

The OHSU Pituitary Unit

Pituitary Tumors and Diseases - A Basic Overview

As discussed in the prior section, the normal pituitary gland releases many hormones necessary for maintaining life. The maintenance of proper pituitary hormone levels is achieved through a delicate mechanism of hormonal feedback from each target organ.

It is therefore not surprising that in the diseased pituitary, this finely tuned control mechanism can be lost and can lead to significant hormonal dysregulation making the individual feel very sick.

Pituitary Tumors

Benign tumors, called adenomas, are the most common cause of pituitary disorders. When thinking about the impact of pituitary tumors on a patient, it is helpful to ask three questions about different kinds of problems:

- Is the tumor causing damage of the normal pituitary leading to pituitary hormone deficiencies?
- Is the tumor producing excessive hormone levels itself?
- Is the tumor causing problems because of its size (headaches, visual problems, cranial nerve problems)? These are often referred to as "mass effects" of the tumor.

Tumors can cause one, two, or all three of these problems.

Non-functioning Adenomas

About 40% of all pituitary adenomas are non-functioning (i.e., don't secrete any hormones). Small "microadenomas" (2-3 mm or about the size of a pepper corn) are common in the general population (about 10% of people have them) and are generally considered to be clinically insignificant. However, these lesions can sometimes grow and cause problems. When pituitary adenomas are greater than 1 cm in size (about the size of a grape) they are referred to as "macroadenomas." Macroadenomas are often discovered when they are 1.5 to 2.5 because of the "mass effects" that they cause. These are as follows:

- **Pituitary hormone deficiencies** may be caused by a growing adenoma as it presses on and damages normal pituitary tissue. The signs and symptoms of these deficiencies can be varied because different combinations of pituitary hormones may be affected. Patients with pituitary adenomas should be evaluated for pituitary hormone deficiencies as discussed in the section: [How Do I Work With My Doctor To Find Out if I have a Pituitary Disorder?](#) Pituitary hormone replacement therapy is discussed in the section: [Pituitary Hormone Deficiency and Replacement](#). Pituitary hormone deficits are rarely reversed completely with tumor removal and often require long-term replacement therapy.
- **Headaches** from the "mass effects" of large tumors are probably due to stretching of the dura (the covering of the brain) by the tumor itself. Removal of the pituitary lesion usually relieves the headaches.

- **Visual loss**, one of the most serious complications of enlarging pituitary tumors, typically (but not always) affects the peripheral vision at first. This happens because the upward growing pituitary tumor presses on the "optic chiasm" where fibers cross that carry information from the peripheral vision. Untreated, these disturbances will typically lead to blindness. Conversely, these visual problems may fully resolve if a tumor is removed while the visual disturbance is still minimal.
- **Cranial nerve disturbances** occasionally result from large pituitary tumors and, when present, typically cause problems with aiming of the eyes.

A sudden, intense headache, often described as "the worst headache of my life" can be the presenting sign of pituitary apoplexy (the dying of the center of a large tumor due to it outgrowing its blood supply).

Macroadenomas can typically be removed with few complications by a neurosurgeon experienced in this procedure. Protecting further visual loss is typically the main objective of surgically removing or debulking non-functional macroadenomas. After debulking, if residual tumor growth continues, radiation is typically used as adjunct treatment. More details on the process of selecting a qualified neurosurgeon are available in the section: [I Have Been Diagnosed with a Pituitary Tumor - What Should I Do Next?](#)

Functioning Adenomas

Functional adenomas over-produce one or more hormones and have characteristic sets of clinical features. There are three major types: Prolactinomas, ACTH secreting adenomas (Cushing's disease), and GH secreting adenomas (acromegaly). These diseases are extremely complex and are each individually discussed in separate sections. Click the quick links below to learn more about each disease.

Other types of functional adenomas occur as well. For example, TSH secreting pituitary adenomas are very unusual but do occur. Interestingly, they are not always associated with hyperthyroidism because the TSH may be dysfunctional. Like non-functional adenomas, "TSH-omas" are treated surgically. Of note, some "non-functioning" adenomas actually secrete inactive hormones or hormone components but these compounds are generally considered to be clinically insignificant. They are also treated surgically if they are growing or are causing "mass effects" due to their size.

Other Pituitary Diseases

Although adenomas (functional and non-functional) account for more than 80% of pituitary lesions, a variety of other causes occur as well. Many of these non-adenomatous lesions impact both the pituitary and hypothalamus. These have been divided below into surgically versus principally

medically treatable disorders. Unlike pituitary adenomas, many of the other "surgically treatable" pituitary lesions are aggressive and require both surgical debulking and irradiation.

Non-adenomatous Pituitary Lesions Requiring Surgery:

Craniopharyngioma are the most common cause of hypothalamic damage in children and young adults but occur in adults as well. These lesions arise from remnants of Rathke's pouch (an embryonic structure) and are typically cystic and partially calcified. They often cause hypothalamic damage (e.g., diabetes insipidus, dysregulation of temperature, water and eating, as well as abnormalities of behavior and consciousness). These lesions are typically hard and "sticky" and often need to be removed through a transcranial (through the skull) approach. Irradiation is often needed as additional therapy.

Meningiomas originate from arachnoidal cap cells and about 10% involve the pituitary and adjacent structures (i.e., third ventricle, cranial nerves and the optic chiasm.) These lesions are rarely metastatic. Complete removal of these tumors can be very difficult to perform without damaging other neurologic structures so debulking of the tumor around critical structures (e.g., optic chiasm) is often the goal of surgery. Radiotherapy is often an additional means of treatment.

Cystic lesions include Rathke's cleft cysts, colloid cysts, and arachnoid cysts. These lesions can be clinically significant causing headaches, optic chiasm compression and pituitary damage. Surgical resection is the treatment of choice when these lesions become symptomatic.

Uncommon pituitary tumors include gliomas, metastatic lesions, chordomas, hamartomas, germ cell tumors, and epidermoid cysts. Treatment includes a combination of surgical, radiological, and medical options.

Medically Treatable Non-Adenomatous Pituitary Disorders:

Empty Sella syndrome occurs when the arachnoid membrane (filled with CSF) herniates into the space normally occupied by the pituitary. This results in the pituitary being pushed against the wall of the sella turcica and can lead to pituitary damage and hormonal deficiencies. This is treated with pituitary hormone replacement.

Irradiation / post-surgery induced hypopituitarism may result from the treatment of other pituitary lesions such as pituitary tumors. Pituitary insufficiency from irradiation may take from years to decades to develop. Post-surgical hypopituitarism occurs more frequently in the hands of non-expert pituitary neurosurgeons. Treatment is with pituitary hormone replacement.

Infiltrative diseases commonly manifest with hypopituitarism and need to be treated with pituitary hormone replacement. Causes include Sarcoidosis, Hemochromatosis, and Histiocytosis X. These diseases also affect other organs in the body and need to be treated by appropriate specialists.

Other causes of hypopituitarism include head injury, immunologic, infectious, and genetic causes among others. Treatment includes pituitary hormone replacement as well as disease specific therapy.

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This section was written by William H. Ludlam, M.D., Ph.D. for: www.OHSUpituitary.com.

Email: pituitary@ohsu.edu