SAINT JOHN'S HEALTH CENTER
BRAIN TUMOR CENTER

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Acromegaly

Introduction

Acromegaly is caused by a growth hormone (GH) secreting pituitary adenoma in over 99% of cases. The problems associated with acromegaly include the effects of abnormally high GH and IGF-1 levels, and in some instances by the tumor compressing the normal pituitary gland and optic nerves. Untreated acromegaly is a serious endocrine condition that can cause dramatic bone and soft tissue changes and serious cardiovascular and metabolic problems. If the tumor develops before bone growth is completed in adolescence, gigantism results. Because of the serious changes resulting from GH excess, treatment is essential.

Symptoms

The most obvious changes of acromegaly are the external physical changes that typically include enlargement of the hands (increase in ring size) and feet (increased shoe size) as well as frontal bossing (enlargement of the forehead) and prognathism (jaw enlargement). There may also be development of an underbite, spreading teeth, an enlarging tongue, increased snoring and sleep apnea. Carpel tunnel syndrome and excessive sweating are also common. More serious problems can include development of hypertension, diabetes mellitus and an increased risk of colon cancer. With GH-secreting macroadenomas, there may be other problems of visual loss, headaches and problems associated with pituitary gland failure including fatigue, depression, impotence and loss of libido in men and menstrual irregularities and galactorrhea (milk discharge from the breast), in women.

Diagnosis

Acromegaly is diagnosed by documenting elevated levels of both GH and IGF-1. An oral glucose tolerance test (lack of suppression of GH to oral glucose administration) is often used to confirm excess GH production. Comparing old and recent photographs will often demonstrate dramatic changes in facial appearance. Following hormonal testing that confirms acromegaly, an MRI of the pituitary should be performed to confirm the presence of a pituitary adenoma.

Treatment

Endonasal Endoscopic Surgery: Endonasal transsphenoidal adenoma removal is considered first-line treatment for acromegaly; long-term remission is seen in 80-90% of patients with microadenomas and in 40-60% of patients with macroadenomas or invasive adenomas. In general, the higher the pre-operative GH level, the lower the chance for cure. Because of improved tumor visualization, the endonasal endoscopic approach is rapidly becoming the preferred method for removal of most pituitary adenomas, including GH-secreting adenomas. Cure (or long-term remission) of acromegaly is often not possible in patients with large or invasive macroadenomas. However, in such invasive tumors, removal of the great majority of the tumor can greatly improve problems associated with acromegaly (such as visual loss, pituitary gland dysfunction and headache) and typically improves hypertension, diabetes and soft tissue swelling. Additionally, such maximal tumor debulking improves the chances of achieving remission with medical therapy using lanreotide or octreotide.

Medical Therapy: For patients with persistent GH elevation after endonasal surgery, octreotide, lanreotide or pegvisomant treatments, SRS, or both are generally indicated. Octreotide (given three times a day by injection or by one monthly injection), or lanreotide (deep subcutaneous injections every 4 weeks) achieve long-term suppression of GH in about 70% of patients. Lanreotide and octreotide also cause tumor shrinkage in 30-50% of patients, and improve soft tissue swelling, headache, joint pains and sleep apnea. Preoperative use of octreotide may facilitate tumor removal and lessen risks of general anesthesia. Side effects may include loose stools, malabsorption, cholelithiasis (gall stones), and local pain at the injection site. Pegvisomant, a GH receptor antagonist, is also effective in lowering IGF-1 levels although it does cause an elevation in GH levels. Bromocriptine and cabergoline are "dopamine agonists" that lower GH secretion in about 15% of acromegalics, however, these medications are more effective in adenomas that co-secrete both GH and prolactin.

Radiosurgery (SRS) or Stereotactic Radiotherapy (SRT): For patients with uncontrolled acromegaly after surgery, SRS (one dose) or SRT (multiple doses), provide precise radiation directly to the tumor, are relatively effective in lowering GH and IGF-1 levels and stopping tumor growth. However, the lowering of GH and IGF-1 levels takes longer with SRT (average 7 years) compared to SRS (average 18 months). Pituitary gland failure often occurs in the years after SRS or SRT. Complications such as visual loss are rare with either SRS or SRT.

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*