ORIGINAL RESEARCH PAPER

Volume-8 | Issue-4 | April-2019 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ISOLATED GROWTH HORMONE DEFICIENCY- A CASE REPORT

Paediatrics

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ABSTRACT

Growth hormone has a well-defined role in promoting childhood growth and in maintaining normal adult body composition. Growth hormone is an anabolic hormone released in pulsatile manner in the circulation. It is one of the uncommon causes of short stature in children and is largely idiopathic.

The case report is about an 8 years old female child who presented with short stature. She was diagnosed to have Isolated Growth hormone deficiency. Growth hormone therapy was started after diagnosis was made. The child improved within 2 years of starting treatment.

KEYWORDS

INTROUCTION

Isolated growth hormone deficiency is a condition caused by a severe shortage or absence of growth hormone. Growth hormone is a protein that is necessary for the normal growth of the body's bones and tissues. Because they do not have enough of this hormone, people with isolated growth hormone deficiency commonly experience a failure to grow at the expected rate and have unusually short stature. This condition is usually apparent by early childhood.

There are four types of isolated growth hormone deficiency differentiated by the severity of the condition, the gene involved, and the inheritance pattern.

Isolated growth hormone deficiency type IA is caused by an absence of growth hormone and is the most severe of all the types. In people with type IA, growth failure is evident in infancy as affected babies are shorter than normal at birth.

People with isolated growth hormone deficiency type IB produce very low levels of growth hormone. As a result, type IB is characterized by short stature, but this growth failure is typically not as severe as in type IA. Growth failure in people with type IB is usually apparent in early to mid-childhood.

Individuals with isolated growth hormone deficiency type II have very low levels of growth hormone and short stature that varies in severity. Growth failure in these individuals is usually evident in early to mid-childhood. It is estimated that nearly half of the individuals with type II have underdevelopment of the pituitary gland (pituitary hypoplasia). The pituitary gland is located at the base of the brain and produces many hormones, including growth hormone.

Isolated growth hormone deficiency type III is similar to type II in that affected individuals have very low levels of growth hormone and short stature that varies in severity. Growth failure in type III is usually evident in early to mid-childhood. People with type III may also have a weakened immune system and are prone to frequent infections. They produce very few B cells, which are specialized white blood cells that help protect the body against infection (agammaglobulinemia).

Growth Hormone deficiency can be isolated or associated with pituitary disturbances[1]. Epidemiological studies suggest that idiopathic isolated growth hormone deficiency occurs more frequently than multiple pituitary hormone deficiency in children whereas in adult onset GH deficiency is frequently due to pituitary adenomas, surgeries and irradiation[2].

Synthetic human growth hormone (somatropin) shows improvement in children with isolated cases of growth hormone deficiency if started early provided that it should be administered under strict supervision.

CASE REPORT

An 8 years old female presented to our institute with history of not gaining height since 2 years. There was no history of perinatal insult, constitutional delay or history suggestive of any systemic causes of short stature. Birth weight and length were adequate and according to race. Dentition was normal. On examination the girl was alert with immature doll like facies, small hands and feet and pot belly (fig-1). Her vitals were stable and no abnormalities detected in systemic examination. There were no dysmorphic features. The anthropometric measurements showed weight 10.79 kg and height 80 cm. Her height for age was below 3rd percentile (CDC) and weight for age was also below 3rd percentiles (CDC) (fig-2). Its grade 3 stunting as per Waterlow classification. However her weight for height was between 90th and 95th percentile (CDC). The mid parental height was 148.5cms. Head circumference was 49 cm (within normal centiles). The ratio of upper and lower segment and arm span suggested being proportionate short stature. There were no secondary sexual characters with sexual maturity rating of P1B1. Investigations showed no abnormalities in routine blood, stool and urine examinations. The blood glucose, renal function test, liver function test and thyroid function tests were all normal. Chest X-ray was normal. Her bone age was normal (Bone age: 7 years) (fig-3).

Clonidine stimulation test (Growth hormone assay) and insulin tolerance test was done which was consistent with Growth hormone deficiency. Insulin like growth factor was below normal (29.47 ng/ml). ACTH, FSH, LH and prolactin levels where within normal limits MRI Brain showed hypoplasia of pituitary gland , 2mm (supero-inferior)*7mm (anteroposterior) * 8mm (transverse diameter), neurohypophysis normal in size and location. Parents were counseled about the condition. A diagnosis of Type II Growth hormone deficiency was made.

Injectable Growth hormone was given at 20 microgram/ m^2 /week (Body surface area-0.483m^2 as per Dubois formula). [1.47 units/ day= 0.49mg/ day, therefore 4 clicks/ day]. Regular follow-up was advised so that frequent blood glucose levels and thyroid hormonal assay could be assessed.

Fig 1 showing comparison of height in two females of same age

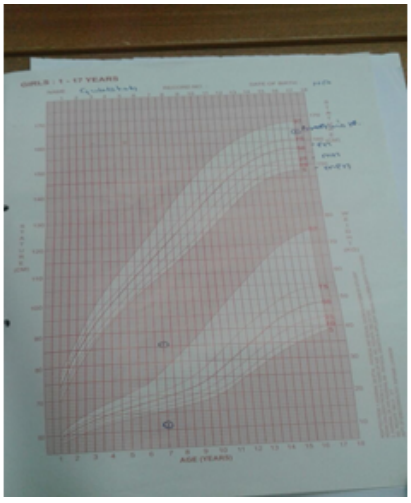
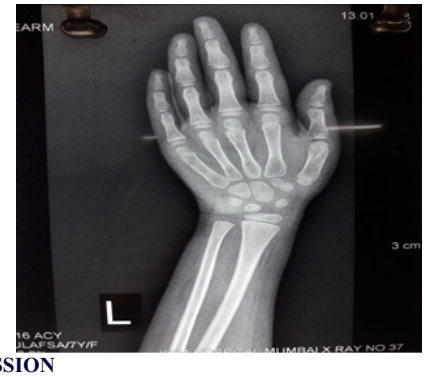


Fig 2 shows CDC charts for height and weight for age



DISCUSSION

Natural growth hormone is released in a pulsatile manner from the adenohypophysis spontaneously and in response to various physiologic stimuli [6]. Its production is stimulated by growth hormone-releasing factor and inhibited by somatostatin, which are both produced by the hypothalamus. Growth hormone binds to receptors on hepatic tissue and other cells, stimulating the production of insulin-like growth factor-I either locally or at the site of growing bone. Growth hormone binds to a specific growth hormone–binding protein (GHBP) and circulates. This GHBP is the extracellular portion of the growth hormone receptor. IGF-1 binds to one of several IGF-binding proteins (IGFBPs) and circulates almost entirely (>99%) in the bound state. IGFBP- 3 accounts for most of the IGF-I binding and this binding protein directly depends on growth hormone[5,7].

Growth hormone deficiency is an uncommon cause of short stature. In one study done in England and Wales, the approximate incidence of growth hormone deficiency was only 1 in every 30,000 births, about half of the patients had idiopathic deficiency and half had deficiency secondary to intracranial disease. These patients with low serum growth hormone levels associated with maternal deprivation were excluded[4]. Several cases apart from being idiopathic causes for a growth hormone deficiency could be due to CNS tumors including (cranio-pharyngiomas) and malformations, perinatal trauma, lack of oxygen at birth[3, 8], Septo-optic dysplasia[3, 7], leukemia, CNS trauma, CNS radiation, abnormalities in the hormone receptors and very rarely it may be due to a genetic defects, which in some instances may also be hereditary. Children with GHD usually present with short stature and a low growth velocity for age and pubertal stage. Alternative causes of poor growth needs to be considered and excluded. Age at presentation can vary from the first few months of life to adolescence. Typically the GH-deficient child has increased subcutaneous fat especially around the trunk. The face is immature with a prominent forehead and depressed midfacial development; this is related to the lack of GH effect on endochondral growth at the base of the skull, occiput, and the sphenoid bone. Dentition is delayed. In males the phallus may be small, and the average age of pubertal onset is delayed in both boys and girls[3]. Radiograph shows bone age lower than the chronological age. Growth hormone levels and binding protein levels (IGF-I and IGFBP-3) will show whether the growth problem is caused by a problem with the pituitary gland. MRI of the head can show the hypothalamus and pituitary glands[9,10]. Treatment with growth hormone will usually result in marked acceleration of linear growth. This is most pronounced in the first two years of therapy. In one study of more than 12,000 children, growth hormone replacement therapy was started at an average age of 9.2 ± 4.1 years and produced an increase in growth velocity from 4.4 cm per year to 10.0 cm per year. The younger the patient at the initiation of treatment and the more severe the growth deficiency, the better the response to early therapy[5].

Index Terms- Growth hormone, pituitary gland.
Conflict of Interest – None
Ethical considerations – Informed consent taken from parents of the patient regarding publishing the data.

Acknowledgements – All staff and residents involved in patient management and treatment. Also, the Dean Sir – Dr Chandanwale and Dr Nita Sutay (Prof and HOD, Pediatrics at Sir JJ Hospital, who permitted for the case report.

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