Intracranial Tumors in childhood – A Review

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Introduction

Most commonly intracranial tumors are located in infratentorial region and hind brain is most common site for their origin. Infratentorial tumors account for about 45– 60% of all pediatric brain tumors, and the most common infratentorial tumors include juvenile pilocytic astrocytoma (JPA), medulloblastoma, ependymoma, and brainstem glioma[1]. An accurate diagnosis of these space occupying lesions is essential and so has important clinical implications related to prognosis and treatment. Craniopharyngiomas are one of the most common tumors arising from the pituitary region in children. Suprasellar gliomas and hypothalamic hamartomas are known for their unique clinical presentations and imaging features. Advanced MR neuroimaging techniques allow assessment of the physiologic features of brain tumors, resulting in better preoperative characterization as well as in better outcome.

Pilocytic astrocytoma

Pilocytic astrocytomas (PA), also known as juvenile pilocytic astrocytomas (JPA), are low-grade, relatively well-defined astrocytomas (WHO Grade I). They tend to occur in young patients and have a relatively good prognosis[1].

They exhibit wide range in appearance on imaging, with the majority presenting as a large cystic lesion with a brightly enhancing mural nodule. Calcification can be present in around one fifth of cases. They typically arise from midline structures, and around 60% are in the cerebellum. Pilocytic astrocytomas are tumors of young people, with 75% occurring in the first two decades of life, typically late in the first decade (9-10 years)[2]. There is no recognized gender predisposition. Although only accounting for between 0.6 -5.1% of all intracranial neoplasms (1.7-7% of all glial tumors) they are the most common primary brain tumor of childhood, accounting for 70-85% of all cerebellar astrocytomas.

Clinical presentation depends on location. In the posterior fossa tumors there is predominantly a mass effect with signs of raised intracranial pressure, especially when hydrocephalus is present. Bulbar symptoms or cerebellar symptoms may also be present.

There is a strong association with neurofibromatosis type 1 (NF1). NF1 associated tumors have a tendency to affect the optic nerves and chiasm and may also have a better prognosis. Pilocytic astrocytomas are seen in up to 20% of all patients with NF1 and typically manifest in early childhood. Approximately one-third of pilocytic astrocytomas involving the optic nerves have associated NF1[3].

By far the most common location is the cerebellum (60%). The distribution within the cerebellum varies with many tumors involving both the vermis and the cerebellar hemisphere.

In general they typically arise from midline structures- optic nerve/optic chiasm (25-30%), very common location in NF1, hypothalamus/adjacent to third ventricle, brainstem, other less common locations, cerebral hemispheres, cerebral ventricles, velum interpositum and spinal cord.

Medulloblastoma

Medulloblastoma is a fast-growing, high-grade tumor. The various types of medulloblastoma include: Classic, desmoplastic nodular, large-cell or anaplastic, medulloblastoma with neuroblastic or neuronal differentiation, medulloblastoma with glial differentiation, medullomyoblastoma, melanotic[4,5]. Medulloblastoma is always located in the cerebellum—the lower, rear portion of the brain. It is unusual for medulloblastomas to spread outside the brain and spinal cord.it is a fast-growing, high-grade tumor. It is the most common of the embryonal tumors—tumors that arise from "emybrynic" or "immature" cells at the earliest stage of their development.

The most common symptoms of medulloblastoma include behavioral changes,

changes in appetite, symptoms of increased pressure on the brain (e.g., headache, nausea, vomiting, and drowsiness, as well as problems with coordination). Unusual eye movements may also occur. Medulloblastoma is relatively rare, accounting for less than 2% of all primary brain tumors and 18% of all pediatric brain tumors[5]. More than 70% of all pediatric medulloblastomas are diagnosed in children under age 10. Very few occur in children up to age 1[6].

Medulloblastoma in adults is less common, but it does occur. About one-third of all medulloblastomas diagnosed are found in adults between the ages of 20-44. The incidence in adults sharply decreases in frequency after age 45, with very few older adults having this tumor. Medulloblastoma occurs more often in men than in women.

Atypical Teratoid Rhabdoid Tumor (AT/RT)

Atypical teratoid rhabdoid tumors (AT/RT) are very rare, aggressive tumors of the central nervous system, occurring mostly in the cerebellum and the brain stem. AT/RT usually occurs by age 3, but arises in older children, as well[7,8,9].

AT/RT's are part of a larger group of malignant tumors called rhabdoid tumors, which can occur outside of the brain in the kidneys, liver or other locations. In most cases, AT/RT is associated with a specific genetic mutation (INI1) that can occur spontaneously or be inherited.

AT/RT was previous thought to be a type of medulloblastoma. However, it is now known to be a separate type of tumor and is treated differently. AT/RT represents only 1 to 2 percent of childhood brain[8].

Ependymomas

Ependymoma is the third most common posterior fossa tumor in children. Incidence peaks in patients 0–4 years old. Approximately 70% of intracranial ependymomas are infratentorial and arise from ependymal cells lining the floor of the fourth ventricle and foramen of Luschka.

These tumors are divided into four major types: Subependymomas (grade I): Typically slow-growing tumors, Myxopapillary ependymomas (grade I): Typically slow-growing tumors, Ependymomas (grade II): The most common of the ependymal tumors[10]. This type can be further divided into the following subtypes, including cellular ependymomas, papillary ependymomas, clear cell ependymomas, and tancytic ependymomas, Anaplastic ependymomas (grade III): Typically faster-growing tumors. Histologically, ependymomas tend to have a high proportion of intracellular myxoid accumulation and cyst formation[11].

The various types of ependymomas appear in different locations within the brain and spinal column. Subependymomas are intra cranial tumors which usually appear near a ventricle. Myxopapillary ependymomas tend to occur in the lower part of the spinal column. Ependymomas are usually located along, within, or next to the ventricular system. Anaplastic ependymomas are most commonly found in the brain in adults and in the lower back part of the skull (posterior fossa) in children. They are rarely found in the spinal cord.

Brainstem Glioma

Brainstem gliomas account for ~25% of all posterior fossa tumors and are most common in children between 7 and 9 years of age 5. There is no recognized gender or racial predilection[12,13,14,15].

Brainstem gliomas are also recognized in adults, although they are rare accounting for only 2% of adult brain tumors. They typically occur in younger adults (third and fourth decade) and tend to be of low grade (WHO I or II). The duration of symptoms is usually much shorter in diffuse gliomas, in which the history is typically very short (a few days). Additionally, diffuse gliomas more frequently have multiple cranial nerve palsies[14].

Recognized histological types of brainstem gliomas include diffuse astrocytoma, focal glioma, exophytic glioma, NF1 associated brainstem glioma.

Hemangioblastoma

Hemangioblastomas account for 1–3% of all intracranial neoplasms, and most occur in middle-aged adults. In children younger than 18 years old, slight male predilection in adults with M:F ratio of 1.3-2.6, these tumors are extremely rare, with an incidence of less than 1 per 1 million[16,17,18].

These are tumors of vascular origin and occur both in patients with von Hippel Lindau (vHL) as well as sporadically. They are WHO grade I tumors that can occur in the central nervous system or elsewhere in the body, including kidneys, liver and pancreas.

These tumors generally present on imaging as sharply demarcated homogeneous masses

composed of a cyst with non-enhancing walls, except for a mural nodule which vividly enhances and often has prominent serpentine flow voids. The main location are 95% in posterior fossa(85% in cerebellar hemisphere, 10% in the cerebellar vermis & 5% in medulla) only rarely do they extend beyond the cerebellum into the cerebellopontine angle.[17,18]

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