v. 3, n. 51, 2023

International Journal of Health Science

TUMORS OF THE CENTRAL NERVOUS SYSTEM IN PEDIATRICS AND THEIR SYNDROMES – LITERATURE REVIEW

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Abstract: Introduction: Neoplasms of the central nervous system are epidemiologically very important, as they are the second most common type of tumors in childhood and adolescence[3]. **Objective**: Central nervous system tumors can present very varied clinical manifestations, depending mainly on the child's age and location[8]. There is a spectrum of syndromes associated with these tumors, which this article aims to describe. **Method**: The search was carried out in the PubMed, BVS and Scielo databases, and was limited to articles between the period 2019 to 2023 that met the criteria of being literature reviews and case reports. **Result**: Based on the findings, the most common syndromes in children with central nervous system tumors, depending on their location and extent, are the intracranial, ataxic, pyramidal, convulsive, neuroendocrine, diencephalic and macrocranial hypertension syndromes.
Keywords: Neoplasms of the Central Nervous System; Pediatrics; Paraneoplastic Syndromes.

INTRODUCTION

The entire classification of primary tumors of the Central Nervous System (CNS), established by the World Health Organization (WHO), is based on the histological origin of the neoplastic cell[1].

Primary CNS (brain and spinal cord) tumors are those originating from the uncontrolled proliferation of their own cells. Less often, neurons, as they have low replication capacity; Glial cells, for example: astrocytes and oligodendrocytes; Meningeal cells; choroid plexus or ependymal cells; And finally, some tumors can develop from embryonic and germ cells[2].

The epidemiology of these neoplasms is important, as it is the second most common type in childhood and adolescence, second only to leukemias3. Most cases are concentrated in children aged five years

or less, with an American incidence of 52 cases/1,000,000 children [4].

Regarding the risk factors associated with the development of brain tumors in childhood, we can highlight, mainly: Genetic syndromes and neurofibromatosis type one [5].

The vast majority are located in the brain and only a small percentage are located in the spinal cord6. Regarding the frequency of location, we observed that in 43.2% of the cases, the location was infratentorial; in 40.9% of the cases it is supratentorial and, in 11% of the cases, the location is in multiple places. In addition, 5% of cases are of medullary tumors[6]. Exceptions to this distribution occur only before one year and after ten years, in which supratentorial tumors are the most frequent [6].

The most common pediatric tumors are astrocytomas, in 30% of cases, medulloblastomas, in 15%, and ependymoma in 6%6. Five-year survival for the 0-19 age group is approximately 65%[7].

Central nervous system tumors can present very varied clinical manifestations, depending mainly on the child's age and location, for example, whether it is supratentorial, midline, infratentorial or medullary8. The picture is usually insidious, progressive, with periods of exacerbation and remission of some symptoms[8].

There is a spectrum of syndromes associated with these tumors, which this article aims to describe.

One of the most frequent, especially in large neoplastic and midline masses, which can cause compression of the ventricular system, is the Intracranial Hypertension (ICH) syndrome, which is characterized by increased brain pressure and can be manifested by headache, usually morning, exacerbated by the Valsalva maneuver[9]. It is important to point out that all headaches of acute onset (< 6 months), always located in the same region, progressive, that intensify with Valsalva maneuvers (coughing, sneezing, defecating), worse in the morning or that wake the patient up at night, must call attention to the possibility of HIC[9]. In addition to vomiting, anorexia, weight loss, behavioral and personality changes, irritability, lethargy, paralysis of the abducens nerve, the sixth pair of cranial nerves, which has a very long intracranial course, and papilledema[9].

With the progression of ICH, signs appear that reflect brainstem dysfunction with imminent herniation: bradycardia, arterial hypertension and changes in respiratory rhythm[9].

Severe and acute cases of decompensated ICH are identified by herniation syndromes, and these define a medical emergency[9]. The most commonly seen herniation is transtentorial, which occurs when the medial portion (uncus) of the temporal lobe moves caudally and medially over the midbrain, compressing the third pair of cranial nerves, the posterior cerebral artery, and the cerebral peduncle[9]. Clinically, there is depression of the level of consciousness due to compression of the activated ascending reticular system, ipsilateral mydriasis, decortication posture and ipsilateral hemiparesis[9].

Another syndrome seen in CNS tumors in children is the ataxic one, with manifestations caused by injury to the cerebellum and its pathways[10]. It is characterized by wide-based or drunken gait, dysbasia (increased base of support to remain standing), incoordination, tremor, dysdiadochokinesia (inability to perform alternating movements), dysmetria (inability to perform finger-to-toe, finger-to-toe tests). nose, hypotonia, and dysarthria (slurred speech)[10].

A syndrome of great importance is the pyramidal syndrome[11]. It manifests when there is compression of the pyramidal pathways such as the internal capsule, corona radiata or cerebral peduncles[11]. Symptoms such as spasticity, deep tendon hyperreflexia, clonus

and pathological reflexes such as Babinski's sign can be found[11].

If the lesion is hemispherical, directly affecting the cortex, the convulsive syndrome, characterized by disordered movements and loss of consciousness of the child, can also occur[12].

Tumors of the third ventricle and suprasellar region can manifest with diabetes insipidus, galactorrhea, pubertal precocity or delay, hypothyroidism or growth disorders due to compression of the hypothalamic-pituitary structures, which are responsible for the endocrine part of our body[13].

In diencephalic syndrome, being more common between 18 months and 3 years, we have growth and development failure, significant weight loss, swelling, increased appetite and euphoric mood[14].

Finally, we will describe the Macrocrania syndrome. In children under two years of age, symptoms related to cranial tumors tend to appear later, as the symptoms of ICH are attenuated by progressive diastasis of sutures and bulging of fontanelles as the tumor expands and, in addition, the most prominent clinical symptoms are nonspecific and easily confused with a number of benign pediatric diseases[15]. For example, repeated vomiting is treated as gastroesophageal reflux disease. The most frequent tumors in this age group are teratomas (< 3 months), medulloblastoma and ependymomas[15].

MATERIAL AND METHODS

The search was carried out in the PubMed, BVS and Scielo databases, and was limited to articles between the period 2019 to 2023 that met the criteria of being literature reviews and case reports.

Then, the keywords of the titles of the articles were analyzed and those whose theme best fits our objective were selected and 15 articles were selected for full reading.

DISCUSSION

Children, especially those under two years of age, with a central nervous system tumor, do not always present these symptoms or part of them, as their neuroplasticity and growth of neurological organs attenuate the compression that tumors can cause[15].

CONCLUSION

Based on the findings, the most common syndromes in children with central nervous system tumors, depending on their location and extent, are the intracranial, ataxic, pyramidal, convulsive, neuroendocrine, diencephalic and macrocranial hypertension syndromes.

CONFLICT OF INTERESTS

nothing to disclose.

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