specifying genes in the absence of SRY and/or inadequate expression of pro-ovary/anti-testis genes such as SOX 8 and SOX9. SOX10, a gene closely related to SOX9 and SOX8, and its overexpression has been suggested as a candidate for 46,XX DSD. Over-expression of SOX10 in mice resulted in the XX DSD. The spectrum of sex reversal phenotypes and gonadal asymmetry
seen in the Sox10 transgenic mice closely mirrors the range of gonadal and reproductive tract anomalies seen in cases of partial duplication of human chromosome 22q13. Although SRY-negative 46,XX testicular DSD is a rare condition, an effort to make an accurate diagnosis is important for the provision of proper genetic counseling and for guiding patients in their long-term management.

Pediatric Endocrinology

PEDIATRIC ENDOCRINOLOGY CASE REPORT

6-Year-Old Girl With a Luteinized Follicular Ovarian Cyst and an Estradiol Level > 1,000 PG/ML

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Background: Precocious puberty in girls is defined as onset of secondary sexual characteristics, such as breast development, before 8 years of age. To differentiate between central and peripheral precocious puberty, laboratory and imaging evaluation is helpful. When gonadotropins are low but estradiol is elevated, results may suggest a primary ovarian source of estrogen production. Small ovarian cysts are not uncommon, are benign and self-resolve. However, large ovarian cysts are rare, let alone ones requiring surgical removal.
Clinical Case: A 6 year 7 month old girl presented with several days of breast tenderness and palpable bilateral breast tissue noted by her mother. There was no history of vaginal bleeding. There were no reported exposures to estrogen-containing products. Her mother reached menarche at age 14 years. The patient was born full term and was otherwise healthy. On exam, her height was at the 90-95th %ile (mid-parental height at the 95th %ile) and her growth velocity was 10.9 cm/yr. She had Tanner 2 breasts (1 cm breast bud on the left and 1.5 cm on the right), Tanner 1 pubic hair and no axillary hair, body odor, acne or café-au-lait macules. A bone age was read as 6 years at a chronological age of 6 years 7 months. A laboratory evaluation revealed an estradiol of 1,029 pg/mL (<15 pg/mL), LH <0.02 mIU/mL, FSH <0.09 mIU/mL, 17-hydroxyprogesterone (17-OHP) 410 ng/dL (<91 ng/dL), AFP 2.3 ng/mL (<6.1 ng/mL), beta-hCG <2 mIU/mL, TSH 2.41 mIU/L (0.5-3.2 mIU/L), and free T4 0.9 ng/dL (0.9-1.4 ng/dL). Pelvic ultrasound revealed a large unilocular cystic structure measuring 6.5 x 4.1 x 6.1 cm in the left adnexal region with no left ovary visualized. The right ovary appeared prepubertal. The uterus was prepubertal in appearance with endometrial thickness of 2 mm. Abdominal ultrasound showed no evidence of a suprarenal mass. A laparoscopic cyst resection was completed, given the risk of left ovarian torsion. Cytology was negative; pathology revealed a luteinized follicular cyst. Repeat labs in one month showed a prepubertal estradiol level of 6.7 pg/mL with LH 0.02 mIU/mL and FSH 0.38 mIU/mL. 17-OHP normalized to 29 ng/dL. Breast tissue had regressed.
Conclusion: This case describes the rare finding of a large luteinized follicular ovarian cyst that required surgical removal in a 6-year-old girl in the setting of a significantly elevated estradiol level. Luteinized follicular cysts have been described in newborns, though rare. To our knowledge, this is the first described case of a luteinized follicular cyst in this patient's age group. Laboratory and imaging evaluation should be considered in girls presenting with precocious puberty, despite the extent of thelarche, as the clinical examination does not always correlate with degree of estradiol elevation. This is especially important if clinical changes are acute and other features are consistent with puberty, such as rapid linear growth.

Pediatric Endocrinology

PEDIATRIC ENDOCRINOLOGY CASE REPORT

A Case of Autoantibody Negative Pediatric Diabetes Mellitus With Marked Insulin Resistance Concomitant With COVID-19: A Novel Form of Disease?

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Background: SARS-CoV-2 uses the angiotensin-converting enzyme 2 (ACE2) receptor to enter human cells. This receptor is avidly expressed in the pancreatic islets, suggesting the virus may target β-cell function. 17% of adults with COVID-19 have evidence of pancreatic injury. There is a direct relation between insulin resistance and COVID-19 severity and mortality with patients with higher insulin resistance presenting with higher inflammatory response. Fulminant type 1 diabetes (fT1D) has abrupt onset of symptoms with insulinopenia without evidence of autoimmunity, usually preceded by viral illness. **Clinical case:** A 12-year-old Hispanic male presented with a week history of polyuria, polydipsia, headache, and fatigue but no weight loss, fever, cough, anosmia, or diarrhea. Laboratory testing revealed new onset diabetes with DKA with HbA1C 11.3%, blood pH 7.04, glucose 381 mg/dL, C-peptide 0.6 ng/mL, β-hydroxybutyrate 8.2 mmol/L, and a positive nasopharyngeal PCR for SARS-CoV-2 but no elevation of inflammatory markers (CRP, ESR and ferritin). There was evidence of mild pancreatic injury (lipase 179 U/L, n:15-110 U/L), and all autoantibodies for autoimmune diabetes were negative. He was pubertal (Tanner 3), non-obese (BMI Z score -0.3) and without acanthosis nigricans. Past medical and family history were non-contributory. He was treated with IV insulin at 0.1 u/kg/h until DKA resolved within 24 h then transitioned to subcutaneous insulin at 1 u/kg/day. He did not have signs of systemic inflammatory response, need for respiratory support, or glucocorticoids but had persistent hyperglycemia prompting an increase of insulin dosing and resumption of IV insulin. His insulin requirements continued to increase up to 4 u/kg/d with sustained hyperglycemia indicative of an exceptional state of insulin resistance. On Day 6 of admission metformin was initiated, on Day 9 insulin requirements declined and he was discharged on an insulin regimen close to 1.5 u/kg/d.