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A CASE OF VAN WYK GRUMBACH SYNDROME

Endocrinology

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ABSTRACT

Van Wyk - Grumbach syndrome (VWGS) is a rare complication of prolonged untreated juvenile hypothyroidism characterized by precocious puberty and enlarged multicystic ovaries. It results from hormonal overlap in negative feedback regulation with increased secretion of gonadotropins, prolactin, and thyroid stimulating hormone (TSH) as a consequence of the chronic hypothyroidism. A 13 yr old female patient presented with lower abdominal pain for 1 week, on palpation mass of 14*10cm size was noted occupying right and left iliac, lumbar and hypogastric region.USG abdomen and pelvis showed bilateral complex ovarian cysts; MRI brain showed bulky pituitary with microadenoma and TSH value >150 mIU/ml. She was started on replacement therapy with thyroxine after which ovarian cyst and pituitary microadenoma regressed.

KEYWORDS

Van Wynk Grumbach syndrome, Hypothyroidism, Prolactin, Ovarian cyst, Precocious puberty

INTRODUCTION:

Association of precocious puberty with primary hypothyroidism causes overlap in hypothalamo-pituitary-ovarian axis. Van Wynk Grumbach syndrome is described as syndrome characterised by development of breast, multicystic ovaries and uterine bleeding in presence of chronic primary hypothyroidism. It is differentiated from other causes of precocious puberty by absence of pubic hair, short stature. Laboratory investigations showed high levels of prolactin and thyroid stimulating hormone (TSH). As TSH, FSH and LH share common Beta subunit, elevated TSH acts at the FSH receptor. Therefore, FSH receptors of ovaries are stimulated producing large amount of estrogen causing bilateral enlargement of ovaries and early onset of menarche.

Case Study:

- A 13-year-old female child presented with lower abdominal pain for 1 week. She attained menarche at age of 8yrs. Her menstrual cycles were irregular, 7-9 days / 2-3 months. She was born out of non-consanguineous marriage, first in birth order. There is no family history of hypothyroidism / precocious puberty. Height - 135 cm (less than 3rd centile for age), weight 32kg (50th percentile for age). BMI -17.6 (underweight)
- On examination, she had pallor and dry skin. On palpation, mass of size 14* 10 cm noted occupying right and left iliac, lumbar and hypo gastric region, with smooth surface, cystic in consistency. Lower border couldn't be palpated separately. On percussion, dull note is heard over the mass.
- MENSTRUAL HISTORY-attained menarche at age of 8 yrs, Irregular,7-9/2-3 months, not associated with pain or clots.
- At the time of Admission: -vital data

Temperature-98.7f
BP-100/70mm hg
PR-83/min

Investigations:

- CBP – microcytic hypochromic anemia, Hb-7.3mg/dl
- Renal function tests and liver function tests-with in normal limits
- Thyroid profile – free T3 – 2.31 pg /ml, free T4 – 0.4 ng/dl, TSH > 150 m IU/ml
- Serum prolactin – 74 ng/ml
- FSH-10.6IU/ml, LH-0.1mIU/ml
- Tumor markers – with in normal limits
- USG abdomen and pelvis: bilateral complex ovarian cysts (right -9 * 9.4 * 8 cm, left -15* 9* 7.5 cm)
- MRI brain – bulky pituitary with micro adenoma

She was started on replacement therapy with thyroxine, after which the ovarian cysts and pituitary microadenoma regressed. TSH and serum prolactin returned to normal levels.

DISCUSSION:

High levels of TSH act on FSH receptors because of molecular similarity.This stimulation is supported by the specific FSH / estrogen dominant clinical picture like breast development, ovarian cysts and menstruation.Hyperprolactinemia is seen either due to pituitary microadenoma or due to direct stimulation of prolactin by TRH.Stimulation of gonadal FSH receptors by TSH is supported by FSH/estrogen dominant clinical features like menstruation, breast development and follicular cysts.High levels of TSH act via FSH receptors causing gonadal stimulation resulting in estrogen production there by development of secondary sexual characters.This case was diagnosed after performing thyroid function test and other hormones profile, Development of ovarian cyst is due increased sensitivity of ovaries to gonadotropins and myxedematous infilteration of ovarian stroma from hypothyroidism. Thus, the pathophysiology of VWG syndrome is complex which is mediated by action of TSH on FSH receptors. Regression of ovarian cyst with normalization of thyroid function test after thyroxine replacement.

CONCLUSION:

It is vital to keep this entity in consideration and; hence, should investigate for thyroid status during the evaluation of ovarian cysts. Thyroxin replacement after establishing the diagnosis early can prevent the patient from going through extensive workup and surgeries.

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