Common and Uncommon Presentation of Childhood Brain Tumour

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Brain tumor is, second to leukemia, the commonest childhood malignancy and it ranks first among all solid tumors. Overseas data show that the annual incidence of brain tumor is around 27 per million per year for those patients less than 19 years of age. According to the Hong Kong Cancer Registry, the local incidence is of similar magnitude.

Types of brain tumor that commonly seen in Hong Kong include astrocytomas, brainstem gliomas, medulloblastomas, intracranial germ cell tumors, craniopharyngiomas, ependymomas and other rarer tumor types. Similar to Japanese series, we see a higher incidence of intracranial germ cell tumors as compared with western countries.

Age is important because it governs the type of tumor and its related symptomatology, Similarly, the pathological diagnosis would affect the location as well as the aggressiveness of the tumor. It is important to note that most of the childhood brain tumors are in the infratentorial region.

The presentations of brain tumor could be classified into four main categories. These include **raised intracranial pressure and associated headache**, **neurological deficit,** partial or generalized **seizure** and **change in behaviour / level of consciousness.** The warning features of headache that may be associated with brain tumor include **recent in onset, progressive increase in severity, localized headache** in the occipital region, headache increased with straining, **nocturnal headache** awakening the child from sleep or **morning headache** especially if associated with vomiting.

At the time of diagnosis of brain tumor, more than **80% have abnormal neurological signs** on examination, which include vomiting, unsteadiness, visual difficulties, educational or behavioral problems, seizures and growth or endocrine abnormalities. The most frequent ones are cranial nerves abnormalities, cerebellar signs and papilloedema. However it should be cautioned that these features may not be present in the initial stage. In Hong Kong, the average time lapse between appearance of first symptom to actual diagnosis is around 3 months. This suggests that the early presenting features of childhood brain tumors can be subtle and a longitudinal follow-up is essential in identifying this group of children early.

For the infant age group, observation such as increase in head circumference disproportional to other body parameters, especially if it is crossing the percentile line, a bulging anterior fontanelle, regression in developmental milestones or focal neurological deficit, and feeding problem with repeated vomiting warrant further attention.

Two common **endocrine abnormalities** that associated with brain tumor are **C**entral **Diabetes Insipidus** and **Pubertal Disorder**. Tumor located in the suprasellar region near the pituitary, for example craniopharyngioma, germ cell tumor and some astrocytomas, could cause destruction of the pituitary gland leading to endocrine dysfunction.

About 2/3 of the intracranial germ cell tumor are located in the pineal gland region, while the other 1/3 are located in the suprasellar region. Intracranial germ cell tumors that are located in the suprasellar region may present with diabetes insipidus as a result of destruction of the posterior pituitary gland. For germ cell tumor that presented with precocious puberty, **precocious pseudopuberty** to be exact, it is mostly mediated through the effect of beta-human chorionic gonadotrophin that is secreted by the tumor rather than the disruption of the pituitary function.

Lastly, **paraneoplastic neurological disorder (PND)** is a rare neurological complication of malignancy (1% of adult patients.) It is triggered by the neuroantigen expressed on the tumor tissue which causes~~d~~ an indirect immune mediated effect on the central nervous system or the peripheral nervous systems. It is rare in children. Perhaps the most familiar one to the paediatric population is the “opsoclonus myoclonus syndrome” associated with neuroblastoma. Other tumors known to be associated with PND in the adolescent age group include Hodgkin or non-Hodgkin lymphoma and ovarian tumor.