

## CASE REPORTS

## Ovarian mucinous cystadenoma in a female with Turner syndrome

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XY gonadal dysgenesis in phenotypic females, confers an increased predisposition to germ cell tumors in rudimentary streak gonads.<sup>1</sup> The description of gonadal neoplasms is confined predominantly to gonadoblastomas and dysgerminoma,<sup>1</sup> 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.<sup>2</sup> If Y chromosome material is identified, gonadectomy is the standard care.<sup>3</sup> 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 eugenic ovaries, have been rarely reported in association with an intersex disorder.<sup>1,4,5</sup> 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 axillary 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). The chromosome length was 400-550 bands. Chromosomal karyotypes (Figure 1) were described according to an International System for Human Cytogenetic Nomenclature.

She was started on estrogen therapy as premarin 1.25 mg daily. She developed breasts and pubic and axillary hair. By the age of 16 years, she had a vaginal break through bleeding and she noticed an increase in her abdominal girth. She was seen by her physician, discovered to have a pelvic-abdominal mass and was referred to gynecology and oncology service. She was asymptomatic. On examination, she was generally well. Abdominal examination revealed a huge pelvic-abdominal mass up to the xiphisternum, which felt cystic and smooth. Ascites was not detected. Hemoglobin was 12 g/dl. Ovarian tumor markers, beta human chorionic gonadotropin, lactate dehydrogenase, Alpha-fetoprotein and CA 125 were within normal limits. Ultrasound showed a huge pelvic multilocular ovarian mass and normal small uterus. Pelvic magnetic resonance imaging revealed that the mass originated from the right ovary.

The patient was prepared for laparotomy through a mid-line incision. There was a large cystic right ovarian mass with an intact capsule, freely mobile, with a streak gonad on the left. Both fallopian tubes were normal and the uterus was small. Peritoneal washing was carried out for cytology. The mass was delivered outside the incision and right oophorectomy was carried out and sent for frozen section and reported as mucinous cyst adenoma, which was confirmed on final pathology.

On gross examination, the excised mass consists of a large cystic structure measuring 25 x 20 x 16 cm with smooth and transparent outer surface. Multilocular cut surfaces were evident on serial sectioning. It was filled with clear mucoid fluid focally adherent to the inner aspect of the cyst wall. No gross papillary projection

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