

# VAN WYK Grumbach Syndrome with Puberty Menorrhagia and Ovarian Cyst - A Case Report

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**Abstract:** ***Aim:** To Report a case of Van WykGrumbach Syndrome a in the Government Maternity Hospital (Tertiary Health Care Center). **Materials and Methods:** A 12year old female from Piler presented to the emergency with h/o heavy menstrual bleeding since 7 days. **Conclusion:** Based on physical, biochemical and imaging studies. Patient was diagnosed with Van WykGrumbach Syndrome.*

**Keywords:** Van Wyk Grumbach Syndrome, Hypothyroidism, Heavy Menstrual Bleeding, precocious puberty

## 1. Introduction

Precocious puberty is a common gynaecological problem. Association of primary hypothyroidism with precocious puberty leads to hormonal overlap in hypothalamo-pituitary-ovarian axis. In 1960 Van Wyk and Grumbach first described a syndrome characterized by breast development, multi cystic ovary and uterine bleeding in the presence of long-standing primary hypothyroidism. Distinguishing feature of this syndrome from other causes of precocious puberty is the lack of pubic hair, short stature and delayed bone age. Laboratory investigation reveals very high level of Thyroid Stimulating Syndrome (TSH) and high level of Follicle Stimulating Hormone (FSH), Prolactin and 17- $\beta$  estradiol with suppressed Luteinizing Hormone (LH). Elevated TSH can act at the FSH receptor as TSH, FSH and LH share a common  $\beta$ -subunit. Thus, with high TSH level, FSH receptors of ovary are stimulated due to molecular mimicry producing high amount of estrogen which causes bilateral enlargement of ovary and onset of menarche

## 2. Case Report

A 12year old female present to emergency with h/o heavy menstrual bleeding since 7 days. On reviewing her history, no obvious findings were noted. She was born at 37 weeks of gestational age with birth weight of 2.8 kg. She attained menarche at the age of 11 years and had regular menstrual cycles, associated with normal flow. She had her regular menstrual period 10 days back with 3 days flow and the present complaint started 2 days after her regular period and continued for 7 days associated with generalized weakness, easy fatigability and breathlessness.

There was no family history of puberty menorrhagia, auto immune disease or any consanguineous marriage. Upon physical examination she had dull face, with peri orbital puffiness, grossly pallor +, Height was-120 cm, weight-30 kg, Pulse rate-52/min, BP-80/50 mmhg, thelarche stage-II, no axillary hair or pubic hair. Thyroid gland is not palpable. On laboratory work up, Hb-2 g/dl, TSH->50 mIU, T3-0.2 ng/ml, T4 < 15 ng/ml. LH-0.31 IU/L, FSH-10.1 IU/L, Prolactin-110 ng/ml, Estrogen-133 pg/ml, progesterone 0.04ng/ml. USG-multicystic ovaries over both sides-?

OHSS. On MRI brain-Pituitary macro adenoma size-1.4 x 2.0 x 1.3 cm. Patient was sent to SVIMS endocrinology I/v/o macro adenoma, there was diagnosed as VWGS. She was started on TAB. LEVOTHYROXINE 50 UG ALTERNATE DAY for 1 month f/b OD. After 6 months, USG revealed no cysts in the ovary, Thyroid levels were normal. Recent TSH is 2.680 mIU/ml. Now she has gained 5 cm height and MRI was normal.

## 3. Case Discussion

In our case, the cause of chronic hypothyroidism is autoimmune thyroiditis as evidenced by elevated thyroid peroxidase antibody. The TSH level was very high which was above 50 microIU/mL. High levels of TSH acts via the FSH receptor causing gonadal stimulation resulting in increased oestrogen production, which stimulates the development of secondary sexual characters. In the index case development of Tanner stage 2 as evidenced by breast enlargement and vaginal bleeding, is seen which is due to increased oestrogen production. Transition from Tanner stage I to Tanner stage II occurs for girls at a median age of 10.5 ( $\pm$  2) years [6]. But the child in index case had already acquired Tanner stage 2 at age of five. Since adrenal hormone synthesis is not increased; pubic and axillary hair is usually absent. VWGS is the only form of precocious puberty in which the bone age is delayed, and this is due to long standing hypothyroidism. Hyperproliferation in VWGS is seen due to either thyrotrophic hyperplasia in the pituitary that compresses the pituitary stalk and disrupts hypothalamic inhibition of prolactin or due to direct stimulation of prolactin release by Thyrotropin releasing hormone (TRH). In our case MRI of head showed macro adenoma.

It is essential to recognize VWGS through clinical and laboratory findings including the relationship between hypothyroidism and cystic ovarian enlargement, to avoid unnecessary surgery. This case of VWGS was diagnosed only after performing thyroid function test and other hormonal profile. Development of ovarian cyst is due to increased ovarian sensitivity to gonadotropins and myxedematous infiltration of the ovarian stroma resulting from hypothyroidism.

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#### 4. Conclusion

Early recognition and initiation of thyroid hormone replacement causes reversal of all symptoms with normalization of hormonal profile. This also avoids further diagnostic tests, fear of malignancy and unnecessary surgery. Regression of ovarian cyst and complete absence of vaginal bleeding along with normalization of thyroid function test was seen in our patient after six months of thyroxine replacement.

#### References

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