2023 Slide Survey Online

Cases 23-06 – 23-10

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Release Date: August 31, 2023

Expiration Date: December 31, 2023

Program Overview

Pediatric pathologists often encounter cases for which they've had limited exposure based on the rarity of a particular disease and/or the frequency with which it presents in their research and/or practice. They are presented with cases that are difficult to diagnose and often require additional consultation and/or work-up. Small specimen size and tissue sampling at earlier stages of a disease add to the diagnostic challenge. With the new developments and refinement of diagnostic criteria, particularly as related to genomic etiologies or associated molecular alterations, pediatric pathologists face challenges in staying up to date with new information that is frequently changing based on rapid advancements in technology. Information, tools, and strategies to address challenging cases are necessary in order to improve the accuracy of diagnoses.

This activity will provide up-to-date information, including the latest research, diagnostic criteria, and diagnostic tools and strategies, on a variety of diagnostic entities across the pediatric spectrum for pediatric pathologists to apply this information to their practice and/or research.

Target Audience

The target audience for this educational activity is pediatric pathologists engaged in the study and diagnosis of pediatric diseases.

Learning Objectives

1. Describe advancements in diagnostic criteria and techniques.
2. Apply diagnostic criteria to develop differential diagnoses.
3. Integrate the most recent advances in diagnostic techniques into practice.

American Board of Pathology (ABPath) Continuous Certification

This activity is registered for American Board of Pathology (ABPath) Lifelong Learning (CME) credit and Improvement in Medical Practice (Part IV) requirements of the ABPath Continuing Certification Program. For more information about CME credits contact the SPP at spp@aoeconsulting.com.

Successful completion of this CME activity, including a passing score on the post-tests and participation in the evaluation component and the post-activity follow-up survey, enables the participant to satisfy the Lifelong Learning (CME) credit and Improvement in Medical Practice (Part IV) requirements in the American Board of Pathology's (ABPath) Continuing Certification (CC) program. It is the CME activity provider's responsibility to submit participant completion information to the ACCME for the purpose of granting credit.

Successful completion of the CME activity allows the participant to earn up to 5.0 Lifelong Learning (CME) credits and up to 5.0 Improvement in Medical Practice (Part IV) credits.

To receive credit, you must complete the post-tests with a score of 80% or better, the evaluation, and the post-activity follow up survey. You must also provide your ABPath Diplomate ID and Date of Birth (MM/DD). Your information will be shared with the ABPath through the ACCME Program and Activity Reporting System (PARS).

Canadian Physicians

Through an agreement between the Accreditation Council for Continuing Medical Education and the Royal College of Physicians and Surgeons of Canada, medical practitioners participating in the Royal College's MOC Program may record completion of accredited activities registered under the ACCME's "CME in Support of MOC" program in Section 3 of the Royal College's MOC Program.

Accreditation Statement

The Society for Pediatric Pathology is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

Credit Designation Statement

The Society for Pediatric Pathology designates this enduring material for a maximum of 5.0 *AMA PRA Category 1 Credits™*. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

The Slide Survey Online enduring material is released in three separate case sets: Case Set 1, Case Set 2, and Case Set 3. Each case set is worth 5.0 credits.

Fees for this activity are as follows:

Practicing or Retired Pathologist: $125 for SPP members, $150 for Non-Members.

Pathologist-in-Training: No fee.

For information about the accreditation of this program, please contact the Society for Pediatric Pathology at (720) 625-8271.

Instructions to Receive Credit

To receive a Certificate of Credit (physicians) or a Certificate of Participation (non-physicians), the participant must complete the evaluation/application for credit form and complete the post-tests with a score of 80% or better. Participants are allowed to take the post-tests multiple times. The participant must also complete the post-activity follow-up survey. The estimated time for completion of this activity is 5 hours (1 hour per case). Participants must complete the requirements and claim credit by December 31, 2023.

Instructions for Online Responses

1. Download the Accreditation & Questions Booklet.
2. Review the case content including the virtual slide, images, and clinical history.
3. Complete the pre-test. The pre-test may only be completed once. Your answers do not affect your ability to complete the course. You must click "Finish" for your answers to be submitted.
4. Review the Discussion. The discussion may only be accessed after the pre-test is completed.
5. Review the case content again, then complete the post-test. Correct any incorrect answers from the pre-test. (Note: You must score 80% on each post-test. You have an unlimited number of attempts). You must click "Finish" for your answers to be submitted.
6. The Discussion & References Booklet will become available after you pass each post-test.
7. Complete the evaluation.
8. Claim Credit: Once you have scored 80% or higher on each post-test, you may claim credit.
9. Download your certificate.

Post-Activity Follow-Up Survey

Please complete the post-activity follow-up survey that will be sent via email upon completion of the Case Set.

Disclosures

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Faculty, Authors, and Content Developers

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Bradford Siegele, MD, JD; José Velázquez-Vega, MD; Louis Samson, MD; Anita Nagy, MD; Yoo Minh Anh, DO; Shannon Kelley, MD; Natalia Belova, MD; David Chiasson, MD; Robert Byrd, MD

Planners and Managers

All planners and managers have **no relevant financial relationships with ineligible companies:**

Lauren Parsons, MD; Amy Coffey, MD; Consolato Sergi, MD, PhD, MPH, PRCPC; Shipra Garg, MD; Shahrazad Saab, MD; Larry Wang, MD, PhD; Bradford Siegele, MD, JD; Julie Fanburg-Smith, MD; Anita Nagy, MD; Athena Chen, MD; Ali Saad, MD; Caitlin Alexander, MD, MPH; Shannon Kelley, MD; Jose Velazquez-Vega, MD; Sandy Cope-Yokoyama, MD; Jiancong Liang, MD; Dehua Wang, MD; Lea Surrey, MD; Sam Sirotnikov, DO; Nick Shillingford, MD; Kristen Thomas, MD; Oscar Lopez-Nunez, MD; Kathryn McFadden, MD; Naseem Uddin, MD; Dina El Demellawy, MD, PhD; Sean Ricciardo, MD; Camilla Stefanovici, MD; Diana Corao-Uribe, MD; Melissa Gener, MD; Jenna Bodmer, MD; Natalia Belova, MD; Jessenia Guerrero, MD; Robyn Reed, MD, PhD; Mikako Warren, MD; Robert Byrd, MD; Louis Samson, MD, FRCPC; Sarah Porter, CHCP; Lyndsay Cole

Faculty and Authors

José E. Velázquez, MD, is a pediatric pathologist and neuropathologist at Children's Healthcare of Atlanta, an Adjunct Assistant Professor at the Emory University School of Medicine, and a member of the SPP Education Committee and Slide Survey Subcommittee**.**

Louis Samson, MD, is a pathologist at CHU de Québec – Hôtel-Dieu de Québec, in Québec, Canada, and is a member of the SPP Education Committee and the Slide Survey Subcommittee.

Anita Nagy, MD, FRCPath, is a pathologist at the Hospital for Sick Children and an Assistant Professor at the University of Toronto and is a member of the SPP Education Committee and the Slide Survey Subcommittee.

Yoo Min Ahn, DO, is an anatomic pathology resident at the University of Texas Southwestern Medical Center, Dallas.

Shannon Kelley, MD, is a pediatric pathologist at Children’s Medical Center, Dallas, an assistant professor of pathology at the University of Texas Southwestern Medical Center, Dallas, and a member of the SPP Education Committee and Slide Survey Subcommittee.

Bradford Siegele, MD, JD, is a pediatric pathologist and hematopathologist at Children’s Hospital Colorado and an assistant professor of pathology at the University of Colorado School of Medicine. He is a member of the SPP Education Committee and is Chair of the Slide Survey Subcommittee.

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Disclaimer

Participants have an implied responsibility to use the newly acquired information to enhance patient outcomes and their own professional development. The information presented in this activity is not meant to serve as a guideline for patient management. Any procedures, medications, or other courses of diagnosis or treatment discussed in this activity should not be used by clinicians without evaluation of patient conditions and possible contraindications on dangers in use, review of any applicable manufacturer's product information, and comparison with recommendations of other authorities.

Case Submissions

The Slide Survey Subcommittee welcomes your comments and submission of cases. Case submissions require a single slide, either glass or virtual (.svs format). The slide, de-identified pathology report, relevant clinical history, and any ancillary digital images should be submitted to:

Bradford Siegele, MD, JD, Slide Survey Chair
Assistant Professor
Children's Hospital Colorado
Anschutz Medical Campus
13123 East 16th Avenue
Aurora, CO 80045
Phone: 720-777-0560
E-mail: Bradford.siegele@childrenscolorado.org

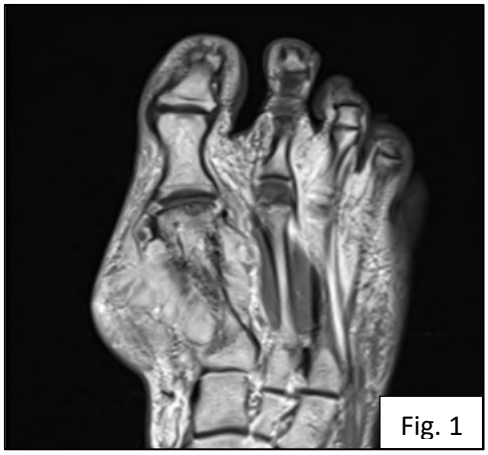


Fig. 1

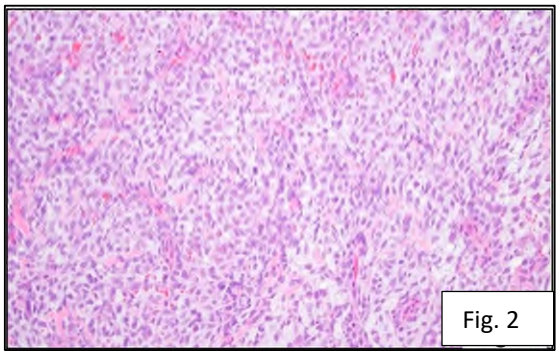


Fig. 2

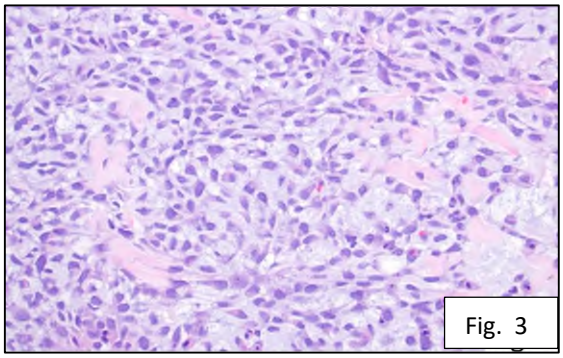


Fig. 3

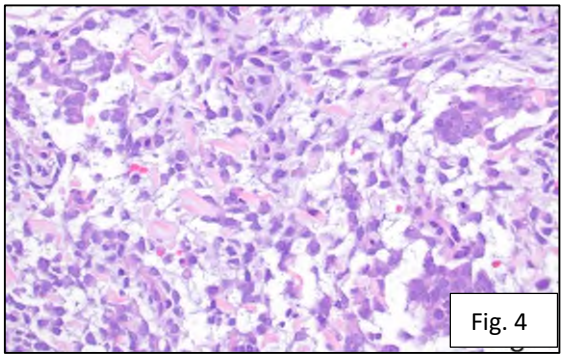


Fig. 4

SPP Slide Survey Case 23-06

A 12-year-old female with no significant past medical history presented for evaluation of a left foot mass that had shown progressive growth over the course of the preceding four months. She initially noted the mass as a small, skin-colored bump on the medial aspect of her left foot that grew despite initial treatment with incision and drainage and antibiotics. She denied any preceding trauma to the area, pain, issues with weight bearing or ambulation, or limitations to playing sports. She endorsed no associated fatigue, joint pain, weight loss, changes in appetite, fevers, chills, or night sweats. Physical examination revealed a large, non-tender left foot mass protruding from the medial aspect of the first metatarsal with a central area of ulceration and necrotic rim without active drainage.

Magnetic resonance imaging (MRI) of the left foot (Fig. 1) revealed a 6.6 cm heterogeneously enhancing mass involving the first metatarsal and the surrounding soft tissues. Chest CT revealed multiple, bilateral subcentimeter lung nodules consistent with metastatic disease.

A whole slide image (H&E) of the left foot mass is provided (see also Figs. 2-4). Immunohistochemistry for SATB2 (Fig. 5) is positive, as are immunohistochemical stains for TLE-1 (not provided) and FLI-1 (not provided), as well as a diagnostic immunohistochemical stain (Fig. 6). NKX2.2 (not provided) is focally positive. CD99, WT-1, desmin, myogenin, myoD1, SMA, S-100, HMB45, pancytokeratin, EMA, CD34, ERG, MUC4, and ALK1 (not provided) are negative. BAF47/INI-1 staining (not provided) is retained.

EWSR1 break-apart and *SS18 (SYT)* fluorescence in situ hybridization studies were negative.

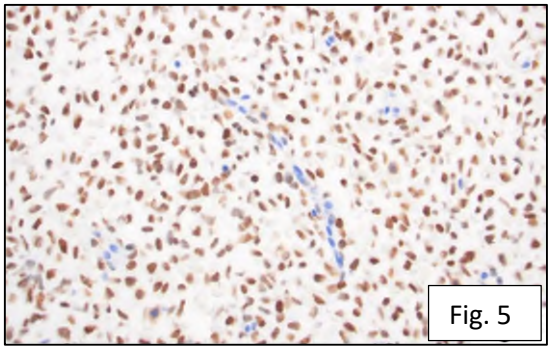


Fig. 5

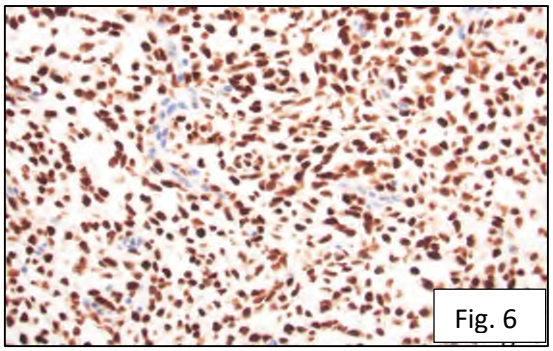


Fig. 6

Diagnostic List:

Please select the most likely diagnosis below:

a. Anaplastic large cell sarcoma (ALCL), ALK-negative
b. *CIC*-rearranged sarcoma
c. Ewing sarcoma
d. Osteosarcoma, high-grade
e. Rhabdomyosarcoma
f. Sarcoma with *BCOR* genetic alterations
g. Synovial sarcoma

1) Which of the following tumors is associated with the most aggressive behavior and lowest overall survival?

a. *CIC*-rearranged sarcoma
b. Ewing sarcoma, *EWSR1::ERG* rearranged
c. Ewing sarcoma, *EWSR::FLI1* rearranged
d. Sarcoma with *BCOR* genetic alterations
e. Synovial sarcoma

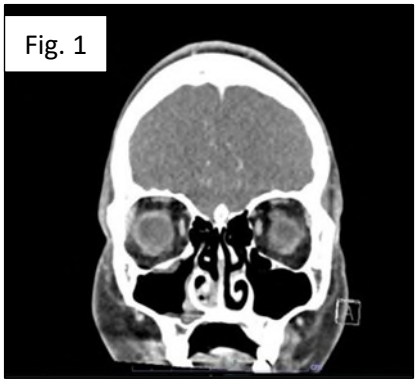
2) In addition to sarcoma with *BCOR* genetic alterations, which of the following tumors can be strongly positive for BCOR immunohistochemistry?

a. Angiosarcoma
b. Chondrosarcoma
c. Desmoplastic small round cell tumor (DSRCT)
d. Low-grade fibromyxoid sarcoma (LGFMS)
e. Synovial sarcoma

3) Which of the following is a characteristic clinicopathologic feature of Ewing sarcoma?

a. CD99 immunohistochemistry characteristically shows strong and diffuse nuclear expression.
b. Nuclear positivity for NKX2.2 is characteristic in cases with *EWSFLI1* rearrangement.
c. Tumor nuclei most often exhibit prominent nucleoli and significant pleomorphism.
d. Tumors most often arise in children <10 years of age.
e. Tumors most often arise in the viscera.

Fig. 1



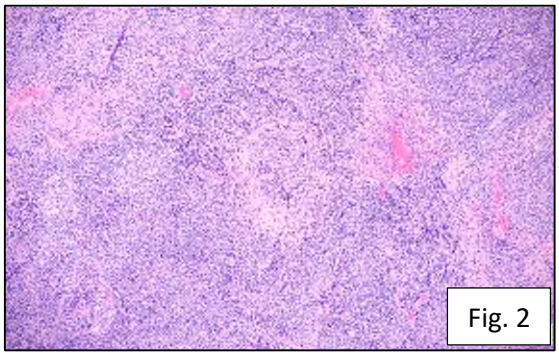


Fig. 2

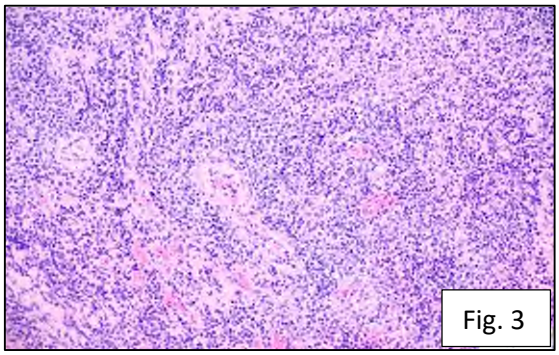


Fig. 3

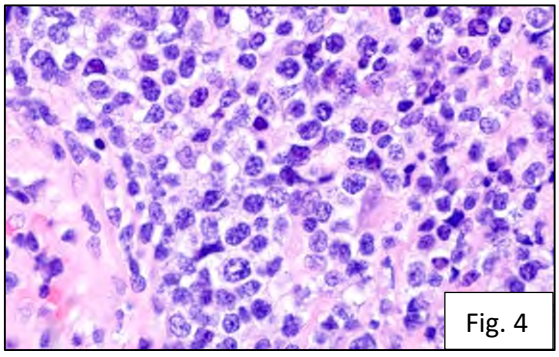


Fig. 4

SPP Slide Survey Case 23-07

A 14-year-old Hispanic male with no previous medical history presented to the emergency department with a 2-month history of rapidly worsening purulent nasal discharge. During the week prior to presentation, increased pain and swelling of the right nasal ala were noted. Review of systems was additionally positive for loss of appetite, fatigue, and fever. Physical exam revealed an erythematous and swollen right nasal ala with partial obstruction of the naris by white purulent discharge. Bilateral cervical lymphadenopathy was palpated. Computed tomography (CT) imaging studies (Fig. 1) showed a right facial and nasal soft tissue mass extending into the nasal cavity with focal erosion of the anterior hard palate. Peripheral blood cell counts were unremarkable.

An emergent operative biopsy was performed with portions of the specimen sent for microbiologic cultures and pathologic assessment.

A whole slide image (H&E) of representative tissue from the biopsy is provided (see also Figs. 2-4), as are immunohistochemical studies for CD3 (Fig. 5) and CD56 (Fig. 6). Epstein-Barr virus encoding RNA (EBER) *in situ* hybridization studies are additionally included (Fig 7). Lesional cells were additionally positive for CD7 and negative for CD20, PAX5, CD34, CD4, and CD8 (not provided).

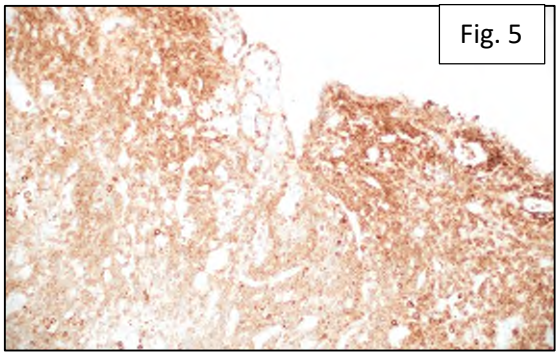


Fig. 5

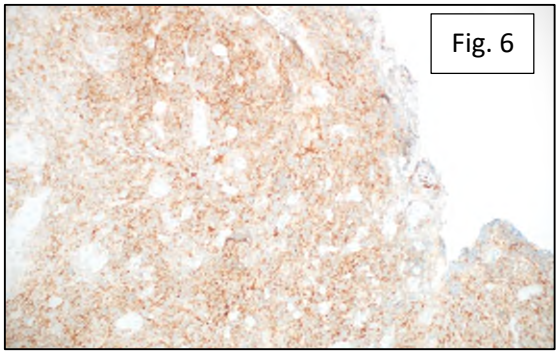


Fig. 6

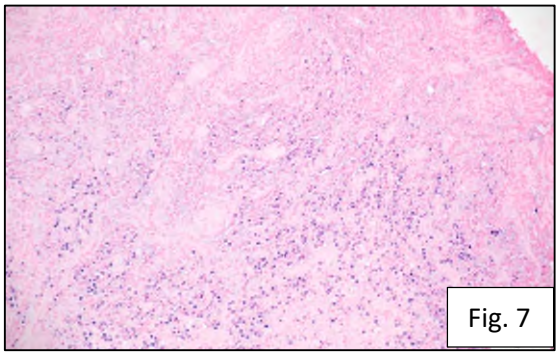


Fig. 7

Diagnostic List:

Please select the most likely diagnosis below:

a. Acute EBV+ cytotoxic T-cell lymphoid hyperplasia of upper aerodigestive tract (EBV+TLH)
b. EBV+ diffuse large B-cell lymphoma
c. Extranodal NK/T-cell lymphoma (ENKTL), nasal type
d. Lymphomatoid granulomatosis

1) What is the molecular pathway most frequently altered/activated in the pathogenesis of extranodal NK/T-cell lymphoma (ENKTL), nasal type?

a. JAK/STAT pathway
b. NF-κB signaling pathway
c. P53-MDM2 pathway
d. Wnt signaling pathway

2) Which of the following is a major clinicopathologic feature of acute EBV+ cytotoxic T-cell lymphoid hyperplasia of the upper aerodigestive tract (EBV+TLH) permitting differentiation from neoplastic mimics?

a. EBV+TLH affects predominantly the nasopharynx and cervical lymph nodes.
b. EBV+TLH exhibits acute onset with spontaneous complete remission within one month with no cytotoxic therapy.
c. EBV+TLH is almost exclusively seen in the context of immunosuppression.
d. EBV+TLH is characterized by atypical lymphoid infiltrate that is CD5+ and CD56+.

3) Which of the following morphologic features is characteristic of lymphomatoid granulomatosis?

a. Angiocentric and angiodestructive polymorphous lymphoid infiltrate with involvement of small to large caliber vessels.
b. Diffuse sheet-like proliferation of atypical large lymphoid cells.
c. “Hallmark” cells with abundant cytoplasm, horseshoe-shaped nuclei, and perinuclear eosinophilic regions.
d. Large transformed immunoblasts, Hodgkin/Reed-Sternberg-like (HRS-like) and lymphocyte-predominant-like cells scattered in a background of small lymphocytes, plasma cells, and histiocytes.

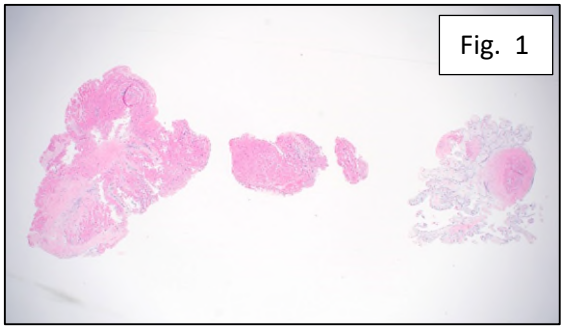


Fig. 1

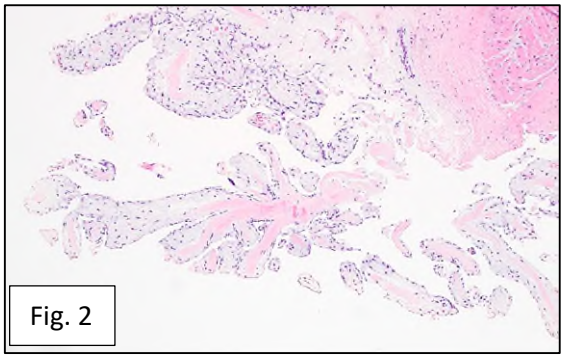


Fig. 2

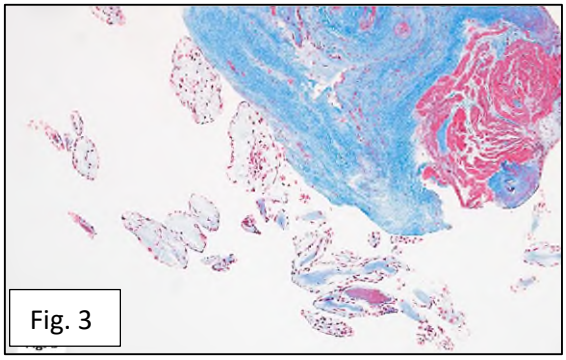


Fig. 3

SPP Slide Survey Case 23-08

A 7-year-old female with a past medical history significant for an orthotopic heart transplant three years earlier presented for follow-up cardiac catheterization and surveillance biopsy. Three months prior, she had been found to have elevated brain natriuretic peptide (BNP) levels, abnormal hemodynamics, and an "abnormal" endomyocardial biopsy, and was treated for rejection with anti-thymocyte globulin (ATG).

A whole slide image (H&E) of her endomyocardial biopsies is provided (see also Figs. 1-2) along with a trichrome stain (Fig. 3). A Verhoeff-van Gieson (VVG) (not provided) special stain was positive. The immunohistochemical stain for C4d (not provided) was negative.

Diagnostic list:
Please select the most likely diagnosis below:

a. Cardiac myxoma.
b. Cardiac thrombus.
c. Lambl excrescence.
d. Papillary fibroelastoma.
e. Valvular vegetation.

1) Which of the following correspond to the most characteristic features of papillary fibroelastoma?

a. Attached to the atrial septum. Pedunculated or villiform architecture. Carney complex association.
b. Complex branches. Narrow avascular papillary fronds. Fibroelastic core. Stromal mucopolysaccharide.
c. Polygonal myxoma cells. Blood vessels in the stroma. Mucopolysaccharide-rich stroma. Calretinin positivity.
d. Small papillary processes. Arranged along free cuspal / leaflet edges. Stromal mucopolysaccharide.
e. Variably cellular stoma. Valvular destruction. No elastic fibers. Clinical symptoms of endocarditis

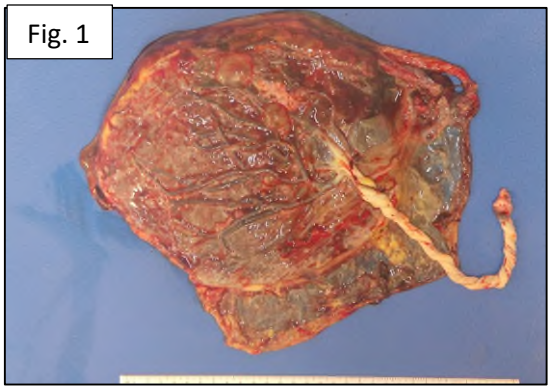
2) Which of the following findings is appropriately matched with the diagnosis?

a. *KRAS* mutation in a subset of cases – Cardiac myxoma.
b. Located in the atrial appendages or region of ventricular aneurysm – Cardiac thrombi.
c. Loss of *PRKAR1A* expression within the myxoma cells – Papillary fibroelastoma.
d. Solitary in a majority of cases – Lambl excrescence.
e. Typical location at the line of valve closure - Papillary fibroelastoma.

3) Which of the following statements is true about cardiac myxomas?

a. A minority of the cases are sporadic.
b. An elastic stain is not helpful to differentiate cardiac myxoma from papillary fibroelastoma.
c. Myxoma cells variably express vimentin, NSE, S100, synaptophysin, SMA and desmin.
d. Myxomas typically occur in the right ventricle and are attached to the ventricular septum by a stalk.
e. Symptoms are most frequently related to local and disseminated spread of metastatic disease.

Fig. 1



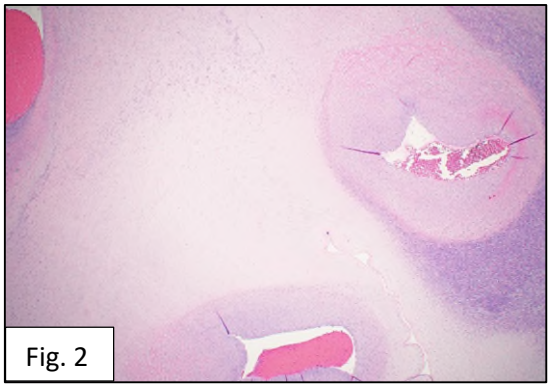


Fig. 2

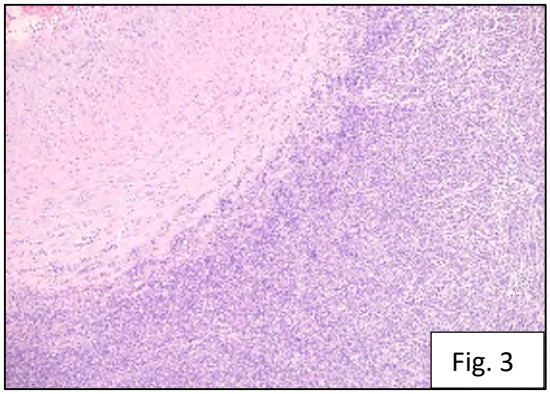


Fig. 3

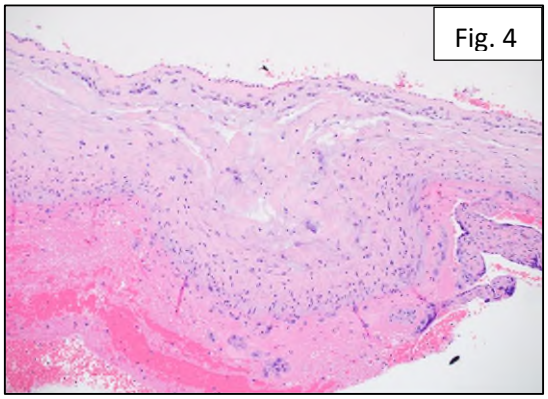


Fig. 4

SPP Slide Survey Case 23-09

A 23-year-old G5P4 female at 31 weeks, 5 days gestation with a history of two spontaneous abortions and recent viral illness presented to an outside hospital with a complaint of post-coital bleeding. Physical examination demonstrated mildly elevated blood pressures, and laboratory studies showed an elevation in liver function tests. The initial clinical concern was for placental abruption in the setting of preeclampsia. Work-up showed no clinical signs of preterm labor. Ultrasound imaging revealed a hydropic fetus with polyhydramnios, hepatosplenomegaly, ascites, and an elevated middle cerebral artery (MCA) peak systolic velocity by Doppler examination. The placenta appeared thickened. Prenatal laboratory tests for maternal HepA, HBsAg, HCV, RPR, and HIV serologies were negative. Group B streptococcus (GBS) studies were negative. Chlamydia and gonorrhea testing was negative. The maternal blood type was O+.
The fetus was otherwise unremarkable by imaging with appropriate size and fetal movement. A fetal echocardiogram showed increased cardiac output and mild cardiomegaly.

After transfer to a high-level fetal care center, ultrasound-guided fetal hepatic vein sampling revealed fetal anemia, and a fetal intrauterine packed red blood cell (PRBC) transfusion was performed. Cytomegalovirus (CMV) and parvovirus B19 serologies performed on fetal blood were negative.

Six days later, preterm premature rupture of membranes (PPROM) was diagnosed, and a viable male infant was delivered via low-transverse cesarean section.

Gross examination of the placenta revealed a large for gestational age placenta (740 grams, 99th percentile for 32-33 weeks) with an edematous appearance and green discoloration

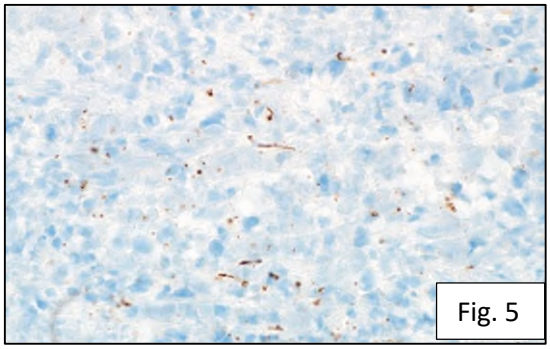


Fig. 5

of the fetal membranes. The three-vessel umbilical cord was diffusely hypercoiled (>3 coils/10 cm) and peripherally inserted (see Fig. 1).

Whole slide images (H&E) of cross-sections of the umbilical cord and a membrane roll (A) and placental parenchyma (B) are provided (see also Figs. 2-4), as is a disease-specific immunohistochemical stain (Fig. 5). A Grocott-Gomori methenamine silver (GMS) special stain (not included) was negative for fungal elements.

Diagnostic List:
Please select the most likely diagnosis below:

a. Congenital cytomegalovirus (CMV)
b. Congenital herpes simplex
c. Congenital parvovirus B19
d. Congenital syphilis
e. Congenital toxoplasmosis
f. Fetal vascular malperfusion
g. Maternal vascular malperfusion
h. Villitis of unknown etiology

1) What is the Amsterdam Classification stage and/or grade of the fetal inflammatory response shown in Figures 2 and 3?

a. Stage 1, Grade 2
b. Stage 2, Grade 1
c. Stage 2, Grade 2
d. Stage 3

2) Which of the following infections is transmitted more commonly perinatally during the intrapartum period, versus hematogenously during gestation?

a. Herpes simplex virus (HSV)
b. Listeria monocytogenes
c. Parvovirus B19
d. Toxoplasmosis
e. Treponema pallidum

3) Which of the following gross and/or histologic features is most commonly associated with fetal vascular malperfusion?

a. Hypercoiled umbilical cord
b. Inflammation of the maternal decidua
c. Inflammation of villi
d. Small placental size
e. Thick umbilical cord

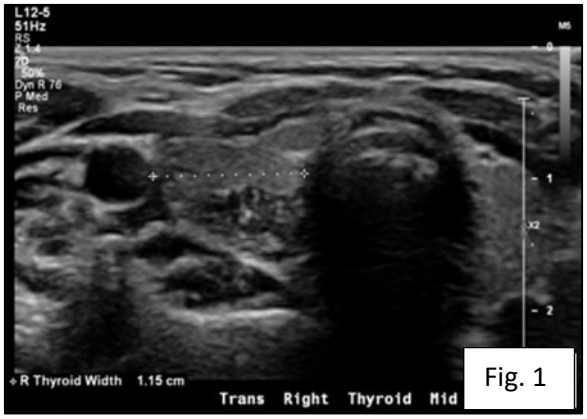


Fig. 1

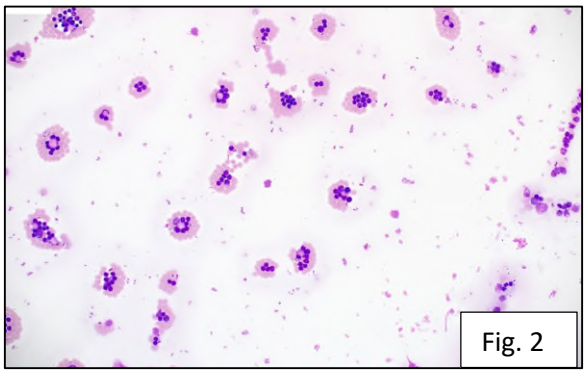


Fig. 2

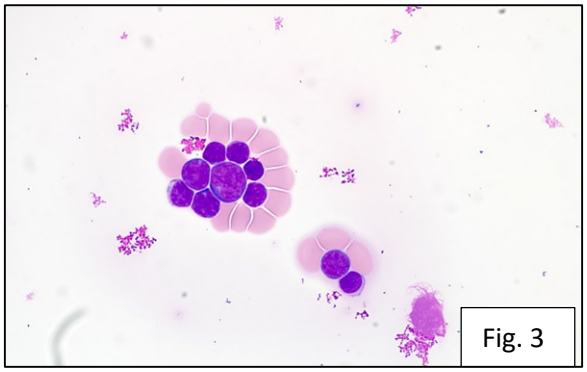


Fig. 3

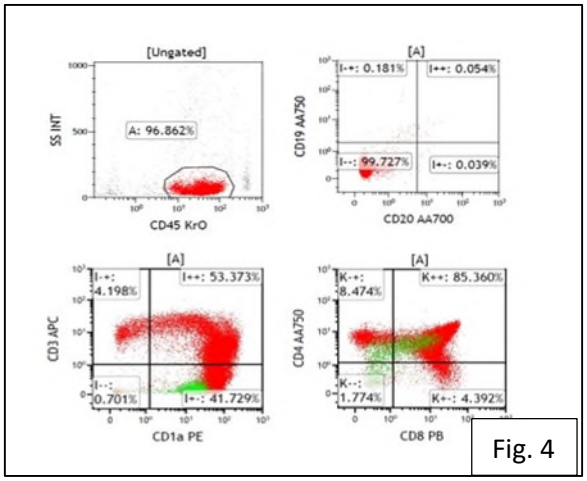


Fig. 4

SPP Slide Survey Case 23-10

A 6-year-old female with a past medical history significant for a right branchial cleft cyst presented for fine needle aspiration (FNA) of an incidentally identified right thyroid nodule. Ultrasound imaging (Fig. 1) showed a solid, hypoechoic, lobulated nodule, with sharp margins, 1.0 cm in greatest dimension, in the right lobe of the thyroid gland.

A whole slide image (Romanowsky) of a representative aspirate smear is provided (see also Figs. 2&3), as are selected flow cytometry scattergrams (Fig. 4).

Diagnostic List:
Please select the most likely diagnosis below:

a. Chronic lymphocytic thyroiditis (Hashimoto thyroiditis)
b. Ectopic intrathyroidal thymic tissue
c. Intrathyroidal lymph node tissue
d. Intrathyroidal parathyroid tissue
e. T-lymphoblastic leukemia/lymphoma (T-ALL/LBL)

1) Which of the following is an appropriate clinicopathologic association for chronic lymphocytic thyroiditis (Hashimoto thyroiditis) in pediatric patients?

a. Antithyroid peroxidase (TPO) antibodies are not detected; antithyroglobulin antibodies are detected in 80-90% of cases.
b. Associated with development of Burkitt lymphoma.
c. Female:male ratio is approximately 1:1.
d. Most pediatric cases are sporadic, though there is an increased incidence with HLA haplotypes DR3, DR4, and DR5.

2) Which of the following represents a phenotype characteristic of ectopic thymic tissue?

a. CD20 positive, with polytypic kappa and lambda light chain expression.
b. Complete loss of T cell markers CD3, CD2 and/or CD5.
c. Heterogeneous expression of CD4 and CD8, with subpopulations of cells with CD4 only expression, CD8 only expression, and CD4 and CD8 coexpression.
d. Homogeneous CD4 and CD8 coexpression.

3) Which of the following morphologic features is characteristic of parathyroid tissue?

a. Dual population of small-to-medium sized, discohesive cells with scant cytoplasm, smooth nuclear borders, and variably open chromatin and epithelioid-to-spindled epithelial cells with moderate amounts of cytoplasm.
b. Large, monolayered sheets of crowded cells with nuclear grooves, nuclear pseudoinclusions, and scattered psammoma bodies.
c. Mixed population of small, discohesive cells with scant cytoplasm and condensed nuclear chromatin, medium-sized cells with eccentric nuclei and perinuclear hofs, and clustered cells with abundant cytoplasm, large round nuclei, and centrally located nucleoli.
d. Round-to-ovoid cells arranged in loose two-dimensional groups, with stippled nuclear chromatin and without significant pleomorphism or mitotic activity.