e-ISSN : 2455-5258

Research in Pharmacy and Health Sciences

Case Reports

Primary Amenorrhoea with Pituitary Dwarfism: A rare case report

Siva S*[1], Divya Gopineni[1], Shafi P[2], Chandra sekhar[2]

[1]Department of Pharmacy Practice, Sri Padmavathi School of Pharmacy, Tiruchanoor-517503, AP, India

[2]Department of Surgery and General Medicine, Sri Venkateswara Ramnarain Ruia Government General Hospital, Tirupati-517501, AP, India

Abstract:

Females with pituitary dwarfism and a multiple deficiency of pituitary hormones show ovarian dysfunction due to hypogonadotropism. Primary amenorrhea can be diagnosed if a patient has normal secondary sexual characteristics but no menarche by 16 years of age. A 16 year-old female patient admitted in general medicine department with chief complaints of shortness of breath on exertion since 15 days, swelling of both legs since 10 days, loss of weight since 5 months, loss of appetite since 3 months, history of pain during swallowing. Pelvis scan examination reveals that uterus measures 3.2×0.5×0.5cm; uterus is hypo plastic, ovaries not visualized. Patient parents reveled that from patient birth to 11years of age her growth and other developments were normal, after that her growth is stopped and no changes were observed in development since 5 years. Patient has hypothyroidism so pituitary gland make an important role to maintain hormone levels, pituitary gland produces thyroid stimulating hormone (TSH) which stimulates thyroid gland to produce thyroid hormones. Primary Amenorrhea, short stature and poorly developed secondary sexual characters which could have been contributed and should be subjected for karyotyping. This type of Pituitary Dwarfism is very difficult to manage.

Received: 10-02- 2016

Revised: 09-03-2016

Accepted: 03-04-2016

***Correspondence to:**
Dr. Siva S.
Email:
sivasanumala@gmail.com

Funding: Nil

Competing Interests: Nil

Keywords: Pituitary Dwarfism, Primary Amenorrhoea, Thyroid Stimulating Hormone, Hypogonadotropism.

INTRODUCTION:

Pituitary dwarfism is often complicated by disorders related to the reduced production of pituitary hormones. Females with pituitary dwarfism and a multiple deficiency of pituitary hormones show ovarian dysfunction due to hypogonadotropism.[1] Such patients would be expected to respond favorably to treatment with human gonadotropins. A thorough history and physical examination as well as laboratory testing can help narrow the differential diagnosis of amenorrhea.[2] Patients with primary amenorrhea, the presence or absence of sexual development should direct the evaluation, Constitutional delay of growth and puberty commonly causes primary amenorrhea. Primary amenorrhea can be diagnosed if a patient has normal secondary sexual characteristics but no menarche by 16 years of age.[3,4] If a patient has no secondary sexual characteristics and no menarche, primary amenorrhea can be diagnosed as early as 14 years of age.[5] The normal menstrual cycle involves a complex interaction between the hypothalamic pituitary- ovarian axis and the outflow tract.[6,7] Any disruption in this interaction can cause amenorrhea.[8] The treatment of primary and secondary amenorrhea is based on the causative factors that are Hyper prolactinemia, Empty sella syndrome, pituitary adenoma, enzymatic metabolism, liver failure, renal failure, gonadoblastoma, hypergonadotropic, hypogonadism, postmenopausal ovarian failure, premature ovarian failure, congenital androgen insensitivity syndrome and mullerian agenesis and others pregnancy, thyroid disease. [9,10]

CASE REPORT:

A 16 year-old female patient admitted in general medicine department with chief complaints of shortness of breath on exertion since 15 days, swelling of both legs since 10 days (Ruleout Echo), loss of weight since 5 months (Ruleout TB), loss of appetite since 3 months, history of pain during swallowing. On examination, her weight is 15kg and height 2.5inches, patient appeared as pallor. On laboratory examination, liver is in normal size and echotecture shows no focal lesions, CBD (common bile duct) and PV (portal vein) are normal and no IHBD (intra hepatic biliary dysplasia). Gallbladder wall thickness showed normal and no calculi observed, spleen and pancreas were normal in size and echotexture, Retro peritoneum aorta IVC (inferior vena cava) was normal. No lymphadenopathy observed. Both kidneys are normal in size, no calculi and no hydronephrosis observed. On examination uterus measures about 3.0×0.9×1.1cm, ET (Endometrial thickness) not appeared. After one month pelvis scan examination revealed that uterus measures 3.2×0.5×0.5cm without any infection, uterus was hypoplastic, ovaries not visualized. Laboratory examinations demonstrated FSH (follicle stimulating hormone) level of 2.02 mIU/mL (before puberty 0 - 4.0), LH (luteinizing hormone) of 0.12 mIU/mL (1.8 – 8.6mIU/mL), prolactin of 4.1 ng/mL (2 – 29ng/mL), and a free T4 of 2.5 ng/dL and HGH (human gonadotropine hormone) 6.11ng/ml were recorded. MRI (magnetic

regimen imagine), CT (computerized tomography) brain, USG (ultra sound sonography) abdomen were also performed. She was treated with B-complex and Iron Folic Acid for vitamin supplement, sucralfate for prevention of gastric irritation, furosemide for increasing urine output. Ampicillin 250mg, pantoprazole 40mg and injection human albumin were also given to the patient. After 2 months she was died with unknown reasons.

DISCUSSION:

Pituitary dwarfism is a condition of growth retardation char acterized by abnormally short stature with normal body proportions in which the pituitary gland does not make enough growth hormone. Primary amenorrhea is the absence of menstruation. Menstruation is a woman's monthly period, amenorrhoea is of two types primary amenorrhoea and secondary amenorrhoea. Primary amenorrhea is when a girl has not yet started her monthly periods; it can be diagnosed if a patient has normal secondary sexual characteristics but no menarche by 16 years of age. The patient has no secondary sexual characteristics and no menarche; primary amenorrhea can be diagnosed as early as 14 years of age. As this patient is not yet started her monthly periods, physicians confirm the diagnosis as pituitary dwarfism with primary amenorrhea. The treatment is given with furosemide as the patient having edema, B-complex as a vitamin supplement because patient is not taking sufficient food. While taking food she feels abdominal distention so for further supplement physician prescribe B-complex. IFA (iron and folic acid) is prescribed to this patient to improve hemoglobin levels as pallor and low hemoglobin levels are observed. Pantoprazole is prescribed to prevent gastric irritation, which may be due to ingestion of various medications. Sucralfate started on fifth day because patient feels abdominal discomfort and belching recurrently, Sucralfate acts as ulcer protective. After one month patient has not recovered so the treatment is changed with other category of drugs like Sucralfate replaced by aluminum and magnesium hydroxide syrup, B-complex replaced by B-complex, nicotinamide and pantothenate. Human albumin for fluid carrying protein, here patient serum Bilirubin concentration is 0.9mg% normal range of Bilirubin is <2mg%. The additional treatment includes depending on cause progestin therapy, MOPA, nor-ethindrone acetate, low dose of oral contraceptive pills, For gonadotropins treatment oral MOPA 1-12 days, low dose of oral contraceptive pills, anti-prolactin therapy, thyroid hormones and growth hormones.

CONCLUSION:

These types of patients needs early evaluation of hormones around 14years and should be diagnosed and treated early. Constitutional delay of growth and puberty commonly causes primary amenorrhea. Primary amenorrhea is an uncommon presentation in clinical medicine. Any symptoms of Primary Amenorrhea, short stature and poorly developed secondary sexual characters which could have been contributed and should be subjected for karyotyping. Karyotyping decides these critical regions and thus various congenital malformations can be observed for the chromosomal abnormalities. Dwarfism has been known throughout recorded history, but the relation of the pituitary to certain kinds of dwarfism has been recognized only recently. This type of Pituitary Dwarfism needs treatment with multiple hormones many continuous, some intermittent (Luteinizing hormone, Growth hormone) and infections may prevent due to lack of inflammatory effect of steroid hormones, so careful antibiotic selection and nutritional supplements need to be given.

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Cite this article as: Ayub M, Khurram S, Maqbool U, Malick A, Muneer F, Fatima SF, Bibi S. Role of Community Pharmacist in the prevention and management of Cardiovascular Diseases: A pilot study. Res Pharm Healt Sci.2016;2(2):145-147.