Brain tumours are the most solid tumours in childhood and the second most common form of cancer, ranking only behind leukemia in terms of frequency of diagnosis. Mortality tends to be high as many children succumb to this illness. There is strong evidence that the incidence of pediatric brain tumours may be rising, but others have argued that this observed increase may be due to advancement and increased availability in diagnostic technologies.

Contrary to adult counterparts, almost 70% of pediatric brain tumours arise from the infratentorial compartment. The clinical presentation of these tumours may follow distinct symptoms as the disease advances. However, early diagnosis is often challenging due to initial non-specific symptoms. The child may go through several investigations for months before a final diagnosis is made through imaging techniques.

Anatomy of posterior fossa

The intracranial compartment is divided into anterior, middle and posterior fossas with the latter two being separated by tentorium (a thickened bilayer of dura).

The infratentorial compartment contains critical brain structures that include the cerebellum, the brain stem and cranial nerves.

Another important structure that exists in the posterior fossa is the fourth ventricle which is connected to the remainder of the ventricular system in the supratentorial compartment. The ventricular system is involved in the production and flow of cerebrospinal fluid (CSF) that eventually leaves the fourth ventricle, bathing the surfaces surrounding the brain (Figure 1).

Classification

Most common pediatric posterior fossa tumours can be divided into two major categories: primitive neuroectodermal tumours (PNETs) and glial tumours (Table 1).

PNETs are highly malignant, undifferentiated neoplasms affecting children. The prototype is medulloblastoma, commonly found in the posterior fossa.

PNETs are highly malignant, undifferentiated neoplasms affecting children. The prototype is medulloblastoma, commonly found in the posterior fossa. PNETs represent nearly one-third of all posterior fossa tumours with a 2:1 ratio of male predominance. It typically affects children between the ages of one to 10, with the rate of incidence peaking at the age of seven. Nearly 40% of medulloblastomas invade the brain stem and CSF.

Interestingly, the primary central nervous system (CNS) neoplasms seldom display systemic metastases. However, > 5% of medulloblastomas can spread to other sites, such as bone and lungs. The average period for diagnosis, after the onset of symptoms, is usually three months.
Glial tumours

Glial tumours can be divided into three main classes:
- astrocytomas,
- ependymomas and
- brain stem gliomas.
In general, glial cells exist in the microenvironments of neural tissue and play several important roles, such as nerve regeneration, cellular migration, cell-to-cell adhesion, as well as providing structural support.

Glial cells are divided into four cell types:
- astrocytes,
- oligodendrocytes,
- microglial cells and
- ependymal cells.

Astrocytomas

Astrocytomas are the most common glial tumours found in the posterior fossa. Cerebellar astrocytomas (CAs) represent 20% of pediatric brain tumours with half located in the posterior fossa. These neoplasms affect children between the ages of one and 10, with the mean age of about seven. It rarely affects children less than one-year-of-age. There is no sex predominance and the average duration of

Table 1
Classification of posterior fossa tumours

<table>
<thead>
<tr>
<th>1. Primitive neuroectodermal tumours</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Medulloblastoma</td>
</tr>
<tr>
<td>- Cerebral neuroblastoma</td>
</tr>
<tr>
<td>- Pineoblastoma</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>2. Glial tumours</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Astrocytoma</td>
</tr>
<tr>
<td>- Ependymoma</td>
</tr>
<tr>
<td>- Brain stem glioma</td>
</tr>
<tr>
<td>- Diffuse intrinsic pontine glioma</td>
</tr>
<tr>
<td>- Cervical medullary</td>
</tr>
<tr>
<td>- Dorsal exophytic</td>
</tr>
<tr>
<td>- Tactile</td>
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</tbody>
</table>

Figure 1. Depiction of the various structures of a posterior fossa tumour
symptoms, before diagnosis, is usually five to nine months. Most CAs are low-grade neoplasms and invasion is a rarity.

**Ependymomas**

Ependymomas are derived from the ependymal lining of the ventricular systems and make up the third most common posterior fossa tumours in the pediatric population.

They are more prevalent in children less than three-years-of-age. Most ependymomas arise from the fourth ventricle and generally carry a poor prognosis (about 60%).

The rest arise in the lateral ventricular systems. The median time of diagnosis, after the onset of symptoms, is about three months.

**Brain stem gliomas**

Brain stem gliomas are astrocytomas that mostly arise from the pons. Diffuse intrinsic pontine gliomas make up about 80% of these tumours. The median age for occurrence is five to nine years. The time of diagnosis is usually within two months of symptoms onset. The incidence is the same in both females and males. These tumours usually considered inoperable. The prognosis is poor; a five year survival rate is less than 20%, despite aggressive chemotherapy and radiation.

**Clinical presentation**

Clinically, pediatric posterior fossa tumours present with distinct symptoms (Table 2). Diagnosis is often missed in their early presentation and a child may go through several investigations before the tumor is discovered with imaging techniques such as MRI or CT scans. Therefore, a careful history and neurological examination is of paramount importance for early diagnosis. The majority of the cases, especially astrocytomas and medulloblastomas, present with features associated with increased intracranial pressure (ICP).

### Table 2

**Signs and Symptoms of posterior fossa in children**

- Initial behavioral changes
- Lethargy, irritability, decreased appetite
- Characteristic headache:
  - Worse at night and early morning
  - Improves with vomiting
  - Worse with recumbency, cough and defecation
- Head tilt, nuchal rigidity, ataxia, nystagmus, cranial nerve dysfunction
- Papilledema

### Table 3

**Key points for diagnosis**

- Consider posterior fossa tumours as differential
- Diagnosis of a child with non-specific behavioural changes (i.e., lethargy, decreased appetite and irritability)
- Distinct features of headache as disease progresses
- A detailed neurological examination most notably fundoscopic exam
- Consider early CT or MRI

Initially, a child may present with non-specific symptoms such as:
- irritability,
- decreased appetite and
- lethargy.

With ICP, a triad of headache, lethargy and vomiting manifests. The nature of the

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Diagnosis is often missed in their early presentation and a child may go through several investigations before the tumour is discovered with imaging techniques, such as MRIs or CT scans.

headache is classic and deserves special attention by clinicians to ensure an early diagnosis.

Typically, the child complains of early morning and nighttime headaches. Headaches improve after vomiting in the morning and the child may continue to function well for the remainder of the day.

Recumbency and straining manoeuvres such as coughing or defecation, may worsen the headaches. From a physiological point of view, vomiting causes hyperventilation which decreases carbon dioxide which, in turn, decreases ICP. On the contrary, sleep causes hypoventilation, increasing ICP and worsening the headache.

With the involvement of the cerebellum and the invasion into the cervical subarachnoid space, more commonly with ependymomas and CAs, the child presents with:
• ataxia,
• nystagmus,
• nuchal rigidity and
• head tilts.

Therefore, chronic torticollis should alert one about the possibility of a posterior fossa mass. In the case of brain stem gliomas or invasion of tumours into the brain stem there may be associated facial or bulbar palsies or evidence of other cranial nerve abnormalities. Sixth nerve palsy may occur as a result of hydrocephalus and increased ICP and may not be associated with brain stem involvement. Examination of fundi is of paramount importance as about 50% of patients have papilledema at the time of diagnosis.

Conclusions

The initial presentation of pediatric posterior fossa tumours often pose a diagnostic challenge to the FP, as the child may present with vague and non-specific symptoms. A basic knowledge in the types of posterior fossa tumours and their often distinct clinical signs and symptoms may aid clinicians in early diagnosis and referral.

As far as the management of these tumours is concerned, despite significant advancements in both neurosurgical techniques, as well as adjuvant treatments, brain tumours remain the most leading cause of cancer death in children.

As in the case with other CNS tumours, future advances in genetic analysis and an understanding of cellular and molecular mechanisms in the micro-environments underlying local formation, growth, or invasion of these tumours may lead to a more tailored approach to adjuvant therapy, which may, in turn, lead to improvements in outcome.

Resources