Poster 01: **Hypertrophic Pachymeningitis as a manifestation of IgG4 Related Disease in a pediatric patient: A case oriented review**

Luna-Garcia, P [1,2]; Perez-Berenguer, Juan [1,2]; Labat, E [2,3]; Ferrer-Torres, L [2,4]; Zambrana-Flores, E [5]; Correa-Rivas, Maria [1,2]. [1] Department of Pathology and Laboratory Medicine, University of Puerto Rico, School of Medicine [2]Puerto Rico Medical Service Association [3]Department of Radiology, University of Puerto Rico, School of Medicine [4]Hato Rey Pathology Immunopathology Laboratory Division, Caguas [5]Pediatric Rheumatology, HIMA-San Pablo, Caguas

IgG4-related disease is a recently discovered disease that is histopathologically characterized by fibrosis and IgG4-positive plasma cell infiltration. This condition may involve any site of the organism with varying clinical presentation. The most recognized affected tissue is the pancreas, but it may also involve the biliary tract, salivary and lacrimal glands, kidneys, orbital tissues, lymph nodes, lungs, and others. More recently and in rare cases, it has been demonstrated to involve the central nervous system as a pattern characterized by hypertrophic pachymeningitis that may have compressive effects. We report a case of an 8-year-old male patient presenting with eyelid and parotid gland swelling that on MRI revealed multiple discrete intracranial extra-axial lesions causing severe stenosis with craniomedullary compression.

Poster 02: **Sudden Death Due to Uterine Rupture**

Lyvia Alvarez [1] ; Stephany Suarez [2]; **Frances M. Gomez[3]** . [1,2]Institute of Forensic Sciences of Puerto Rico [3]Department of Pathology and Laboratory Medicine, University of Puerto Rico, School of Medicine.

Gestational trophoblastic disease comprises a series of tumor and tumor-like conditions that have in common the proliferation of placental derived tissue. Here, we describe a case of a 22-year- old female that was found dead at her home. She had a prior history of a uterine curettage three months' prior to death with the diagnosis of hydatidiform mole. She refused follow up treatment after the curettage. The autopsy revealed blood in the abdominal cavity, and a ruptured uterus. Macro and microscopic findings reiterate the diagnosis of hydatidiform mole. In this article, we will present a case of an invasive hydatidiform mole of a 22-year old female.

Poster 03: **Plasmablastic lymphoma and its association with immunosuppression and HIV infection**

Jorge I. Chéverez Ocasio [1,2]; Román Vélez Rosario[1,2] ; Luz M. Rodriguez [3]
[1]Department of Pathology and Laboratory Medicine, University of Puerto Rico, School of Medicine, [2]Administración de Servicios Médicos de Puerto Rico, Anatomic Pathology Laboratory [3]Laboratorio de Patología Dr. Noy, Bayamon Medical Center

This is a case series and review of literature of plasmablastic lymphoma (PBL) in an HIV positive patient and in a patient with an unknown immunosuppressive status. The aim of this presentation is to review and discuss the key features and diagnostic criteria of this entity. PBL is a subtype of diffuse large B cell lymphoma (DBLCL) with a distinct blastic morphology. It was first described in 1997 as a series of aggressive non-Hodgkin lymphomas arising in the oral cavity of HIV positive patients. Since then, PBL has been strongly associated with HIV infection. Nevertheless, cases of PBL in patients with other immunosuppressive conditions are reported. Histologically, PBL is characterized by a diffuse proliferation of large neoplastic cells that resemble B immunoblasts or plasmablasts.

Immunophenotypically, it expresses plasma cell markers instead of B-cell markers typical of DBLCL. The Ki-67 proliferation index is often high, reflecting its aggressive biology. Epstein-Barr virus is frequently detected in the majority of cases. Therefore, differentiating plasmablastic lymphoma from other lymphoproliferative disorders requires immunohistochemical analysis.

Poster 04: **Symptomatic Female Carrier of Duchenne Muscular Dystrophy: Case report**

Jorge Asenjo Biaggi[1]; Juan Luis Perez Berenguer[2]; Edwardo Ramos Cortes [3]; Luis Ferrer Torres [2]; Aurelio Segundo Diaz[4] . [1]UPR-RCM, [2] Hato Rey Pathology Labs, [3] Instituto de Rehabilitación del Caribe, [4] HIMA Surgery Department

The dystrophinopathies are a group of diseases caused by mutations in the dystrophin gene. These include Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) which are X-linked recessive diseases and therefore expressed in males. Symptomatic female carriers of Duchenne muscular dystrophy (DMD), however, have been reported in the literature. Reported cases have demonstrated common characteristics in terms of genetic and histopathological presentation. Clinical manifestations can be variable, with some patients presenting with almost no symptoms and others presenting with symptoms as severe as those seen in affected males [3, 5]. Even in female manifesting carriers (MCs) that do not exhibit symptoms, signs of DMD can be present, such as elevated serum creatine phosphokinase (CPK) [9]. We present a case of a 7 y/o female patient with a history of elevated serum CPK levels, muscle biopsy with characteristic features of MCs, and genetic deletions in exons 46-48 of the DMD gene, a common mutation found in MCs.

Poster 05: **Cerebellar Liponeurocytoma: Case Report**

Brandon Torres Rivera [1,2], Román Vélez Rosario [1,2], Yannina Colón Sanchez [1,2], Juan Pérez Berenguer [1,2] . [1]Department of Pathology and Laboratory Medicine, UPR School of Medicine, [2]Puerto Rico Medical
Services Administration.

Cerebellar liponeurocytoma is a rare entity with cells showing neurocytic and lipomatous differentiation that tend to affect only adults. They are typically located in the cerebellum with a predilection for the cerebellar hemispheres. It has a low proliferative potential, but a high recurrence rate, therefore, it is classified as a WHO grade II tumor. The first case was reported in 1978 as a lipomatous medulloblastoma and since then, multiple terms have been used to describe it, the latest being cerebellar liponeurocytoma, adopted in the 2000 World Health Organization Classification. We present the case of a 58-year-old male patient with a right cerebellar cystic intra-axial tumor. Conclusion: Even though cerebellar liponeurocytomas share some features with malignant aggressive tumors, such as medulloblastomas, it is a distinct entity with a favorable prognosis.

Poster 06: **Ebstein Barr Virus Positive Mucocutaneous Ulcer, Report of One Case and Review of Literature**

Yannina M. Colón Sánchez [1,2]; Roman Velez Rosario [1,2]. [1]Department of Pathology and Laboratory Medicine, UPR School of Medicine [2]Puerto Rico Medical Service Administration.

Epstein Barr Virus (EBV) positive B-cell lymphoproliferative disorders are a spectrum of diseases that range from self-limiting, localized conditions to aggressive lymphomas. Epstein Barr virus is a widespread organism, attaining

asymptomatic lifelong carrier status in a large proportion of the world's population. Epstein Barr Virus Mucocutaneous Ulcer (EBVMCUs) is a rare lymphoproliferation that gained recognition as a new entity in the 2016 revisions to the World Health Organization Classifications and was first identified in 2010. This condition has been reported in the setting of iatrogenic immunosuppression, advanced age-associated immunosenescence and primary immunosuppression. Iatrogenic immunosuppression, being the most common risk factor has been linked to the use of immunosuppressive drugs, such as, methotrexate. We present the case of an 89-year-old woman who presented with multiple oral mucosal ulcers. Biopsy of EBVMCUs demonstrate surface ulceration with infiltrates of atypical lymphoid cells and can have similar histologic appearance with various entities such as Classical Hodgkin lymphoma and more aggressive lymphomas. However, it has a favorable prognosis as most cases regress spontaneously or with reduction of immunosuppressive therapy. Conclusion: The successful diagnosis and treatment of EBVMCUs rely on a multidisciplinary and patient-centered approach. Improvement in care coordination can prove pivotal for the diagnosis and outcomes of patients with this disease.

Poster 07: **Collision tumor; mantle cell lymphoma and acinar adenocarcinoma of the lung: A case report**

Jasmine Figueroa Díaz [1]; Gladys Cruz Santiago [2]; Laura Vega Vázquez [2]. [1] Department of Pathology and Laboratory Medicine, UPR School of Medicine, [2] Department of Pathology, San Juan VA Medical Center.

Collision tumors are defined by the presence of at least two tumors of independent origins within the same anatomical site. They are sharply demarcated and lack significant tissue admixture. The tumors may originate from the same organ or metastasize from other sites. They are rare, but well documented, and may be composed of benign or malignant tumors, most commonly a carcinoma and sarcoma or carcinoma and lymphoma. They have been encountered in various organs, including the gastrointestinal tract, lymph nodes, and lungs. Primary lung cancer with lymphoma at the same site is rare and, in most cases, the reported lymphoma is either mantle cell lymphoma (MCL) or extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). We present a rare case of mantle cell lymphoma and acinar adenocarcinoma of the lung within the same lung lesion. High suspicion and proper diagnosis are important to guide appropriate treatment.

Poster 08: **Unusual Biclonal IgA Plasma Cell Myeloma with Aberrant Expression of High-risk Immunophenotypes in Puerto Rico: First Report of a New Diagnostic and Clinical Challenge**

Carlos A. Monroig-Rivera[1]; Clara N. Finch Cruz[2]; Adalberto Mendoza[2] . [1]Ponce Health Sciences University, School of Medicine, [2]Southern Pathology Services, Inc.

Introduction: Biclonal IgA plasma cell myelomas (PCM) are rare (<2% of PCM). Therefore, no literature reports are found on immunophenotypes associated with high-risk disease in patients with unusual PCM. Hence, we report the first case of a Puerto Rican patient with biclonal IgA PCM expressing high-risk immunophenotypes and analyze its clinical relevance. Case Presentation: A 65-year-old Puerto Rican female with acute abdominal pain was admitted to the Hospital due to laboratory tests showing hypercalcemia, increased blood urea nitrogen and creatinine levels. Peripheral blood evaluations showed normocytic anemia, slight leukoerythroblastosis and rouleaux formation. Serum protein electrophoresis with

immunofixation (PEP/IFx) found high IgA levels and detected a biclonal IgA gammopathy with Kappa specificity. Urine PEP/IFx detected increased Ig in the IgA region without a well defined band. Bone marrow aspiration and biopsy samples showed hypercellularity, increased abnormal plasma cells (32%) of intermediate-type morphology replacing over 50% of the marrow stroma. Immunohistochemistry revealed that plasma cells were positive for CD138, MUM1, for OCT-2 and c-MYC (both high-risk markers). Flow cytometry studies detected CD45-negative plasma cells expressing CD33, another high-risk marker. Upon discharged, the patient was lost to follow-up.

Poster 09: **Morphological Findings in Early Cardiac Deaths and Obtaining Genetic Material for SNP Characterization**

Arismendy Benítez [1]; Estefani Sánchez Marte [1]; Claudia Suero Guzmán [1]; Víctor Calderón [2]; Robert Paulino-Ramírez [1]. [1]Instituto de Medicina Tropical & Salud Global, Universidad Iberoamericana, Santo Domingo, República Dominicana. [2]Laboratorio Biotecnológico TIOTE BIOTECH

Background: Cardiovascular diseases are challenging for health systems for high mortality rate association, accounting for 31% of deaths worldwide. They lead majority of death causes among all ethnic groups in people under 65 years old. Most deaths include cerebral vascular and ischemic diseases, heart failure, and arterial hypertension. Modifiable and non-modifiable risk factors have been substrates for genetic studies seeking to determine the causal mutations. The purpose of this study is to evaluate the morphological and molecular characteristics of early cardiac deaths in the Dominican Republic. Methods: Complete autopsy of cases was performed. We obtained data of morphologic findings from myocardium, coronary arteries, tissue processing and H&E staining. We extracted DNA from blood. Purification was performed with the Pure Link® kit of Invitrogen to evaluate the quality of the samples. Results: We examined 26 cases. The mean age of the cases was 50 years. The 27% corresponding to women, mean age was 38.5 years, among men (73%), mean age was 54 years. The average heart weight for women was 458.5 grams. For men the mean 583 grams. Areas of acute infarction appeared in men with 90%, compared to women with 2%, OR 45 with confidence index of 95% (3.3-604), p 0.00038. The volume of collected blood ranged from 1.5 to 4 ml. DNA was obtained in all the cases. Quality of the extracted material showed a higher ratio 260/280 nm. Conclusions: Deaths occurred predominantly in men. Presence of myocardial infarction was frequent. Good quality genetic material was obtained.

Poster 10: **Parasellar Paraganglioma, Case Report**

George Santiago Quiñones [1]; Juan L. Perez Berenguer [1]; Luis E. Ferrer Torres [2]. [1]Department of Department of Pathology and Laboratory Medicine, UPR School of Medicine, [2]Hato Rey Pathology Laboratory.

Background: Paragangliomas are neuroendocrine tumors of paraganglionic tissue which are extremely rare in the sellar area. Their clinical course characteristically begins with the appearance of endocrine disorders manifested by progressive hypophyseal insufficiency and visual field changes with progressive tumor extension. A specific feature of paragangliomas of the chiasma-sellar area is their rich vascularization, which is demonstrated by angiography and verified in all cases during operation. The rich vascularization of paragangliomas of the chiasma-sellar area calls for rapid and complete removal and poses a challenge for neurosurgical management and achievement of complete hemostasis. We present a case of parasellar paraganglioma with

suprasellar extension and involvement of the cavernous sinus on a 30 y/o male patient with visual changes and endocrinopathies for the past few months. Conclusion: Paraganglioma must be considered in the differential diagnosis of parasellar tumors, even when secretory symptoms are absent. The radiologic findings are obscure, and it is difficult to differentiate it from more common tumors of this region such as meningioma, schwannoma, and pituitary adenoma. Surgical excision followed by radiotherapy is the mainstay of management

Poster 11**: A case of atypical spindle cell lipomatous tumor in the parotid gland.**

María S. Oliver Ricart [1]; Román Vélez Rosario [1]; María De Lourdes Marín [2]. [1] University of Puerto Rico, Medical Sciences Campus, Department of Pathology and Laboratory Medicine [2] Hospital Oncológico Dr. I. González Martínez.

Atypical spindle cell lipomatous tumor is a benign adipocytic neoplasm, characterized by ill-defined tumor margins and the presence of mild to moderately atypical spindle cells, adipocytes, lipoblasts, pleomorphic cells, multinucleated giant cells, and a myxoid or collagenous stroma. It has a wide anatomical distribution occurring predominately in middle-aged males. It has a low tendency for local recurrence if incompletely excised. Unlike conventional atypical lipomatous tumors, there is no risk for dedifferentiation. Tumor cells variable express CD34, S100, desmin. Expression of MDM2 or CDK4 is rare. There is loss of RB1 nuclear expression in most cases. Atypical spindle cell lipomatous tumor's wide variety of microscopic appearances and histologic overlap with diverse mimics may make it a challenging diagnosis. We present a case of atypical spindle cell lipomatous tumor in a 49 year-old male with a left parotid gland lesion.

Poster 12: **Cardiac Rhabdomyoma- A pediatric autopsy case report**

Mirliam N. Saldoriga Merced [1,2], María Correa Rivas [1,2], Enrique Carrión Vargas [3], María del Carmen González Ríos[4]. [1]Department of Pathology and Laboratory Medicine, UPR - Medical Science Campus, School of Medicine, [2]Administración de Servicios Médicos de Puerto Rico, Anatomic Pathology Laboratory, [3]Department of Pediatrics Cardiology Section and [4]Department of Pediatrics Medical Genetic Section, UPR -Medical Science Campus, School of Medicine.

The most common pediatric cardiac tumor is rhabdomyoma; a rare benign neoplasia arising from the myocardium. Most of the cases are seen in children younger than 1 year old. They are highly associated with tuberous sclerosis complex (TS); 70% to 80% of infants with rhabdomyoma have a subsequently confirmed mutation in the TSC1 and TSC2 genes. The clinical manifestations of cardiac rhabdomyomas are determined by their size, multiplicity, location and whether they expand into a chamber. Larger rhabdomyomas can project from the ventricular wall or septum into the cardiac cavity, obstructing cardiac flow and valvular motion. In addition, they may disrupt the conduction system causing arrythmias. Cardiac rhabdomyomas usually present a complete or partial regression with consequent resolution of symptoms. We present a preterm adequate for gestational age (PTAGA) newborn with a diagnosis of cardiac masses, functional tricuspid stenosis, arrhythmia, and hydrops fetalis, that expired three weeks after birth. The autopsy confirmed multiple cardiac rhabdomyomas and raised a flag to evaluate other family members for the tuberous sclerosis complex, as no known previous family history was documented in the medical record.