Pituitary tumours

Tumours of the pituitary gland are almost always benign and are usually curable.[1] Pituitary tumours can cause problems by:

- Excessive hormone production
- Local effects of the tumour
- Inadequate hormone production by the remaining pituitary gland

Pituitary tumours may be associated with multiple endocrine neoplasia type 1 (MEN1), which is a rare hereditary endocrine cancer syndrome characterised by tumours of the parathyroid glands, endocrine gastroenteropancreatic tract (eg, gastrinomas, insulinomas and carcinoid tumours) and anterior pituitary - eg, prolactinomas.

Types of tumour

Pituitary tumours include (in decreasing order of frequency):

- Non-functioning adenomas
- Prolactinomas
- Growth hormone (GH)-secreting
- Adrenocorticotrophic hormone (ACTH)-secreting
- Thyroid-stimulating hormone (TSH)-secreting
- Leutinising hormone/follicle-stimulating hormone (LH/FSH)-secreting tumours

Hormone production

- The tumours that are often hormonally active are the eosinophilic GH-secreting adenomas, basophilic ACTH-secreting adenomas and prolactin-secreting adenomas. These tumours may protrude outside of the pituitary fossa (sella turcica):

    - ACTH-producing tumours: basophilic adenoma, presents with Cushing's disease. Enlargement of the tumour is usually slowly progressive. Initially confined to the sella turcica, but may enlarge and become invasive after bilateral adrenalectomy (Nelson's syndrome).
    - Prolactin-producing adenomas: usually intrasellar; are often small (less than 10 mm) but may become large enough to enlarge the sella turcica.
    - GH-producing tumours: eosinophilic - results in gigantism in children and acromegaly in adults. Suprasellar extension is not uncommon. Enlargement of the tumour is usually slowly progressive.

- Non-functioning tumours: they cause symptoms by extension beyond the sella, resulting in pressure on surrounding structures. In the absence of endocrine symptoms, visual loss is the usual initial manifestation.

How common are pituitary tumours? (Epidemiology) [1]

- Pituitary tumours account for 10% to 25% of all intracranial neoplasms.
- Adenomas account for the largest portion of pituitary neoplasms (estimated at approximately 17%). Only a minority of adenomas are symptomatic.
- Adenomas may invade the dura mater, cranial bone, or sphenoid sinus.
- Carcinomas account for 0.1% to 0.2% of all pituitary tumours.

- Prolactinoma is the most common pituitary adenoma in children. Non-functioning (non-secreting) pituitary adenomas are the second most common.[2]

Since neuroimaging techniques have improved, pituitary tumours are more often diagnosed incidentally. About 16.7% of the general population show changes in the pituitary gland.[3]

Pituitary tumour symptoms

Depends on the hormone secreted by the tumour as well as the pattern of growth of the tumour within the sella turcica.

- Local effects resulting from an expanding pituitary mass:
    - An expanding mass within the pituitary fossa may give rise to headache, neuro-ophthalmological defects or facial pain according to the size and direction of expansion:
        - Headaches: are classically retro-orbital or bitemporal. They tend to be worse on waking. Sudden catastrophic headaches may result from pituitary apoplexy. Very large pituitary tumours may cause obstruction of CSF, resulting in hydrocephalus and expansion of the lateral ventricles.
        - Visual field defects: these are common but often asymptomatic. Bitemporal hemianopia is the classic abnormality but any unilateral or bilateral visual field defect may occur.
        - Ocular nerve palsies cause a squint.
        - Extensive extension into the hypothalamus may result in disorders of appetite, thirst, temperature regulation and consciousness.

- Anterior pituitary hormonal deficiency:
    - Panhypopituitarism or varying degrees of loss of any of the six hormones may occur.
    - Hypopituitarism tends to occur in the following order of LH, GH, TSH, and lastly ACTH and FSH.
    - Therefore the presentation in adults tends to be infertility, oligo/amenorrhoea, decreased libido and erectile dysfunction. Deficiency of LH and GH may result in decreased muscle bulk, decreased body hair, central obesity and small, soft testes.
    - In children, hypopituitarism commonly presents with delayed puberty or impairment of growth.
    - Diabetes insipidus is rarely a presenting feature but may occur following surgery for a pituitary adenoma.
- Hypersecretion of the involved pituitary hormone - eg, acromegaly, hyperprolactinaemia, Cushing's disease, thyrotoxicosis.

Investigations

- Endocrine studies for hormone hyposecretion and hypersecretion. See also the separate article on Pituitary Function Tests.
- Lateral skull X-ray: may incidentally show enlargement of the fossa but is not a definitive investigation.
- Visual fields: common defects are upper-temporal quadrantanopia and bitemporal hemianopia.
- MRI scan is the preferred imaging investigation and is superior to CT scanning.[3]

Differential diagnosis

- Other neoplasms of the sellar region include craniopharyngiomas, Rathke's cleft cysts, and, less commonly, meningiomas, germinomas, and hamartomas.[4]

- Craniopharyngiomas are benign, cystic tumours found above the sella turcica. They present with headaches, visual field defects and hypopituitarism (including growth failure, as often present in childhood and adolescence).
- Other causes of headache, visual field defects, visual disturbance and endocrine dysfunction.

Pituitary tumour treatment

Treatment depends on the type of pituitary tumour and whether it extends into the brain around the pituitary. Hormone-secreting tumours can be treated by surgery, radiation therapy or by drugs such as bromocriptine (prolactin-secreting adenomas) or somatostatin analogues (GH-secreting adenomas). Small non-functioning adenomas and prolactinomas in asymptomatic patients do not require immediate intervention and can be observed.[5]

Surgery

Trans-sphenoidal surgery is the usual treatment of choice for lesions confined within the sella turcica and ACTH-secreting adenomas. Frontal craniotomy is rarely required. Lesions extending beyond the confines of the pituitary are most frequently non-functioning chromophobe adenomas and require additional radiation therapy. Rapid deterioration of vision is an immediate indication for surgery.

Postoperative pituitary dysfunction may include adrenal insufficiency, diabetes insipidus, syndrome of inappropriate antidiuretic hormone, and cerebral salt wasting syndrome. Neurosurgical complications may include visual disturbance, cerebrospinal fluid leak, subdural haematoma, and epistaxis.[6]

Radiotherapy

Radiotherapy is reserved for patients whose tumour has been incompletely resected or who remain hypersecretory after surgery.[7]

Somatostatin analogues

See also the separate article on Acromegaly.

Bromocriptine

Drug therapy with bromocriptine has been used with success in patients with prolactin-secreting tumours. See also the separate article on Hyperprolactinaemia and Prolactinoma.

Recurrent pituitary tumours

- Patients who develop recurrence following surgical resection can be treated with radiation therapy.[1]
- Re-irradiation of recurrent pituitary adenomas in selected patients is reported to have achieved long-term local control with improvement or stabilisation of visual symptoms.[1]

Complications

Pituitary apoplexy - sudden-onset hypopituitarism caused by an acute infarction of a pituitary adenoma. See the separate article on Acute Pituitary Failure.

Prognosis

Remission can be obtained in up to 90% of patients with microadenomas and in about 50% to 60% of those with macroadenomas.[1]

Authored by:	Peer Reviewed by: Dr Colin Tidy, MRCGP	
Originally Published: 20/11/2023	Next review date: 22/01/2023	Document ID: doc_2615