Brief Communication

Pituitary Gigantism: A Case Report

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ABSTRACT

Objective: To present a rare case of gigantism. **Case Report:** A 25-year-old lady presented with increased statural growth and enlarged body parts noticed since the age of 14 years, primary amenorrhea, and frontal headache for the last 2 years.She has also been suffering from non-inflammatory low back pain with progressive kyphosis and pain in the knees, ankles, and elbows for the last 5 years. There was no history of visual disturbance, vomiting, galactorrhoea, cold intolerance. She had no siblings. Family history was non-contributory.Blood pressure was normal. Height 221 cm, weight 138 kg,body mass index (BMI)28. There was coarsening of facial features along with frontal bossing and prognathism, large hands and feet, and small goitre. Patient had severe kyphosis and osteoarthritis of knees. Confrontation perimetry suggested bitemporal hemianopia. Breast and pubic hair were of Tanner stage 1. Serum insulin like growth factor-1 (IGF1) was 703 ng/ml with all glucose suppressedgrowth hormone (GH)values of >40 ng/ml. Prolactin was 174 ng/ml. Basal serum Lutenising Hormone (LH), follicle stimulating Hormone (FSH) was low. Oral glucose tolerance test (OGTT), liver and renal function tests, basal cortisol and thyroid profile, Calcium, phosphorus and Intact Parathyroid hormone (iPTH) were normal.Computed tomographyscan of brain showed large pituitary macroadenoma. Automated perimetry confirmed bitemporal hemianopia. A diagnosis of gigantism due to GH secreting pituitary macroadenoma with hypogonadotrophichypogonadism was made. Debulking pituitary surgery followed by somatostatin analogue therapy with gonadal steroid replacement had been planned, but the patient refused further treatment.

Key words: Pituitary, gigantism

Introduction

Gigantism refers to a condition characterized by extreme physical size and stature. By definition, this originates during infancy, childhood or adolescence, when epiphyseal growth plates remain open.Although the term gigantism may be applied to a number of non-hormonally mediated overgrowth conditions in children, it is often used to specifically denote growth hormone(GH) excess. GH excess during childhood and adolescence is extremely rare, with the total number of reported cases thus far numbering only in the hundreds.Herein we present such a case.

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Case Report

This 25-year-old lady,hailing from a rural area of West Bengal, presented to us with history of increased statural growth and enlarged body parts noticed since the age of 14 years, primary amenorrhea, and frontal headache for the last 2 years.She has also been suffering from non-inflammatory low back pain with progressive kyphosis and pain in the knees, ankles and elbows for the last 5 years. At the time of presentation, she was unable to walk without assistance. There was no history of visual disturbance, vomiting, galactorrhoea, cold intolerance, acroparaesthesia, overt gastrointestinal bleeding, or snoring. She had no siblings. There was no history of similar illness in the family.

On examination, vitals were stable with normal blood pressure. Height was 221 cm; weight was 138 kg with BMI of 28. There was coarsening of facial features along with frontal bossing and prognathism. She had large hands and feet [Figures 1-3]. Her voice was deep. There was a small goitre. Patient could not stand straight due to severe

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kyphosis and osteoarthritis of knees with limited joint mobility. Osteoarthritic changes were also present in the small joints of the hand.Confrontation perimetry suggested bitemporal hemianopia. There was no visceromegaly. Breast was Tanner stage 1, and there was no axillary or pubic hair.

Laboratory investigation showed OGTT of 136 mg/dl with normal liver and renal function test. Serum IGF1 was 703 ng/ml with all glucose suppressed GH values of >40 ng/ml. Prolactin was 174 ng/ml. Basal cortisol and thyroid profile were normal. Pooled sera showedFSH of 1.05 and LH of 0.272 mIU/ml. Serum calcium, and phosphorus were normal. iPTH was 16.7 pg/ml. Hemoglobin was 11.1 g/dl.Computed tomographyscan of brain showed large pituitary macroadenoma. Magnetic resonance imaging (MRI) could not be done as the patient could not be negotiated within the MRI machine. Automated perimetry confirmed bitemporal hemianopia. ultrasound (USG) showed small, prepubertaluterine volume (1.5 ml). X-rays showed osteoarthritic changes of spine, knees, and small joints of the hand. Epiphyses were seen to be fused in X-rays of hands and knees.

A diagnosis of gigantism due to GH secreting pituitary macroadenoma with hypogonadotrophichypogonadism was made. Debulkingpituitary surgery followed by somatostatinanalogue therapy with gonadal steroid replacement had been planned, but the patient refused further treatment.

Discussion

Gigantism is an extremely rare condition caused by hypersecretion of GH.In 1909, when Harvey Cushing examined the skeleton of an Irish patient who lived from 1761-1783, he noted an enlarged pituitary fossa.[1] Subsequently it has been observed that GH hypersecretion in childhood is usually caused by pituitary adenoma or hyperplasia.[2] Gigantism differs from the acromegaly by the time of onset, i.e. before and after epiphyseal fusion respectively. This condition may occur sporadically or may show familial predilection. Extreme tall stature is very rare nowadays because of early presentation and treatment. Acromegaloid features may develop in older children.[3] The most consistent biochemical abnormality observed in patients with gigantism is an elevated IGF-1. The gold standard for making the diagnosis of GH excess relies on the inability to suppress serum GH to an appropriate level (usually <1 ng/ml) following 75 g oral glucose load. Hyperprolactinemia is also commonly seen, which is usually due to co-secretion by mammosomatotroph.[4]All these features were present in our patient. Other pituitary cell lines may or may not be affected. Our patient showed features of hypogonatophichypogonadism. Due to delay in epiphyseal fusion, some patient with hypogonadism may continue growing until twenties or thirties. Epiphyseal fusion was

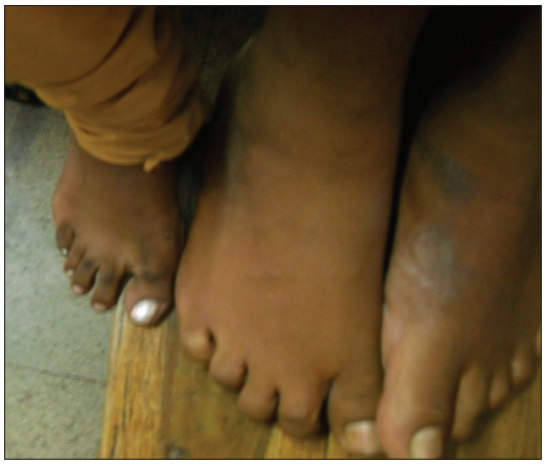

Figure 1: Large feet (comparison with normal adult female)

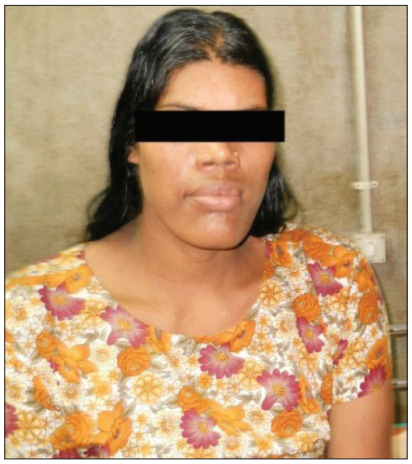

Figure 2: Facial characteristics

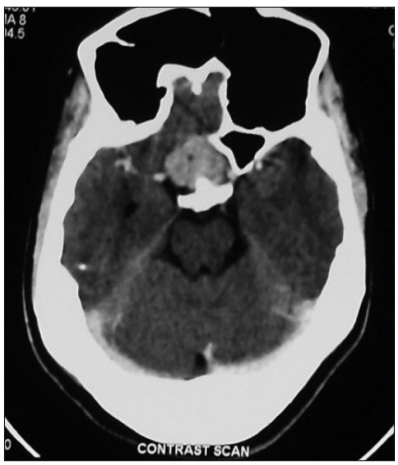

Figure 3: Contrast CT scan shows pituitary macroadenoma

noted in our patient at presentation. Pituitary imaging usually showedmacroadenoma. No large-scale studies evaluating various therapeutic approaches to the treatment of GH excess in pediatric patients are available. Therefore, the optimal treatment of gigantism has traditionally been extrapolated from the adult literature in association with anecdotal case reports detailing the experience of a few clinicians dealing with a handful of patients. Three separate modalities are available for the treatment of children and adolescents with GH hypersecretion, viz. surgery, radiation, and medical therapy. Trans-sphenoidal or trans-cranial surgery may be performed depending on tumor size and extension. Somatostatin analog with or without dopamine agonist has been used as medical therapy. As far as authors' knowledge, until date there is no experience with the use of pegvisomant for the treatment of gigantism in children or adolescents. Gonadal steroid may be used in children to promote early epiphyseal fusion. Radiotherapy is rarely used in children but may be used in adults.

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