Precocious Puberty Due to an Autonomous Ovarian Cyst in a 3-Year-Old Girl: Case Report

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Abstract: Background: Precocious puberty is the occurrence of secondary sexual characteristics in girls before the age of 8. The diverse etiology of premature puberty is crucial to determine whether it is true precocious puberty, depending on the activation of the hypothalamic-pituitary-gonadal axis, or pseudo-precocious, which is independent of the activation of this axis. Whatever the cause, premature action of the sex hormones leads to the common symptoms of various forms of puberty. These include the development of sexual characteristics, acne, acceleration of growth rate and acceleration of skeletal maturation. Due to the possible genetic basis of the disorders, an interdisciplinary search for the cause is needed. Case report: The case report concerns a patient of a pediatric gynecology clinic who, at the age of two years, developed advanced thelarhe (M3) and started recurrent vaginal bleeding. In August 2019, gonadotropin suppression initially and after LHRH stimulation and high estradiol levels were reported at the Endocrinology Department. Imaging examinations showed a cyst in the right ovary projection. The bone age was six years. The entire clinical picture indicated pseudo- (peripheral) precocious in the course of ovarian autonomic cyst. In the follow-up ultrasound performed in September, the image of the cyst was stationary and normalization of estradiol levels and clinical symptoms was noted. In December 2019, cyst regression and normal gonadotropin and estradiol concentrations were found. In June 2020, white mucus tinged with blood on the underwear, without any other disturbing symptoms, was observed for several days. Two consecutive USG examinations carried out in the same month confirmed the change in the right ovary, the diameter of which was 25 mm with a very high level of estradiol. Germinal tumor markers were normal. On the Tanner scale, the patient scored M2P1. The labia and hymen had puberty features. The correct vaginal entrance was visible. Another active vaginal bleeding occurred in the first week of July 2020. The considered laparoscopic treatment was abandoned due to the lack of oncological indications. Treatment with Tamoxifen was recommended in July 2020. In the initiating period of treatment, no maturation progression, and even reduction of symptoms, no acceleration of growth and a marked reduction in the size of the cysts were noted. There was no bleeding. After the size of the cyst and hormonal activity increased again, the treatment was changed to Anastrozole, the effect of which led to a reduction in the size of the cyst. Conclusions: The entire clinical picture indicates alleged (peripheral) puberty. Premature puberty in girls, which is manifested as enlarged mammary glands with high levels of estrogens secreted by autonomic ovarian cysts and prepubertal levels of gonadotropins, may indicate McCune-Albright syndrome. Vaginal bleeding may also occur in this syndrome. Cancellation of surgical treatment of the cyst made it impossible to perform a molecular test that would allow to confirm the diagnosis. Taking into account the fact that cysts are often one of the first symptoms of McCune-Albrigt syndrome, it is important to remember about multidisciplinary care for the patient and careful search for skin and bone changes or other hormonal disorders.

Keywords: McCune Albrigth's syndrome, ovarian cyst, pediatric gynaecology, precocious puberty

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