, even for rather unspecific symptomatology.

Particularly, neurosurgeons and neuropediatricians should more actively teach about posterior fossa tumors and their symptoms, diagnostics, therapy, and outcome.

We need to pay attention to the parents who present their children with specific complaints and symptoms as well as parent who state that there is just something wrong with their child.

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