MAVEN e-ISSN: 2581-4443 | www.idahow.org

CASE REPORT

Pituitary Dwarfism: A Case Report

Dr. Harshvardhan Jha[1], Dr. Rupam Sinha[2], Dr. Soumyabrata Sarkar[3], Dr. Shubhadeep Maity[4], Dr. Ishita Banerjee[4] , Dr. Biyas Bhowmik[4]

[1]PG Student, [2]Professor and HOD, [3]Reader, [4]Senior Lecturer, Dept. of Oral Medicine and Radiology, Haldia Institute of Dental Sciences and Research, West Bengal.

Corresponding author: Dr. Harshvardhan Jha, PG Student, Dept. of Oral Medicine and Radiology, Haldia Institute of Dental Sciences and Research.
Email- harshvardhan.spartan@gmail.com

Abstract

Hypopituitarism describes combined deficiency of any of the anterior pituitary hormones. The clinical features vary and are based upon the underlying hormonal deficiency. Growth hormone deficiency is the prime cause of dwarfism due to pituitary insufficiency. The pa- tient has a short stature, limited intelligence, truncal obesity and certain orofacial manifes- tations like retained deciduous teeth, delayed eruption. This article reports a case of pitui- tary dwarfism in a female patient of 18 years and discusses its clinical findings. (2018, Vol. 02; Issue 02: Page 26 - 31)

Key words: Hypopituitarism, anterior pituitary, Growth hormone, Truncal obesity, Short stature.

Introduction

Hypopituitarism is an endocrinal disorder involving the hormones of anterior pitui- tary. Different hormone deficiencies can lead to different clinical features (1). Growth hormone deficiency has mainly seen to have caused dwarfism. A patient with pituitary dwarfism shows short stat- ure, truncal obesity, coarse skin, loss of hair, mid face hypoplasia, depressed nasal bridge, the size of the limbs are altered, re- tained deciduous teeth, delayed eruption of permanent dentition. Other hormones like Thyrotropins, Vasopressin, Adreno- corticotrophic hormone (ACTH), Gonado- tropins are also low in many cases. Causes can range from trauma to the pi- tuitary gland, a tumour most commonly Craniopharyngioma near to the pituitary gland, absence of pituitary gland etc. but in most cases no underlying cause is found (1-3).

Case report

A female patient of age 18 years reported to the Department of Oral Medicine And Radiology, Haldia Institute of Dental Sci- ences And Research, Haldia ,West Bengal with a complain of pain in upper left back teeth region since 2 weeks. Pain was sud- den in onset, intermittent, mild, localised, non-radiating, dull throbbing type which aggravated upon taking cold beverages and upon chewing food and was relieved upon taking medications prescribed by a local practitioner.

The family history was non-contributory, the mother of the patient reported us that the patient has yet not attended puberty and no menstrual cycles were reported till date. She also reported to us that the pa- tient talks in a proper language, under- stands instructions, is irritable at times and doesn't go to school. Patient's past dental history was non-contributory.

Extraoral examination revealed different features like shunted physical growth, truncal obesity, brachycephalic head, de- pressed nasal bridge, thick negroid lips, hypotrichosis, oblique slant of both eyes, mid-face hypoplasia, proportionate upper to lower limb ratio, increased intercanthal distance, coarse facial and body skin (Fig 1A-D). Upon inspection intra orally, mul- tiple retained deciduous teeth were pre- sent namely 51, 54, 55, 61, 64, 65, 73, 74,

75, 83, 85. Also lingually erupted 31,32,41,42 were present. Class II caries involving pulp chamber was present in re- lation to 64 (Fig 2A & B).

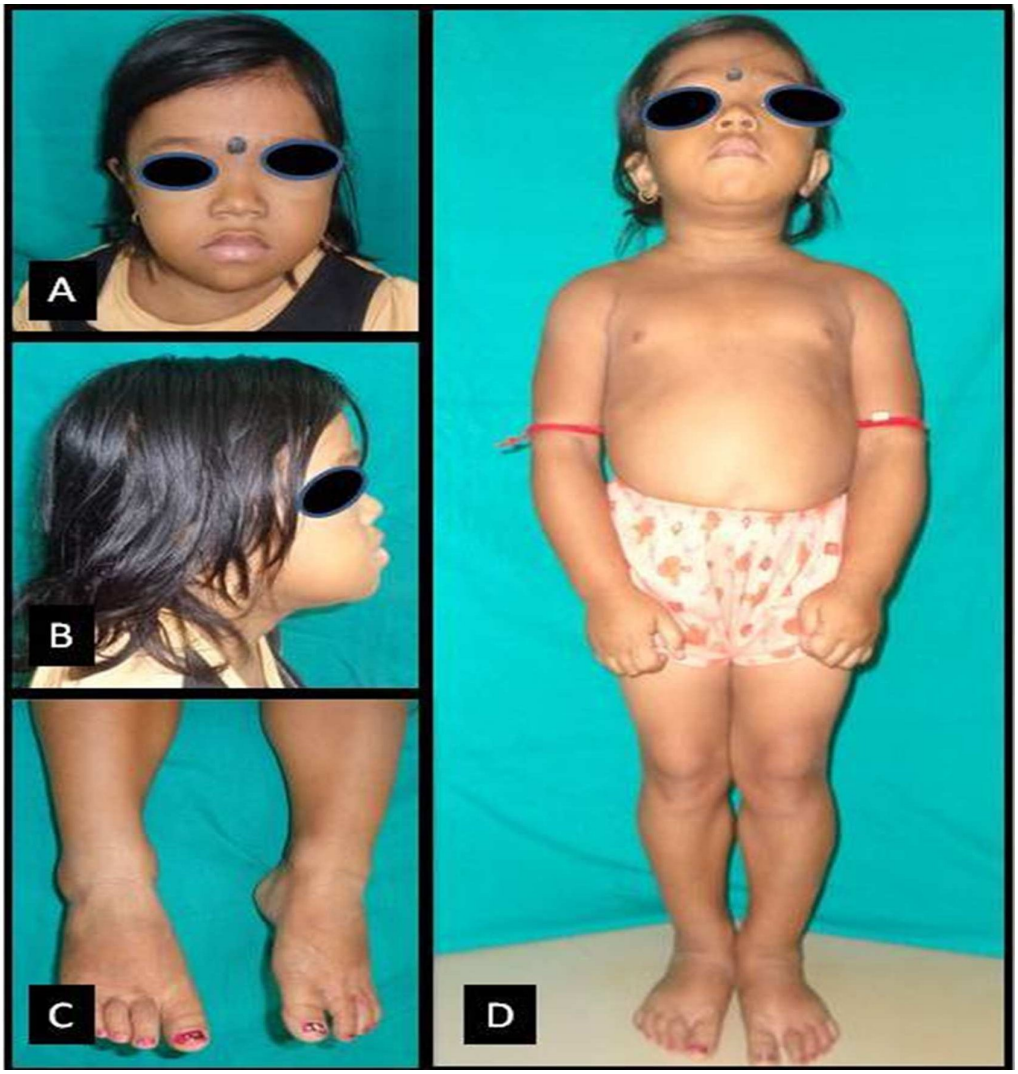

Fig 1: Extraoral view showing shunted physical growth, truncal obesity, brachycephalic head, depressed nasal bridge, hypotrichosis, mid-face hypoplasia, proportionate upper to lower limb ratio, increased intercanthal distance, coarse facial and body skin.

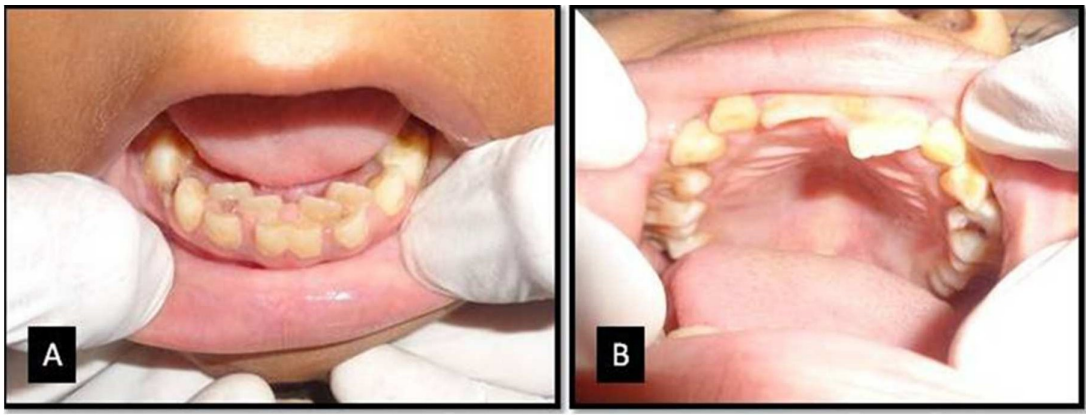

Fig 2: Intraoral view showing multiple retained deciduous teeth, lingually placed 31,32,41,42 and caries involving pulp chamber in relation to 64.

We advised the patient for an orthopantomograph, a PA skull view, a lateral skull view, a lateral arm view and an AP view of the leg.

The orthopantomograph shows multiple retained deciduous teeth namely 51, 54, 55, 61, 64, 65,73, 74, 75,83,85. It also showed us a diffuse radiolucency involving enamel, dentin and pulp in relation to 64. It showed that there was complete absence of tooth buds of 3rd molars in upper and lower arches (Fig 3).

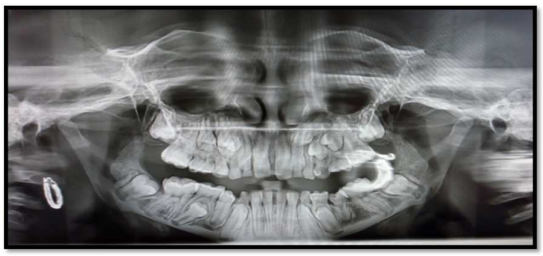

Fig 3: Orthopantomograph

The PA skull view was showing a brachycephalic skull (the skull appeared short and broad), also the anterior fontanelle was still persisting and has not fused yet. The lateral skull view also showed a short and broad brachycephalic skull along with a small sella (Fig 4A & B).

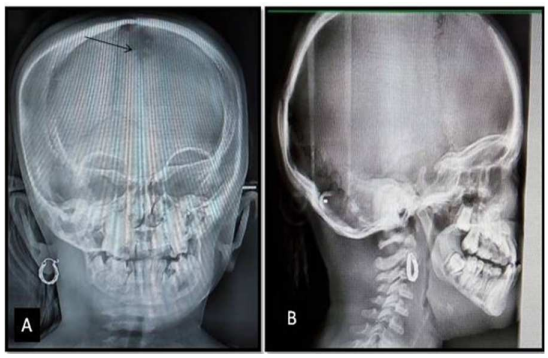

Fig 4: A- PA view of skull showing anterior fontanelle (black arrow), B- Lateral skull view showing broad brachycephalic skull with small sella.

The lateral view of the arm was showing bilateral erosion in the head of the Humerus and the shaft was shortened. The AP view of leg showed tibial metaphyseal flaring, fibula appeared elongated and the lower end of both femoral epiphysis appears distorted and sclerotic (Fig 5A & B) Upon viewing all the clinical features along with radiographic features, we gave a provisional diagnosis of Dwarfism with differential diagnosis of Achondroplasia, Cretinism.

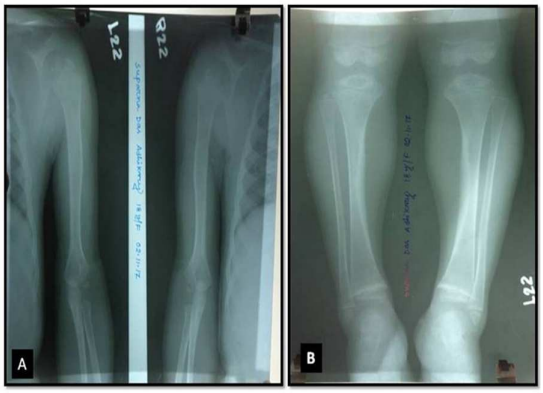

Fig 5: A- Lateral view of the arm showing bilateral erosion in the head of the Hu- merus, B- AP view of leg showing tibial metaphyseal flaring, fibula appeared elongated and the lower end of both fem- oral epiphysis appears distorted and scle- rotic.

Furthermore we send the patient for bio- chemical tests which included serum lev- els of T3, T4, TSH, FT4 and growth hor- mone assay. The reports came back and the values of T3 was 0.31ng/ml (reference range=0.60-1.81), T4 was 0.27mcg/dl (ref- erence range=4.50-10.90), FT4 was 0.19ng/dl (reference range=0.80-2.80), TSH was 96.50 mIU/ml (reference range=0.35-5.50). The value of growth hormone was 0.113ng/ml (reference range=0.0-8.0).

The biochemical reports clearly showed that the patient was having severe growth hormone deficiency along with deficiency of T3, T4 and FT4. All these data pointed towards hypopituitarism related dwarfism along with hypothyroidism.

The patient was first referred to the De- partment of Pedodontics and Preventive Dentistry Haldia Institute of Dental Sci- ences And Research for the treatment of class II caries in 64. Afterwards the patient was referred to an endocrinologist for fur- ther opinions and treatment.

Discussion

The pituitary gland, or hypophysis cerebri, is a reddish-grey, ovoid body, about 12 mm in transverse and 8 mm in anteropos- terior diameter, and with an average adult weight of 500 mg. The pituitary has two major parts, the neurohypophysis and ad- enohypophysis, which differ in their origin, structure and function. The neuro- hypophysis is a diencephalic downgrowth connected with the hypothalamus. The adenohypophysis is an ectodermal deriva- tive of the stomatodeum. Neurohypophy- sis includes the pars posterior (pars ner- vosa, posterior or neural lobe), infundibu- lar stem and median eminence. Adenohy- pophysis includes the pars anterior (pars distalis or glandularis), pars intermedia and pars tuberalis. The neurohormones stored in the main part of the neurohy- pophysis are vasopressin (antidiuretic hormone; ADH), which controls reabsorp- tion of water by renal tubules and oxyto- cin, which promotes the contraction of uterine smooth muscle in childbirth and the ejection of milk from the breast during lactation. Most of the hormones synthe- sized by the adenohypophysis are trophic. They include the peptides growth hormone (GH), involved in the control of body growth, and prolactin (PRL), which stimu- lates both growth of breast tissue and milk secretion. Glycoprotein trophic hormones are the large pro- opiomelanocortin pre- cursor of adrenocorticotrophin (ACTH), which controls the secretion of certain su- prarenal cortical hormones; thyroid-stim- ulating hormone (TSH); follicle-stimulat- ing hormone (FSH), which stimulates

growth and secretion of oestrogens in ovarian follicles and spermatogenesis (act- ing on testicular Sertoli cells); and lutein- izing hormone (LH), which induces proges- terone secretion by the corpus luteum and testosterone synthesis by Leydig cells in the testis (4, 5).

Pituitary dwarfism is may be caused by a deficiency of growth hormone or due to a lack of peripheral action of growth hor- mone. Lack of peripheral action of growth hormone can be due to different factors namely;inability of growth hormone to generate growth promoting substances like somatomedins and insulin like growth factors, also it may be due to unrespon- siveness of target tissues to such growth promoting factors (6).

The patients with such condition presents with short stature, proportionate ratio of upper and lower limbs, truncal obesity due to improper fat deposition, normal in- telligence, decreased amount of hair on the body resulting in a coarse skin, de- layed skeletal development, delayed sex- ual maturity (6, 7).

Delayed skeletal development can be shown by features like non fusion of the fontanels especially anterior fontanel at an adult age (8). There is presence of flaring of metaphyses and symmetrical shorten- ing of limbs (9). Studies have shown that hypopituitarism may result in small sized sella. Sometimes a J-shaped sella is seen as well (10). MRI can be useful to detect any tumour involving pituitary gland mainly Craniopharyngioma or any anom- alies of pituitary size and shape (9).

Studies have shown that variable orofa- cial findings are present as well namely hypoplasia of midface, crowding of teeth in both arches, numerous retained decidu- ous teeth, delayed eruption of permanent teeth, complete absence of buds of wisdom teeth even in 4th decade of life (11).

Biochemical values shows abnormal de- crease in serum growth hormone level, de- crease in insulin like growth factors (IGF- 1) level, hypothyroidism may also be pre- sent which is shown by a decrease in T3, T4, FT4 serum values along with in- creased serum TSH values. In some cases serum FSH and LH values are also de- creased showing delayed sexual develop- ment (1).

Treatment modalities include patient and family counselling, treating the underlying cause and symptomatic treatment (1, 2, 12).

In our case we had many features that co- relates to pituitary dwarfism mainly short stature, normal intelligence, delayed pu- berty and delayed menstrual onset, pro- portionate limb ratio, hypotrichosis along with a coarse skin, mid face hypoplasia, crowding of teeth in both upper and lower arches, multiple retained deciduous teeth, delayed eruption of permanent teeth, com- plete absence of tooth buds of all 3rd mo- lars, metaphyseal flaring in limbs, persis- tence of large anterior fontanel, small sized sella. Along with these features the biochemical investigation was suggestive of severe deficiency of growth hormone and hypothyroidism. These features prove that our patient was suffering from pitui- tary dwarfism.

Conclusion

Pituitary dwarfism is one of the rare cases of endocrinal field which involves both systemic and oro-facial findings. As a den- tist it is our prime job to diagnose such

cases at the earliest if it comes to us. A dentist can provide a firsthand counsel- ling to the patient and family and then can work in co-ordination with an endocrinol- ogist for the treatment of this multi feature disorder. Family and society too has an important role to play by cordially welcom- ing such patients to the normal living ways of the society rather than making them isolated. This disorder needs more and thorough research and studies for ad- vancement in the treatment modalities.

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