Pediatric Genitourinary Cases

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2 year-old boy with painful hematuria
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Pathology: Polyp of the verumontanum
Urethral Polyp

Regarding urethral polyps, which of the following is TRUE?

A. More commonly affects girls than boys
B. Most frequently occurs in the anterior urethra.
C. Typically solitary and pedunculated.
D. Treated with surgical resection due to malignant potential.
Urethral Polyp

• Presenting symptoms
  • Intermittent urinary retention
  • Hematuria
  • Lower urinary tract symptoms
  • Protrusion of the polyp at the urethral meatus in girls

• Etiology: Developmental error in the invagination process of the submucosal, glandular portion of the inner zone of the prostate
Urethral Polyp

Reference:

16 year-old with long-standing lump at the base of the left testes
16 year-old with long-standing lump at the base of the left testes

Diagnosis: Left accessory testicle, microlithiasis
16 year-old with long-standing lump at the base of the left testes

Regarding accessory testes, which of the following is TRUE:

A. More commonly left-sided
B. Typically present as a painless mass
C. Unlikely to contribute to reproductive function
D. Usually presents in the neonatal period
16 year-old with long-standing lump at the base of the left testes

Reference:

2 month-old boy with hypoplastic left heart, status post Norwood and Sano
2 month-old boy with hypoplastic left heart, status post Norwood and Sano

The provided images suggest which possible abnormality?

A. Ectopic air
B. Renal calculus
C. Tube malposition
D. Skeletal dysplasia
2 month-old boy with hypoplastic left heart, status post Norwood and Sano
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Diagnosis: Perforated ND tube with tip in right kidney

Reference:

Newborn with ambiguous genitalia
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Newborn with ambiguous genitalia

This patient’s imaging features are most compatible with:

A. Female pseudohermaphrodite
B. Male pseudohermaphrodite
C. True hermaphrodite
D. Mixed gonadal dysgenesis
Newborn with ambiguous genitalia

Diagnosis: Ovotestes, true hermaphrodite.

Reference:

Disorders of Sexual Development (DSD)

Congenital conditions in which chromosomal, gonadal, or anatomic sex is atypical

- Female pseudohermaphrodite: (46, XX DSD) Female genotype, 2 ovaries, variable virilization
- Male pseudohermaphrodite: (46, XY DSD) Male genotype, 2 testes, variable degree of external feminization
- True hermaphrodite (Ovotesticular DSD): Have both testicular and ovarian tissue in the gonads. Usually 46, XX
- Pure gonadal dysgenesis (PGD): Both gonads are streak gonads. 45, XY/XO mosaic
- Mixed gonadal dysgenesis (MGD): Have a testes on one side and a streak gonad on the other. Variable genotype
6 year-old girl with intermittent abdominal pain and suspected constipation
6 year-old girl with intermittent abdominal pain and suspected constipation
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6 year-old girl with intermittent abdominal pain and suspected constipation

Pathology: Ovarian fibroma.

Ovarian fibromas are rare in children and may be associated with Gorlin syndrome in some cases.
Nevoid Basal Cell Carcinoma Syndrome (Gorlin Syndrome)

Which features characterize ovarian fibromas in the setting of Gorlin Syndrome:

A. Calcified and bilateral.
B. Calcified and unilateral.
C. Non-calcified and bilateral.
D. Non-calcified and unilateral.
Nevoid Basal Cell Carcinoma Syndrome (Gorlin Syndrome)

Manifestations:
- Basal cell carcinoma (80%)
- Palmoplantar pits (87%)
- Ondontogenic keratocysts (65-70%)
- Ovarian or cardiac fibromas (14-24%)
- Medulloblastoma (<4%)
- Other
  - Intracranial and dural calcifications
  - Craniofacial malformations
  - Lymphomesenteric cysts
Nevoid Basal Cell Carcinoma Syndrome (Gorlin Syndrome)

Reference:

8 month-old girl whose parents noticed a left abdominal mass
8 month-old girl whose parents noticed a left abdominal mass.
8 month-old girl whose parents noticed a left abdominal mass

Pathology: Clear cell sarcoma with tumor extension into soft tissue of the renal sinus and lymphatic invasion present.
Clear Cell Sarcoma of the Kidney

Regarding clear cell sarcoma of the kidney, the MOST COMMON site of metastases is:

A. Bone
B. Brain
C. Lung
D. Hilar lymph nodes
Clear Cell Sarcoma of the Kidney

- Second most common renal tumor in children
- Typically presents in children younger than 4 years old
- Can mimic Wilms tumor, congenital mesoblastic nephroma, and rhabdoid tumor of the kidney
- 29% have evidence of lymph node metastases at presentation

Reference:
15 year-old girl with progressive abdominal fullness and discomfort
15 year-old girl with progressive abdominal fullness and discomfort
15 year-old girl with progressive abdominal fullness and discomfort
15 year-old girl with progressive abdominal fullness and discomfort
15 year-old girl with progressive abdominal fullness and discomfort

Pathology: Vaginal mass, Biopsy: Ewing sarcoma/primitive neuroectodermal tumor. FISH analysis performed confirms presence of EWSR1 rearrangement. There is no evidence of an SYT rearrangement.
Extraosseous Ewing Sarcoma

Compared to Ewing sarcoma of bone, extraskeletal Ewing sarcoma:

A. Typically occurs in younger patients.
B. Has a predilection for the trunk.
C. More commonly affects boys than girls.
Extraosseous Ewing Sarcoma

Imaging features
• Radiography
  • Large soft tissue mass
• US:
  • Typically hypoechoic mass; increased blood flow on Doppler
• CT:
  • Non-specific soft tissue mass, attenuation similar to muscle
  • Calcifications in 25%-30%
• MR:
  • T1-weighted images: Soft tissue mass similar in intensity to muscle
  • T2-weighted images: Intermediate to high signal intensity mass

Reference:
12 year-old girl with acute onset urinary retention and back pain
12 year-old girl with acute onset urinary retention and back pain
12 year-old girl with acute onset urinary retention and back pain
12 year-old girl with acute onset urinary retention and back pain

Pathology: Pelvic tumor biopsy: Ewing sarcoma

Reference:
2 year-old boy with past history of a febrile urinary tract infection
2 year-old boy with past history of a febrile urinary tract infection
2 year-old boy with past history of a febrile urinary tract infection

Of the following choices, which diagnosis is LEAST likely:

A. Calyceal diverticulum
B. Cystic Wilms tumor
C. Hemorrhagic cyst
D. Renal abscess
2 year-old boy with past history of a febrile urinary tract infection

Pathology: Right kidney, partial nephrectomy: Lobular renal dysplasia with reflux changes.

Pathologist’s note: This is an unusual lesion in that the lobule is so intact, and that the papillae is pyramidal rather than the expected concave with reflux.

Radiologist’s note: VCUG was negative.
12 year-old girl with recurrent abdominal pain
12 year-old girl with recurrent abdominal pain

Pathology: Right fallopian tube: Extensive necrosis with calcification and macrophage response, consistent with torsion. Right ovarian cyst: Ovarian tissue with corpus luteum cyst.
22 year-old woman with abdominal pain
22 year-old woman with abdominal pain
22 year-old woman with abdominal pain
22 year-old woman with abdominal pain

Pathology: Hemorrhagic, necrotic ovary and fallopian tube consistent with torsion.