Craniopharyngiomas

Craniopharyngiomas are a range of slow-growing tumours that arise from remnants of the craniopharyngeal duct and/or Rathke's cleft. They generally occupy the sellar and suprasellar region, most commonly arising on the pituitary stalk and projecting into the hypothalamus. The craniopharyngeal duct extends from the pharynx to the sella turcica and the third ventricle and tumours can occur anywhere along its length. Tumours can therefore be:

- Sellar - suprasellar (most cases), infrasellar or intrasellar.
- Prechiasmic.
- Retrochiasmic.

Rare locations include extradural and extracranial-nasopharyngeal, posterior fossa or extending down the cervical spine. These tumours are avascular on angiography and may encase or displace the vessels forming the circle of Willis. Craniopharyngiomas are histologically benign but may invade surrounding structures and can recur after an apparently total resection. Based on their histology, craniopharyngiomas can be further classified as adamantinomatous (paediatric type, cystic in nature, most common type), papillary (adult type, solid in nature) or mixed.

Epidemiology

- The overall incidence is 0.5-2.0 new cases/million population/year. Approximately 30-50% of all cases present in childhood.
- They account for about 3% of primary intracranial tumours overall, 5% of childhood intracranial tumours and 4.2% of all childhood tumours.
- There is bimodal age distribution with peaks occurring at 5-14 years and 65-74 years of age.
- There is possibly a slight preponderance in males.

Presentation

Symptoms and signs may be insidious and include those of a space-occupying lesion. The range of presentation and severity is broad. Typical clinical features at diagnosis include headache, visual impairment, polyuria/polydypsia, poor growth and significant weight gain.

- Endocrine dysfunction:
  - Hypothyroidism, adrenal failure and diabetes insipidus.
  - There may be panhypopituitarism and obesity.
  - Other endocrine features include decreased sexual drive and impotence. Most women complain of amenorrhoea.
  - There may be delayed or precocious puberty and growth failure occurs in most young people.

- Headaches: typically slowly progressive, dull and positional.
- Visual disturbances:
  - Caused by compression of the optic nerve and/or chiasma.
  - Includes bitemporal inferior quadrantanopia progressing to bitemporal hemianopia and optic atrophy.
  - Children rarely become aware of visual symptoms until advanced and irreversible damage has occurred.
Neuropsychological problems:

- Mild-to-severe dementia (due to frontal expansion) may occur.
- Other psycho-behavioural manifestations include hyperphagia and obesity, psychomotor delay, emotional immaturity, apathy, short-term memory deficits and incontinence.

- Features of hydrocephalus: caused by obstruction of CSF outflow at the third ventricle, resulting in headache, papilloedema and visual impairment.

Growth failure, delayed puberty and headaches are the most common presentations in children.

Differential diagnosis

- Other brain tumours.
- Other causes of intracranial space-occupying lesions.
- Other causes of hypothalamic pituitary dysfunction.
- Multiple sclerosis.
- Lyme disease.
- Tolosa-Hunt syndrome.
- Migraine.

Investigations

- Imaging:
  - Plain skull X-ray shows a calcified cyst in/above the pituitary fossa in 80% of cases. Such calcifications are more common in children (90%) than in adults (50%).
  - CT scan may reveal a lesion of mixed density in the suprasellar region containing both solid and cystic components. Administration of contrast enhances the cyst definition.
  - Magnetic resonance imaging (MRI) and magnetic resonance angiogram (MRA) clarify the precise position of the tumour with respect to the third ventricle and the relation of the major vessels to the tumour respectively.

- Biochemical: pituitary stimulation tests to assess the need for pituitary hormone replacement therapy.
- The patient needs a full neuro-ophthalmological evaluation with formal documentation of the visual fields.
- Some patients may also benefit from a psychiatric assessment.

Management

Non-aggressive surgery followed by radiotherapy is currently the most widely used management option. The management of choice for patients with favourable tumour localisation is complete resection but ensuring maintenance of the optic nerve and hypothalamic-pituitary axes.

Surgery

- Treatment for most craniopharyngiomas is surgical but this is associated with significant postoperative morbidity and mortality and recurrence is very common.
- An endonasal extended endoscopic approach may provide an alternative to transcranial surgery for some patients with suprasellar craniopharyngiomas.
- Total tumour excision followed by radiotherapy: the main risk is removal of the hypothalamus. Although recurrence rates are low, there is a mortality rate of up to 10%.
- Partial removal of tumour followed by radiotherapy is associated with a recurrence rate of up to 50% (much higher without radiotherapy).
- Drainage of the cystic areas followed by either external irradiation or implantation of radioactive yttrium-90.

Intracystic treatments

- Systemic chemotherapy does not work but intracystic chemotherapy has been performed with reasonable outcomes and minimal side-effects. Intracystic interferon alfa and bleomycin have been shown to be effective.
Brachytherapy and radioisotopes are recommended for solitary cystic craniopharyngiomas. Stereotactic aspiration of the cystic content is followed by instillation of a beta-emitting isotope. Intracystic phosphorus-32 (P-32) can be effective in controlling cystic components of craniopharyngiomas as a primary treatment or after other treatments but progression of solid tumour components often occurs.\cite{12} Other radioisotopes used include rhenium 186, gold 198 and yttrium-90.

Radiotherapy

- Radiotherapy is used following total or partial tumour excision.
- Stereotactic radiation (highly focused radiotherapy using three-dimensional imaging) has been used for further treatment of residual solid tumour after brachytherapy.
- Gamma knife radiotherapy for small tumours between the retrochiasm and anterior stalk. Although only useful in a select number of tumours, there are fewer risks with gamma knife radiotherapy - notably, no neuroendocrine sequelae.\cite{13}

There is research into the role of new systemic therapies (eg, interferon alfa-2a for progressive or recurrent craniopharyngiomas) which is showing some promising results.

Complications

- Related to the tumour: these relate to the effects of a space-occupying lesion.
- Related to the treatment:
  - Total resection is associated with a mortality rate as high as 10% (lower in adults). There may be significant ensuing psychosocial consequences if the hypothalamus is damaged.
  - Permanent endocrinopathy (eg, diabetes insipidus) occurs in up to 75% of adults and 93% of children. 80-90% of patients will need replacement of at least two anterior pituitary hormones.\cite{14}
  - Obesity occurs in over 50% of patients.\cite{2}
  - Other complications include:
    - Seizures.\cite{14}
    - Visual disturbances.
    - CSF leakage.
    - Herpes simplex encephalitis: there has also been a case report of this postoperatively.\cite{15}

- Recurrence occurs in up to 75% of patients after 2-5 years. Early intervention significantly minimises this risk, and tumour size also has a bearing on risk (20% if less than 5 cm and 83% if over 5 cm). Recurrences usually occur at the primary site; ectopic and metastatic recurrences are extremely rare.

Prognosis

- This depends on the exact nature of the tumour involved - histological type and location.
- The overall 10-year survival rate is between 60% and 90%.
- The outlook for younger patients is generally very good (reported to be between 69% and 99% survival at five years for those less than 20 years old).\cite{14}
- Older patients do less well (38% survival at five years for those over 65 years old, although there may be confounding factors such as unrelated comorbidity).
- Hypothalamic obesity is associated with increased mortality.\cite{16}

Further reading & references

- Paediatric Endocrine Tumours; British Society for Paediatric Endocrinology and Diabetes (2005)

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