A 5-year-old patient was brought by her parents to our pediatric endocrinology Outpatient clinic with history of progressive bilateral breast budding and enlargement since 3 months ago. Her previous medical history were uneventful; there was no family history of precocious puberty. Parents were married, nonconsanguineous, she has 1 other sibling who is well. At presentation, our patient was a well looking girl. She had a full female phenotype: On initial physical examination the breasts were abnormally developed compatible with Tanner stage III. The gynecological exam reveals normal external female genitalia, the vagina and hymen were seen, but Pubic hair was not. Normally her weight was 24 kg (90 percentiles on CDC growth charts), her height was 124 cm (95th percentile on CDC growth charts), there was no advanced bone age in X Ray. Excepting elevated estrogen levels other hematological and biochemical profiles including thyroid function test were normal. The levels of gonadotropins were measured and found (FSH 3.18 mIU/mL, LH 0.8 mIU/mL); estradiol was 64 pg/ml but initial ultrasonographic study of abdomen and pelvic ultrasonography showed no abnormality, brain MRI was also normal. After getting all this investigation, we came to conclusion the patient may be suffered from constitutional precocious puberty. Despite this she was regularly monitored, after 3 months her breasts began to grow rapidly became as large as tanner stage of IV and she had 4 cm increasing in her height. Repeat of hormonal assay showed high levels of estradiol, 145pg/ml but tumor markers levels were normal, with a total BHC of 0.1ml/L and an alpha-fetoprotein of 0.9IU/mL. At this time second thorough abdominal and pelvic ultrasonography workup revealed a round solid hypo echo and vascular structure mass measuring about 28.23mm in her left pelvic cavity. Surprisingly The pelvic MRI also detected, the lack of uterus and ovaries and short blind end vagina, with oval shape structure measuring about 10.6mm in right side of pelvic cavity. For this reason The blood sample was sent to the molecular karyotyping laboratory for detection of chromosomal abnormality. This test confirmed the suspected diagnosis of testicular feminization syndrome 46XY. Our patient underwent an explorative laparotomy 3 months after her initial presentation to our clinic and a solid tumor localized to her left side of pelvic cavity was identified. After resection of tumor, gross examination showed the well-circumscribed yellow-pale mass measured about 2 x 2.5 cm in diameter. The mass was capsulated and congested blood vessels were seen at the outer surface of tumor. Following surgery Sample sent for pathologist, histopathological report confirmed the diagnosis of a Sertoli cell tumor composed of proliferated variable sized tubules lined by polygonal cells with small uniform nuclei and abundant cytoplasm with a mild atypical mitosis less than 5 per 10hpf, no necrosis microscopic criteria in favor of benign pattern. Immunohistochemical study of the tumor cells showed negative staining for EMA alpha-fetoprotein and positive staining for inhibin. Our patient is well now without evidence of tumor recurrence or metastasis during six months of post-surgery follow-up. Levels of plasma estrogen rapidly returned to the normal pubertal range and breast enlargement also. Since testicular estrogen and peripheral conversion of androgen to estrogen help to patient feminization, Our plan is follow her every three months and removal of other testes after puberty when feminization will be complete.

**Conclusions**

Although in most of girls with precocious puberty, the etiology is idiopathic, Sertoli cell tumor with secretion of estrogen should be considered in the differential diagnosis for a prepubescent girl with an abdominal mass.