Case Report

Ovarian mucinous cystadenoma in a female with Turner syndrome

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ABSTRACT

The development of an epithelial tumor, especially mucinous type, in a female with a streak gonad is rare and not fully understood. We report a case of a 19-year-old; a single female known to have Turner syndrome presented with an increased abdominal girth and was found to have a huge pelvic and abdominal mass. Ultrasound and magnetic resonance imaging revealed a huge cystic ovarian mass with no ascites. Laparotomy and right oophorectomy were performed for the ovarian mass. Histology revealed a large mucinous cyst adenoma. Further study of these tumors may help to eludicate the underlying cause and pathogenesis.

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XY gonadal dysgenesis in phenotypic females, confers an increased predisposition to germ cell tumors in rudimentary streak gonads.[1] The description of gonadal neoplasms is confined predominantly to gonadoblastomas and dysgerminoma,[1] these tumors are occasionally identified in childhood, and the risk increases with age from an estimated 2% at age 10 years to 27.5% at age 30 years.[2] If Y chromosome material is identified, gonadectomy is the standard care.[3] In Turner syndrome without any Y chromosomal material, there is no increase in the chance of having an ovarian tumor. Ovarian epithelial tumors, the most common histological type in eugenetic ovaries, have been rarely reported in association with an intersex disorder.[1,4,5] We describe the finding of an epithelial tumor in a female with known Turner syndrome.

Case Report. The patient was a product of spontaneous vaginal delivery after a full term uneventful pregnancy. Her neonatal and childhood developments were unremarkable. At age 14, she was seen by a pediatric endocrinologist for her short stature and delayed puberty. There was no history of mumps infection or exposure to chemotherapy or irradiation. Her sister had menarche at the age of 10 years and there was no family history of premature menopause or autoimmune disease. Physical examination showed, short stature (135cm, weight 40kg), no dysmorphic features, no breast development (Tanner stage I), no pubic or axilary hair, and no other features of Turner syndrome. Her chest and heart were normal; there were no pelvic masses, ascities or organomegaly. Local pelvic inspection revealed normal external genitalia and patent vagina. Hormonal assays confirmed the diagnosis of primary ovarian failure with elevated serum follicle stimulating hormone of 71 iu/L and luteinizing hormone 24 iu/L with, low estradiol level of 90 pmol/L. Blood karyotyping confirmed the diagnosis of Turner syndrome 46, X, i(X) (q10).

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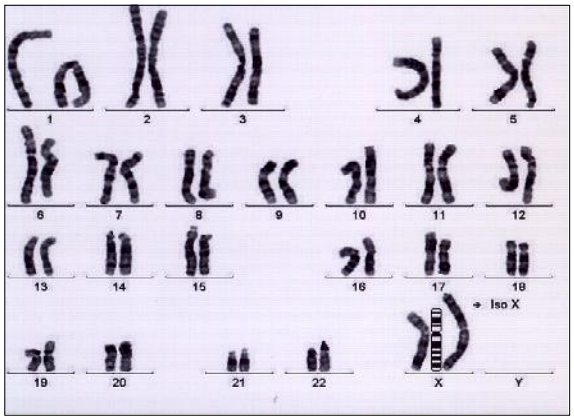
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