Primary Hypothyroidism Presenting as Bilateral Ovarian Torsion

Van Wyk–Grumbach Syndrome, first described in 1960, is characterized by isosexual precocious puberty, delayed bone age and bilateral multicystic ovaries in the background of primary hypothyroidism (secondary to autoimmune thyroiditis) [1,2]. The inherent pathophysiology involves the action of raised Thyroid-stimulating hormone (TSH) on Follicle-stimulating hormone (FSH) receptors, leading to the development of pseudoprecocity. These hormones share a common α-subunit which acts through similar transmembrane receptors [3]. Such cases may present with bilateral ovarian masses and end up with bilateral oophorectomy if the primary physician is unaware of underlying pathophysiology.

An 11-year-old girl presented to us with acute abdominal pain of 7 days duration, associated with vomiting and abdominal distension. The patient had achieved menarche 6 months ago. There were no neurological complaints. The patient had short stature, obesity, and features of peritonism and shock. Contrast-enhanced computed tomography scan showed two multiloculated adnexal masses. Serum α-fetoprotein and β-human chorionic gonadotropin levels were normal.

On surgical exploration, massive hemorrhagic ascites was revealed. Both ovaries were enlarged (15 cm on right and 11 cm on left) with multiloculated cysts. They had undergone torsion and were grossly gangrenous. Bilateral salpingo-oophorectomy was done. Histopathologic evaluation of the specimens revealed bilateral follicular cysts with surrounding edema with large areas of hemorrhage.

Three months post-operatively, the child complained of abnormal weight gain along with muscle weakness. Thyroid profile showed raised TSH level (>100 uIU/mL) with low free T3 and T4, normal FSH and LH and raised estradiol. On imaging, there was a delayed bone age. Patient was treated with thyroid hormone replacement and within 1 month, she improved symptomatically with a reduction in weight and improved muscle weakness. At present, she is 18-year-old with normal breast and feminine hair development, on oral estrogen and thyroid hormone replacement therapies, and is clinically and biochemically euthyroid.

Any child of isosexual precocious puberty presenting with bilateral ovarian masses should undergo bone age estimation, as delayed bone age in this context clinches the diagnosis of Van Wyk-Grumbach syndrome, which can further be confirmed by hormonal analysis. Initiation of thyroid hormone replacement results in improvement in endocrine abnormalities, final height achieved as well as resolution of ovarian masses, thus preserving ovarian reserve in the child [4]. Only in case of ovarian torsion and ischemia, surgery is warranted. In all other less acute cases, pharmacotherapy may be sufficient and avoids unnecessary surgery [5].

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REFERENCES