Acromegaly

Acromegaly is an overproduction of growth hormone (GH) during adulthood (Copstead-Kirkhorn & Banasik, 2013).

Pathophysiology

GH production is increased during childhood and teen years in order to prepare and stimulate the body for growth. The level of GH decreases steadily throughout an adult's life at a rate of 50 percent every seven years (Melmed, 2013a). GH is dispersed in a pulse like fashion throughout the day with about 10 disbursements a day. In patients that have acromegaly this pattern is disrupted by an adenoma that produces a constant level GH throughout the day. For adults this stimulates growth but the growth plates have closed which makes the symptoms very different than a child with gigantism. The increase in GH production causes the liver to increase production of Insulin-like growth factor-1 (IGF-1). "The increase in production of GH/IGF-1 causes growth of connective tissue, intracellular swelling, and bony proliferation (McCance & Huether, 2014, p. 22)." The increase in GH/IGF-1 causes a hyperglycemic state due to a reduction in peripheral glucose uptake and an increase liver production of glucose by the liver. The pancreas responds by producing more insulin with an end result of insulin resistance and diabetes type 2. Other major organs affected by acromegaly are the heart and kidneys. Unrestricted Growth in the heart causes cardiomyopathy and all the associated symptoms. The kidneys increase reabsorption (https://en.wikipedia.org/wiki/Reabsorption) of phosphate causing a slight increase in phosphate levels (Melmed, 2013b).

Genetics

Early research is finding that there is likely a mutant gene that causes some cases of acromegaly.

Epidemiology

Acromegaly is a rare disease that affects an estimated 6 people per million a year (Melmed, 2013b). There are no nationwide statics collected making it difficult to collect accurate numbers for the United States. Some studies estimate that the number could be as high as 1000 per million (Melmed, 2014). These numbers are based on random screenings of IGF-1. The age of onset is usually in the early forties.

Disease described

Acromegaly is an over production of GH that stimulates the liver to produce IGF-1. This causes physical and metabolic changes. Physical changes include growth of soft tissue to include skin, cartilage, connective tissue, bone, viscera and epithelial tissue (Melmed, 2014). Progression of the disease is slow and steady with an average interval from onset to diagnoses being 12 years (Colao, Ferone, Marzullo, & Lombardi, 2004). This is usually caused by an adenoma of the anterior pituitary that secretes GH. In a few cases tumors are in remote locations such as the lungs, pancreas, hypothalamus, or adrenal glands (Melmed, 2014). These tumors either produce GH or GHRH.

Sign and Symptoms

(Copstead-Kirkhorn & Banasik, 2013), (Melmed, 2013b)

- severe snoring and sleep apnea in a non-obese patient
- enlarged hands with increased ring size
- increased foot size
- enlargement of the frontal sinus
- growth of the mandible causing an under bite,
- deepening of the voice secondary to vocal cord thickening and large tongue
- skin becomes thick
- hypertrophy of the soft tissue
- increased size of internal organs
- hyperthyroidism
- arthritis
- kyphosis
- peripheral neuropathies due nerve impingement
- cardiovascular disease
- headaches
- vision loss
- type 2 diabetes are possible associated findings

Diagnosis

Initial test for suspected acromegaly is a Serum level IGF-1 (Copstead-Kirkhorn & Banasik, 2013). If negative than the disease is ruled out. If the IGF-1 is elevated then an oral glucose tolerance test is performed along with measurement of GH levels (Melmed, 2014, para. 9). If inadequate suppression of GH then a MRI of the pituitary is

performed. If normal, a CT of the chest and abdomen is performed along with measurements of growth hormone-releasing hormone and the diagnosis is Extra-pituitary acromegaly. If the MRI is positive for mass or empty sella, the diagnosis is GH-secreting pituitary adenoma.

Treatment

The first line treatment is surgical removal of the tumor followed by with octreotide therapy to reduce the size of the remaining tumor and reverses many of the clinical manifestations (Copstead-Kirkhorn & Banasik, 2013), (McCance & Huether, 2014). It is important to note that most of the time the tumors are too big to remove completely. Radiation is utilized when the patient is not a surgical candidate or if the surgery is unable to remove most of the tumor.

Links

- http://endocrine.niddk.nih.gov/pubs/acro/acro.aspx
- http://www.pituitarysociety.org www.pituitarysociety.org
- http://www.pituitary.org.uk/content/view/36/47/
- http://www.nlm.nih.gov/medlineplus/pituitarydisorders.html
- http://www.hormone.org
- https://www.aace.com/files/acromegaly-guidelines.pdf

Related Articles

- Newly diagnosed acromegaly presenting with hypertriglyceridemia pancreatitis with normal amylase and lipase levels. Located at: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3862123/
- Neurocognitive function in acromegaly after surgical resection of GH-secreting adenoma versus naïve acromegaly. Located at: http://www.ncbi.nlm.nih.gov/pubmed/23593161

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