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Case Report

Van Wyk-Grumbach Syndrome: an unusual Presentation of long Standing Hypothyroidism with Early Menarche

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Abstract
Hypothyroidism is generally associated with delayed puberty, but long standing untreated hypothyroidism in young children can lead to pseudo-precocious puberty. Hypothyroidism induced precocious puberty presents with thelarche, menarche, galactorrhea, absence of sexual hair, delayed bone growth also known as van wyk-grumbach syndrome. Here we report an 8 years old girl presented to us with history of cyclical per vaginal bleeding since age of 3 years. Upon further workup there were bilateral polycystic ovaries on ultrasonography, upon subsequent investigations child was diagnosed with hypothyroidism.Due to lack of neonatal screeing for hypothyroisdim, lack of awareness among genral population such cases are missed and often presents late to healthcare pfofessionals.

Kew Words: hypothyroidism; van wyk-grumbach syndrome; galactorrhea

Introduction
This syndrome was first described by van wyk and grumbach in 1960 characterized by breast development, early menarche and multi cystic ovaries in the background of long standing hypothyroidism [1].The presumed hypothesis is that long standing hypothyroidism causes enlargement and hyper stimulation of pituitary gland which in turn causes hyper stimulation of ovaries, ovarian cysts and precocious puberty. Phenotypically, female patients with van wyk grumbach syndrome show breast enlargement, early onset of menstrual bleeding and enlarged multi cystic ovaries and delayed bone growth. The incidence of van wyk grumbach syndrome is reported to be more in females than males [2].the present case report presented a female child with features of van wyk grumbach syndrome. Furthermore a literature review was done regarding its treatment and its outcome.

Case presentation
A female child 8 years of age from Battagram presented to us at Gyne B deaptment of Ayub teaching hospital Abottabd Pakistan on 11 April 2018 with regular cycling per vaginal bleeding since the age of 3 years.she was born at full term, it was a normal vaginal delivery with no antepartum intrapartum or postpartum complications. There was no history of prolonged neonatal jaundice or delayed passage of meconium. Developmental millstones were normal till the age of 3.During her third year of life the child presented with premature menarche, ever since she goes through regular mensuration every month. The child bleeds for 5-6 days and the cycle repeats after every 30 days. Flow was moderate initially but from last one year menstrual flow was heavy. Examination the child was pale due to severe anemia, typical coarse features of the face can be appreciated, and skin was rough and dry. Excessive puffiness of eyes present. Patient gave history of constipation and cold intolerance. Breast development was tanner stage 2 without pubic or axillary hair development. Weight of the patient was 18kilograms(kg), height was 92 centimeter (36inches), FOC was 50centimeter (19inch) and upper segment was 51centimeter (21 inches).Intelligence of the child was average, the child belonged to a family of low socio economic status and there was no history of thyroid disorders or precocious puberty in other siblings.

Pelvic ultrasound findings were adult size uterus, bilaterally enlarged ovaries with multiple follicles of 12-28millimeter size and multiple cysts of 31-36millimeter sizes (fig;1). she had severe anemia with HB of 4.2g/dl(range 11.5-17.5g/dl).hormonal workup was done her serum lactate dehydrogenase(LDH) is 553I.U/L(208-378),serum Alpha-feto protein 5.2IU/L(< 6.7IU/L),serum BHCG less than 2mIU/ml(range <10),serum CA-125 29.30(range 2-30.2)all these tumor markers within normal limits. Her biochemistry revealed t3 level <40ng/dl (ref 58-159ng/dl), free-t4 levels were <0.40ng/dl (ref 0.70-1.48ng/dl),TSH level was > 100µIU/ml (range 25
100µIU/ml).Radiological examination of the arm and wrist bone revealed a bone age of 3 years (Figure 2)

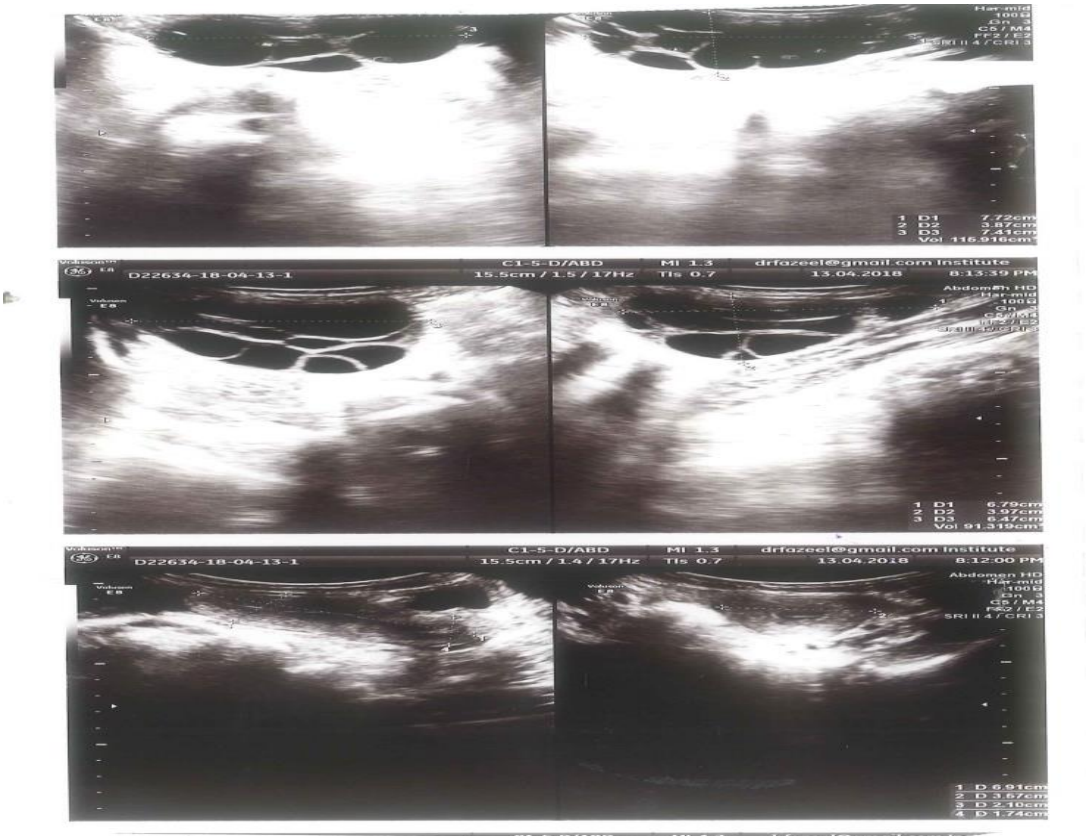

Figure 1

Figure 2

Discussion

Primary hypothyroidism usually causes retardation of linear bone growth and delay of puberty in juvenile patients but long standing untreated hypothyroidism leads to precocious puberty. Van wyk grumbach syndrome is characterized by juvenile hypothyroidism, delayed bone age, is sexual precocious puberty and the patient reverse to prepubertal state following thyroid replacement thearapy. [3] Some girls have irregular vaginal bleeding and solitary or multiple ovarian cysts [4] as evident in our case. The proposed theory explaining the etiology of this syndrome states that the low levels of T3 & T4 caused by primary hypothyroidism elicited a rise in TRH secretion which leads to an increase in levels of TSH, prolactin,and gonadotropins (LH and FSH),these glycoprotein hormones have a common alpha summit.it is presumed that a possible hormonal overlap exist resulting in excess secretion of not only TSH but LH and FSH as well [4].The exact pathophysiology is unclear but the most accepted theory states that high concentration of TSH are sufficient to cause activation of FSH receptor and produce gonadal enlargement. [5] The ovarian cysts are thought to be due to increased ovarian sensitivity to gonadotropins or action of increased levels of TSH on FSH receptors causing gonadal stimulation and thus cyst formation. [6]

Conclusion:

There is no concept of neonatal screening of hypothyroidism indeveloping countries like pakistan,due to which such cases are missed and mostly presents late at hospitals,Timely diagnosis of the above condition can lead to successful treatment of the condition and restoration of all hormonal abnormalities and can prevent unnecessary surgical intervention. Simple therapy with levothyroxine can reverse the symptoms and lead to restoration of normal bone growth.

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