SAINT JOHN'S HEALTH CENTER
BRAIN TUMOR CENTER

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Craniopharyngioma

Introduction

Craniopharyngiomas are benign tumors that arise near the pituitary gland and pituitary stalk and are typically both cystic and solid in structure. They occur most commonly in childhood and adolescence and in later adult life, after age 50. They account for 10-15% of sellar and suprasellar tumors (tumors that occur in and above the pituitary gland) and 50-60% of sellar and suprasellar tumors in children. They are usually not discovered until they impinge upon important structures around them, and are frequently quite large (over 3 cm) when detected. Although they are benign (not malignant) tumors, these tumors tend to become adherent to structures around the pituitary gland and stalk, including the optic nerves, optic chiasm, intracranial arteries and the brain itself. They are thought to arise from remnants of the craniopharyngeal duct or Rathke's pouch which are developmental structures related to the primitive gut. These tumors are thought to be closely related to Rathke's Cleft Cysts (RCCs).

Symptoms

Craniopharyngiomas can cause a variety of symptoms depending upon their location. If the tumor compresses the pituitary stalk or gland, the tumor can cause partial or complete pituitary hormone deficiency which may lead to growth failure, delayed puberty, loss of normal menstrual function or sexual desire, increased sensitivity to cold, fatigue, constipation, dry skin, nausea, low blood pressure, and depression. Pituitary stalk compression can also cause diabetes insipidus (DI), and increase prolactin levels causing a milky discharge from the breast (galactohhrea). If the tumor compresses the optic chiasm or nerves, then visual loss can result. Involvement of the hypothalamus, an area at the base of the brain, may result in obesity, increased drowsiness and temperature regulation abnormalities. Other symptoms especially with larger tumors may include personality changes, headache, confusion, and vomiting.

Diagnosis

The best means of visualizing a craniopharyngioma is with a magnetic resonance imaging (MRI) scan of the pituitary region performed without and with contrast. Many craniopharyngiomas will also be well seen on a computer tomography (CT) scan especially since some are partially calcified (containing calcium deposits). A complete pituitary hormonal blood evaluation should also be performed. Other possible diagnoses to consider when a cystic mass is seen in the area of the pituitary include a cystic pituitary adenoma or an arachnoid cyst.

Treatment

The best initial treatment for a craniopharyngioma is surgical removal. The goal of surgery is to completely remove the tumor while improving vision and brain function and avoiding complications.

The great majority craniopharyngiomas can be removed by either an ***endonasal endoscopic*** *approach (through the nose) or* ***a supra-orbital eyebrow craniotomy****.* Because of their tendency to be adherent to the optic chiasm, other nerves and important blood vessels, a total removal is possible in only 50 – 60% of patients. With incomplete removal, stereotactic radiotherapy (SRT) or stereotactic radiosurgery (SRS), in which special equipment provides a precise dose of radiation directly to the tumor to prevent further growth, is often used. Because hormonal deficiencies can develop many years after radiation treatment, patients treated with radiation should have periodic hormonal evaluations throughout their lifetimes. Additionally, because of the tendency for craniopharyngiomas to recur, repeat MRIs or CT scans should be obtained at least every six months for the first 5 years after surgery or radiation therapy and then at least annually thereafter.

For a visual tour of endonasal and keyhole surgery for brain, pituitary & skull base tumors, visit our BTC YouTube channel: *www.youtube.com/user/BrainTumorCenter*