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 elucidate the underlying cause and pathogenesis.

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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 elucidate the underlying cause and pathogenesis.

**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 axillary hair, and no other features of Turner syndrome. Her chest and heart were normal; there were no pelvic masses, ascites 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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