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Imaging Features of Common Pediatric Intracranial Tumours: A Primer for the Radiology Trainee

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Brain tumours constitute the second most common malignancy in children <15 years of age and the second most common cause of death in children [1]. Whereas supratentorial tumours are more common in children <3 years of age, posterior fossa tumours are more common in children 4-10 years old, and both locations are equally common in children after 10 years of age [2,3]. Clinical presentations depend on the location, size, and extent of the lesion, and are usually secondary to direct compression of underlying brain parenchyma, mass effect causing shift of intracranial contents, raised intracranial pressure, or focal cerebral ischemia. However, symptoms may be nonspecific and include headache, ataxia, nausea, vomiting, weakness, gait disturbance, seizures, torticollis, and visual or speech changes. Although usually insidious, acute presentations due to stroke or obstruction of cerebrospinal fluid flow can occur.

In this paper, we review the imaging appearances and approach to the differential diagnosis of some of the common supratentorial and posterior fossa tumours in children, selected based either on high prevalence or aggressiveness. We provide a broad overview of the imaging features of these selected tumours, which a general radiologist in practice or resident in training is likely to come across, along with the most likely differentials for each, although this is by no means an exhaustive compilation. Where there are unique features in the clinical presentation these will be discussed within the tumour-specific text, to act as a diagnostic aid. Although outside the scope of this basic review paper, effort has been taken wherever needed to reflect the recently added information relative to 2016 World Health Organization (WHO) classification of brain tumours including the relevance of molecular biology in management of some of these tumours [4].

Supratentorial Tumours

Optic Pathway Glioma

Epidemiology and presentation

Optic pathway gliomas (OPGs) are WHO grade I tumours that account for approximately 15% of all supratentorial tumours in the pediatric population, and >75% are diagnosed in the first decade of life [5]. OPG affect 11%-30% of children with neurofibromatosis type 1 (NF1) [6]. Patients can be asymptomatic or present with visual loss, proptosis, strabismus, or endocrine or hypothalamic disturbance, depending on the location along the optic pathway. Histologically, the majority are pilocytic astrocytomas. Pediatric OPG are usually indolent whereas presentation in the non-NF1 general population is often more aggressive [7]. Management approaches include observation, chemotherapy, radiation, or surgery [4,8]. The rapidly improving understanding of molecular markers and newer treatment strategies based on tumour biology has resulted in a paradigm shift in management of these tumours [9]. Prognosis is generally favorable, with a high degree of variability based on histology. In addition, the presence of NF1 and an anterior location are associated with a more favorable

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prognosis, whereas younger age at presentation is associated with a poorer prognosis [10].

Imaging

OPGs most often present as a mass causing enlargement, buckling, or kinking of the optic nerve, apparent as an isodense mass with variable enhancement and calcification on computed tomography (CT). Magnetic resonance imaging (MRI) is the preferred imaging modality, with the tumours appearing iso- to hypointense on T1-weighted imaging (WI) and hyperintense on T2WI with variable degrees of enhancement with gadolinium [10,11] (Figure 1A and B). Bilaterality and hydrocephalus are more commonly seen in the setting of NF1, while extraoptic extension into the adjacent basal ganglia and white matter as well as intratumoural cystic areas is more commonly seen in the non-NF1 population.

Differential

The major differential for intraorbital OPG is optic nerve meningioma, which, although more common in adults, tends to be more aggressive if presenting in childhood. On imaging, OPG infiltrates and deforms the optic nerve, whereas with meningioma the optic nerve is visualized on postcontrast imaging with the tumour extending along the nerve, giving the so-called tram-track appearance [12]. Inflammatory diseases such as sarcoidosis, optic neuritis, or inflammatory pseudotumour are also in the differential.

For chiasmatic-hypothalamic OPGs, the main differential diagnosis includes craniopharyngioma, germ cells tumours, and hypothalamic hamartoma [2]. The presence of optic nerve enlargement and NF1 stigmata (clinical features or radiological focal abnormal signal intensity) favor the diagnosis of OPG. On the other hand, the presence of low T2 signal and diffusion restriction, both suggesting high cellularity [3], are suggestive of germ cell tumours that may also be associated with an additional pineal region tumour. Craniopharyngiomas can sometimes be differentiated from astrocytoma owing to their primary cystic appearance, more calcification, higher T1 signal within the cystic component and by the more heterogeneous appearance of their solid component [13].

Craniopharyngioma

Epidemiology and presentation

Craniopharyngiomas are common suprasellar or intrasellar tumours, WHO grade I, that account for approximately 5%-10% of all pediatric brain tumours and are believed to arise from remnants of the Rathke pouch [14]. There is a bimodal age distribution of presentation, with a peak in the early second decade and a second peak in the sixth decade [14−16]. Incidence is similar in male and female patients. They can arise anywhere along the pituitary stalk from the floor of the third ventricle to the pituitary gland, or rarely below the sella turcica in the sphenoid sinus, within the path of the embryological craniopharyngeal canal [17].

Twenty percent of children and 80% of adults present with visual symptoms [18]. Hormonal changes are also likely due to the infundibular position of the tumour with amenorrhea, short stature, delayed puberty, reduced libido, and diabetes insipidus all described. Symptoms may also result from raised intracranial pressure. Although surgery alone is often the therapeutic standard of care, radiation, or intracystic chemotherapy are options for nonresectable tumours. Prognosis is variable—although craniopharyngiomas are classified as histologically benign tumours, the 20-year survival is around 60%, but once recurrence occurs, survival rates drop to around 25%. Being <5 years of age at diagnosis, the presence of calcifications, and incomplete tumour resection are associated with worse prognosis [19].

There are two histological types of craniopharyngioma: adamantinomatous (90%) and papillary (10%), but a mixed variant also exists.

Imaging

In general, craniopharyngiomas are large tumours with suprasellar and intrasellar involvement; 15%-20% are purely suprasellar whereas a purely intrasellar location is very uncommon (<5%) [18,20]. The papillary subtype may present as a third ventricular lesion. The pediatric variant is usually a

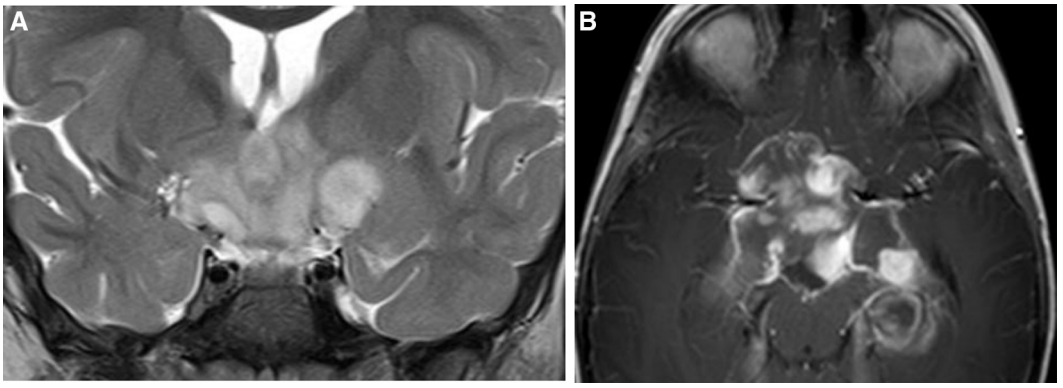

Figure 1. Optic pathway glioma: a 3-year-old boy with an infiltrative supratentorial mass lesion. (A) Coronal T2-weighted imaging showing bright signal and (B) axial postcontrast image showing heterogeneous and rim enhancement (panel B obtained 2 years after panel A). The lesion arises from the optic chiasma and extends into the surrounding structures including bilateral basal ganglia, bilateral mesial temporal lobes, and bilateral basal frontal lobes.