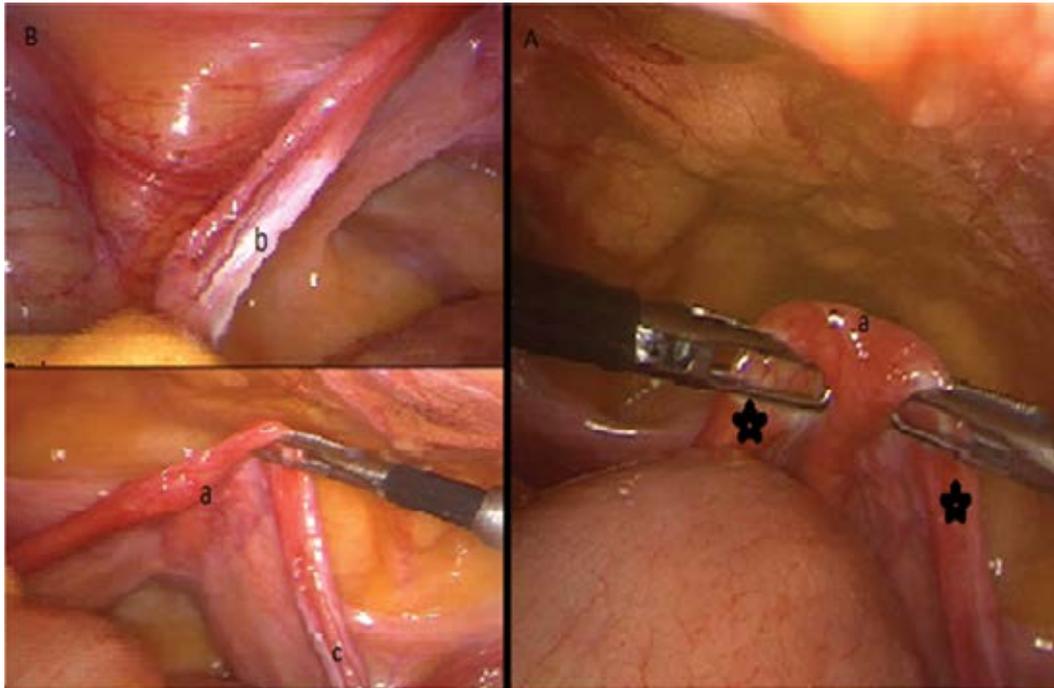


Case Blog

Title: 46, XY Disorder of Sex Development

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46, XY disorder of sex development (46, XY DSD) is an anatomoclinical entity due to a failure of testicular determination. At present 20% of disorders are explained by transformation or deletion in one of the five genes: SRY. Abdominal dysgenetic gonads are at increased risk for gonadal tumors (most commonly dysgerminoma) and should be surgically removed; streak gonads and dysgenetic gonads are at increased risk for gonadoblastoma and should be surgically removed. Typically, hormone replacement therapy is required from puberty onward.

We describe a case of a 22 years old patient presented to us with a chief complain of primary amenorrhea. Physical examination revealed completely female external genitalia with mild clitoral enlargement, breast development Tanner stage II, and height 157 cm. Echography and magnetic resonance imaging revealed an hypoplastic uterus and full vagina, while in the normal pelvic position of ovaries located gonadal streaks.

The chromosomal analysis revealed a 46, XY karyotype, and detected SRY gene. In addition exploratory laparotomy was performed in order to investigate the Mullerian and Wolffian structures, to evaluate gonadal tissue and to prevent future tumorigenesis (Figure 1). The exploration shows an hypoplastic uterus (a) Extended by structures like tubal (stars), they Continues by a white strip on the right (b) and a nodular formation on the left (c). Histological examination of the streak gonads tissue did not detect any signs of malignance. It shows normal ovarian tissue on the right, and normal epididymal tissue on the left.

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