THE CLINICAL CHALLENGES OF PEDIATRIC POSTERIOR FOSSA TUMORS

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ABSTRACT

Pediatric brain tumors occur mainly in the posterior fossa. Clinicians encounter numerous difficulties and challenges in establishing the diagnosis. Symptomatology varies according to the location of tumor, its rate of progression, and the age of the child. Most of them are usually nonspecific and misinterpreted as common benign childhood disorders. Consequently, a significant delay arises between the onset of symptoms and the diagnosis, despite the advent of high-resolution neuroimaging. This only leads to irreversible neurological damage and creates the pathway to a dismal prognosis. The purpose of this article is to present an overview of the complexities in diagnosing pediatric posterior fossa tumors. A particular attention is offered to the nonspecific overlooked symptoms, highlighting the most frequent misdiagnosis. Clinical presentations for specific tumor subtypes are also provided. Prompt diagnosis requires an improvement in the awareness and familiarity of the clinician with the signs and symptoms associated with this type of malignancy.

Keywords: pediatric brain tumors, posterior fossa tumors, increased intracranial pressure

Brain tumors are the most frequent solid tumors in children and the second most common pediatric neoplasia after leukemia [1]. The highest incidence occurs in infants and in children younger than 5 years old [2]. The rate of mortality outranks other childhood malignancies and the morbidity encompasses various physical, neuroendocrinological, psychological and cognitive burdens. More than that, the aggressive therapeutic approach extensively impairs the quality of life [1].

Unlike adults, children develop tumors especially in the infratentorial compartment (approximately 50%-55% of the pediatric brain tumors).

This type characterizes the first decade of life [1]. It comprises medulloblastoma, juvenile pilocytic astrocytoma, ependymoma and brainstem glioma [3].

Due to the neuroanatomical features of the posterior cranial fossa, these tumors are regarded as critical brain lesions, in severe cases causing even cerebral herniation [4]. They exert compression on the cerebellum or on the brain stem leading to predictable signs and symptoms [5].

However, when it comes to children, the clinical features become rather unpredictable. The clinical presentation varies according to the location of the tumor, to its rate of growth, to its aggressiveness and to the age of the child [4]. The clinical features can be classified into two main categories: increased intracranial pressure and localizing signs or symptoms [1]. These classic traits could be preceded by behavioral or personality modifications [2].

Actually, the early diagnosis raises the greatest amount of challenges due to the nonspecific initial symptomatology [6].

It can vary from isolated frequent vomiting which can be misdiagnosed as a gastrointestinal disorder, to even sudden neuropsychological changes [1,2].

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Therefore, children are referred to different pediatric subspecialties for several months before a final diagnosis is established, critical time being lost [6].

**INCREASED INTRACRANIAL PRESSURE**

The clinical mainstay for posterior fossa tumors are signs and symptoms related to increased intracranial pressure caused by the obstruction of the cerebrospinal fluid pathways [7]. Symptomatology is very variable and many times nonspecific. It can consist of vague and generalized symptoms or of severe and life threatening signs. The classic presentation is early morning headaches relieved with vomiting and the classic triad includes headache, nausea and vomiting. In young children, a part of these findings are usually mistaken for common childhood illnesses such as gastrointestinal ailments or influenza [1,2].

The signs are even more subtle in infants who can only present with irritability, poor feeding and lethargy [1]. Macrocephaly can be overlooked because the cranial sutures allow the skull to adapt to the increase in ventricular size [8]. In very young children, even after the closure of the fontanelles, the cranial sutures can still separate. Consequently, head circumference should be routinely measured for all children up to the age of 3 years [8].

The most prevalent symptom is the headache, nearly 40-50% of elementary school children and 60-80% of adolescents diagnosed with brain tumors having a history of frequent or chronic headache. Suspicion is raised when headaches become more frequent or severe, waking the child up or if they are accompanied by vomiting. A relevant sign is the association with an abnormality noticed during neurologic examination (abnormal eye movements, asymmetric motor or sensory examination, coordination problems, disorders of equilibrium and gait). Needless to say, performing a thorough neurological exam on all children with headache is mandatory [1,2,8].

A clinical setting which requires immediate attention is a permanent occipital headache associated with neck pain exacerbated by hyperextension, specific for tonsillar herniation. The back of the head shares common innervation with the components of posterior fossa. As a result, an aggravating occipital headache may represent the sign of increased cranial pressure secondary to an infratentorial tumor [1].

**LOCALIZING SIGNS AND SYMPTOMS**

Localizing signs of posterior fossa tumors are consistent with focal neurologic deficits. Children may present with seizures, cranial nerve dysfunction (nuclei or tracts of the third, fourth, or sixth cranial nerves are commonly involved), cerebellar dysfunction, abnormal or asymmetric strength, unusual sensations or asymmetric deep tendon reflexes [1,4]. Long-standing progressive symptoms in association with papilledema can characterize slow-growing tumors. Rapidly growing tumors can present concomitantly with pyramidal tract signs and disorders of ocular motility and diplopia [7].

Ataxia is suggestive for vermal and cerebellar hemisphere involvement, brainstem dysfunction and chronic hydrocephalus [7]. Truncal ataxia is specific of midline tumors and it consists of a tendency to fall frequently and a widely based gait. Hemi-cerebellar syndrome includes ataxia, nystagmus (usually develops late) and dysmetria. Tumors growing in the cerebellar hemisphere may cause ataxia of the contralateral limbs [4].

A specific clinical syndrome that can be related to infratentorial tumors is Parinaud’s syndrome. It includes paresis of upward gaze, accommodative paresis, nystagmus during attempts at upward gaze (convergence retraction nystagmus), eyelid retraction (Collier’s signs) and “setting-sun sign” (an ominous sign for advances stages of hydrocephaly) [8]. This syndrome is encountered more frequently in pineal region tumors [2].

Seizures occur more often in adolescents than in younger children (twice more often). They can be focal or generalized [8]. Indicators relevant for a brain tumor consist of a change in preexisting seizure features, status epilepticus as first presentation of a seizure, resistance to pharmacological treatment, prolonged postictal focal symptoms or neurological abnormalities. Neuroimaging is recommended for all simple and complex partial seizures and most generalized seizures, especially when focal abnormalities are found on EEG [1].
NEUROPSYCHOLOGICAL AND BEHAVIORAL CHANGES

Perhaps, the most frequent overlooked symptom in school-age children is regression in school performance accompanied by changes in behavioral traits and neuropsychological impairment. These category of children are commonly referred by a psychologist or pediatric psychiatrist after tedious months of unsuccessful therapy for this symptomatology [8].

Tumors in the posterior fossa alter the development of higher mental functions such as linguistic and emotional traits. They alter the neural circuits established between cerebellum and different cortical and subcortical areas of cerebrum resulting in the cerebellar cognitive syndrome. This syndrome encompasses executive function disturbances, personality change, impaired spatial cognition, and linguistic difficulties such as dysprosody, agrammatism and mild anomia. Consequently, the cerebellum damage hinders children’s intellectual ability, producing deficits that are difficult to diagnose during childhood and adolescence [9].

Children with infratentorial tumors suffer from somatic concerns, aggressiveness, anxiety and internalizing disorders. In some cases behavioral changes have been described such as separation anxiety, pathological laughter, aggression, irritability, isolation, depression or anorexia. Personality changes such as disinhibited behavior or blunted affect have also been reported [9].

CLINICAL PRESENTATIONS FOR SPECIFIC TUMOR TYPES

A basic knowledge of the specific types of posterior fossa tumors and their most common distinct clinical features can enable clinicians to establish an early diagnosis and refer the patients for initiation of prompt treatment [6].

Medulloblastoma

Medulloblastoma is the most common pediatric posterior fossa tumor (40% of cases). The incidence reaches two peaks at 3 and 7 years old, with a median age of 5-7 years, affecting more often boys (ratio 2:1). The rapid progression determines a relatively rapid onset of the symptoms (over the course of weeks or 2-3 months) [2,3]. Developing in the midline cerebellum at the level of the fourth ventricle, patients present with signs and symptoms of increased intracranial pressure and cerebellar dysfunction [10]. The onset may consist of irritability, behavioral changes and declining school performance. Afterwards, children present vomiting upon awakening, horizontal diplopia, head tilt, clumsiness and occipital or frontal headaches. After six months of evolution, they develop neurologic signs such as papilledema, strabism, ataxia and weakness. In infants, the signs are macrocephaly, splitting of the cranial sutures or a bulging anterior fontanelle [10].

Cerebellar juvenile pilocytic astrocytoma

Cerebellar astrocytoma is the second most common pediatric posterior fossa tumor (30% of cases). The peak of the incidence is between 5 and 15 years of age, with a mean age of 6.8 years, affecting equally boys and girls. Unlike medulloblastoma it is a slow-growing tumor, patients presenting an indolent, gradual onset of symptoms (over 3 to 6 months) [2,3]. As a result, they are large at the time of the diagnosis [11].

Initial symptomatology is usually mild and nonspecific and is the result of increased intracranial pressure. Headache is the prevalent symptom (75-97% of cases) and is exacerbated by recumbency. In the beginning it is localized in the frontal lobe and later migrates to the occiput. Vomiting is the second most common complaint (64-84% of cases). Papilledema is found in 40-80% of cases in association with signs of cerebellar dysfunction (ataxia, gait disturbance, appendicular dysmetria, wide-based gait). Masses of the cerebellar hemisphere causes ataxia and dysmetria in the ipsilateral limbs, whereas midline masses cause truncal and gait ataxia [11].

Ependymoma

Ependymomas are the third most common pediatric posterior fossa tumors (10-15% of cases). The mean age of presentation is 6 years old, with approximately 40% of cases occurring before 4 years of age. They develop along the floor and the roof of the fourth ventricle [2,3]. They usually invade adjacent structures or extend into the aqueduct of Sylvius, foramen of Magendie, foramen of
Luschka. The duration of symptoms is less than 6 months at the time of diagnosis, half of the children presenting within 1 month from the onset [12].

The clinical presentation consists of signs and symptoms of increased intracranial pressure (vomiting, headache, ataxia). Infiltration into the brainstem and growth through the foramina of Luschka produces cranial nerve palsies, torticollis and meningismus. Children younger than two years of age present with nonspecific signs such as irritability, vomiting, lethargy, macrocephaly and gait imbalance [12].

**Brainstem glioma**

Brainstem glioma are the fourth most common pediatric posterior fossa tumors (10-15% of cases). They equally affect boys and girls [2,3]. They exhibit stereotypical growth patterns. The progression and shape, especially for benign slow-growing tumors, depend strictly on the local anatomical structures. On the contrary, high-grade tumors grow disrespectfully of the surrounding tissue matrix [13]. Most often the clinical presentation is characterized by a relatively rapid onset of symptoms (over 1 to 4 months) [2,3].

The clinical features depend on the location of the tumor and the involved anatomical structures. Focal midbrain tectal gliomas determine hydrocephalus and extracerebral motor palsies. Dorsally exophytic tumors (occur in the medulla) produce signs of increased intracranial pressure and present with failure to thrive in infants. Cervicomedullary tumors (arise in the lower medulla and upper cervical spinal cord) cause dysphagia, dysarthria, nausea, vomiting, failure to thrive in infants, upper motor neuron dysfunction, torticollis and sensory loss. Diffuse intrinsic pontine gliomas show a rapid onset (less than one month): cranial nerves dysfunction (VI, VII), long tract signs (paresis, hyperreflexia, spasticity, Babinski positive) and ataxia [13].

**Atypical teratoid/rhabdoid tumor**

Atypical teratoid/rhabdoid tumor is a rare type (5% of cases) of aggressive embryonal malignancy. It occurs mostly in very young children (before the age of 3), with a mean age at diagnosis of 1-7 months [2,3]. Children present with rapid onset (over days to weeks) signs of increased intracranial pressure along with facial weakness and strabismus [2]. Infants present with recent macrocephaly and older children with truncal ataxia [3]. The prognosis is extremely unfavorable. Median survival is less than 10 months, and most children die within a year after the diagnosis [14].

Establishing a prompt diagnosis for pediatric posterior fossa tumors is of paramount importance for improving not only the long-term prognosis, but also the quality of life of these children. Each child presenting with specific or nonspecific signs of increased intracranial pressure should undergo a thorough clinical and neurological examination, identifying signs suggestive for brain tumors developing in the infratentorial compartment. Increased attention should be given to symptoms qualified as benign pediatric ailments or to sudden behavioral changes. Needless to say, it is better to be cautious and exclude the presence of a brain tumor, than be confronted with the severe consequences of misdiagnosis.

**REFERENCES**


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