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A Unusual Case of Classic Hodgkin Lymphoma (cHL) with Aberrant T-Antigens Expression

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the most common non-recommended indication. The majority of these studies were ordered by the hematology/oncology service, with only a third coming from others. Some possible causes for these inappropriate requests include ordering studies as a battery instead of a step-wise approach, and concern for missing an incipient/occult condition.

Essential Thrombocythemia Associated with Plasma Cell Neoplasm

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Introduction/Objective: The co-occurrence of essential thrombocythemia (ET) and multiple myeloma (MM), two distinct entities with distinct cellular origin, is rare, with a limited number of cases reported.
Methods/Case Report: We report a case of a 63-year-old male who initially presented with thrombocytosis, splenomegaly and elevated LDH. A bone marrow examination showed hypercellularity (90%) with increased abnormal megakaryocytes, as well as a JAK2 V617F mutation, with overall features consistent with involvement by a myeloproliferative neoplasm (MPN), consistent with ET (versus early primary myelofibrosis (PMF)). Additionally, 6% kappa-restricted plasma cells were identified, consistent with involvement by a monoclonal gammopathy of undetermined significance (MGUS). The patient was subsequently treated with hydroxyurea. Four years later, he presented with evidence of paraproteinemia. IgG kappa monoclonal paraprotein was elevated at 3.4 g/dL. A repeat bone marrow examination showed hyper-cellularity (60%), including clusters of abnormal megakaryocytes and mild to moderate reticulin fibrosis. The previously identified kappa-restricted plasma cell population increased to approximately 40% of the total cellularity. Cytogenetic analysis showed a normal male karyotype (46,XY[20]), and a prognostic myeloma-FISH panel including 13q-/-13, 1q32/1q21, p53/NF1, CCND1/IgH t(11;14), FGFR3/IgH t(4;14), IgH/MAF t(14;16) and IgH/MAFB t(14;20) was negative for all tested abnormalities. The overall features were again consistent with involvement by an MPN and progression of the previously identified MGUS.
Results (if a Case Study enter NA): NA.
Conclusion: Only a few cases of concurrent ET and MM have been previously reported in the literature, with most of these cases having a temporal association with alkylating agent therapy. However, MM development has also been reported in a patient with non-cytotoxic treatment of ET. In contrast, our patient was diagnosed with ET associated with MGUS at the initial diagnosis. Notably, the co-existence of early PMF with MM appears to be relatively more established. A large study showed that previous PMF was strongly associated with MM development (OR 24.3; 95% CI:2.9-201.5).

Focal Myositis with CD8+T-Cell predominance: an Inflammatory Myositis Mimicking a Soft Tissue Neoplasm

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Introduction/Objective: FM is a rare self-limiting T-cell rich lesion arising within muscles of young adults as a solitary lesion and can be confused with a variety of neoplastic/inflammatory conditions or lymphoma. Here we describe a rare T-cell rich variant of FM with CD8 predominance.
Methods/Case Report: A 51-year-old female presented with two-month history of left trapezius swelling. MRI showed an enhancing tumor within the muscle, suspicious of sarcoma, less likely myeloma.
Results (if a Case Study enter NA): Biopsy showed diffuse infiltration of small lymphocytes in a fibrotic background and atrophic skeletal muscle. These lymphocytes stained positive for CD2, CD3, CD7, CD8, CD5 (partial), TIA1 (partial), CD43, CD57 and negative for CD4, CD30, CD56, CD57, granzyme B, perforin, BCL-6, BCL-2, Pax5, TCL1, and CD56. Ki-67 was (<10%) with few background CD20+ B cells. By flow cytometry, the CD8+ T-cells co-express CD2, CD3, CD7, with dim CD5 expression. TCR gene rearrangement study by PCR showed no clonal TCR Gamma gene rearrangement.
Conclusion: FM is extremely rare with only 22 cases well-described in the literature, all predominantly composed of CD4+ T-cells, clinically concerning for low-grade sarcomas, as inflammatory myofibroblastic tumor, inflammatory leiomyosarcoma, or liposarcoma. Careful analysis is essential for correct diagnosis. Mimics include other causes of myositis, such as polymyositis and inclusion body myositis. Depending on the extent of lymphocytic infiltrate, gene rearrangement studies might be necessary to rule out clonality. Recognition of this rare entity with excellent prognosis is crucial to provide appropriate management and avoid unnecessary, aggressive procedures.

A Unusual Case of Classic Hodgkin Lymphoma (cHL) with Aberrant T-Antigens Expression

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Introduction/Objective: T-cell-associated antigens (TCA) are rarely expressed by Hodgkin/Reed-Sternberg (HRS)

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cells of classical Hodgkin lymphoma (cHL) and is associated with uncertain clinical significance.

Methods/Case Report: We present a very recent case of a 68 year-old female with no significant past medical history presented with a large right groin mass for 1 month. She denied any fever, night sweats, fatigue or weight loss. CT abdomen/pelvis showed inguinal/pelvic lymphadenopathy. Resection showed effaced nodal architecture with a vague nodular appearance and with large, atypical nucleolated cells in a background of small, mature lymphocytes and histiocytes. Eosinophils are conspicuously absent in the background. The large cells were polylobated lymphocyte and histiocyte rich (L & H) like, classic HRS cells as well as multinucleated in appearance. The morphologic features overlapped between Classic Hodgkin Lymphoma, and Nodular Lymphocyte Predominant Hodgkin Lymphoma. Given the immunohistochemical (IHC) features of the neoplastic cells [CD45 negative, PAX5+ (weak), CD30+ (strong), CD15+ (majority) and MUM1 +] the diagnosis was most compatible with CHL. Interestingly, a subset of the Hodgkin cells (10-20%) aberrantly expressed a number of T-cell markers (CD2, CD3, CD4, and CD8). However, the background smaller T-cells did not show cytologic atypia or aberrancies of T-cell antigen markers. The large cells were negative for CD43 and ALK1. TCR gamma gene rearrangement was also negative for clonal T-lymphocytes, thus excluding the possibility of peripheral T-cell lymphoma with Hodgkin-like cells. Based on the morphology and IHC profile, this case proves to be CHL with multiple TCA expression including CD2, CD3, CD4 and CD8. She was planned for chemotherapy. Four months after the initial diagnosis and after completion of 2 cycles of chemotherapy, her PET CT scan showed improvement with right inguinal adenopathy being smaller in size and decreased avidity, indicating a response to treatment. She is due for 2 more cycles. Till todate she is doing well with no further symptoms.

Results (if a Case Study enter NA): NA.

Conclusion: So far reported cases of classic Hodgkin lymphoma (CHL) with aberrant T-cell antigen (TCA) expression were associated with decreased event-free survival and overall survival when compared with TCA-negative CHLs. Our finding contributes to the literature and improves our knowledge of the disease.

Intracranial Extramedullary Hematopoiesis: a Rare Entity

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Introduction/Objective: Extramedullary hematopoiesis (EMH) is characterized by ectopic proliferation and differentiation of hematopoietic precursor cells outside the bone marrow. In postnatal life, EMH occurs due to hematopoietic disorders, infection or advanced tumors and is usually seen in the spleen, liver, and lymph nodes. Intracranial EMH is an extremely rare entity that has been reported to be associated with hematological disorders, meningioma, subdural hematoma and pilocytic astrocytoma.

Methods/Case Report: To highlight the significance of identifying EMH as one of the differential diagnoses of brain lesions, we present a 67-year-old female with a history of headaches and generalized malaise. Her past medical history was significant for hypertension and mild anemia. As part of the headache workup, magnetic resonance imaging (MRI) of the brain showed a 3.7cm cystic mass in the right inferomedial parietal lobe with medial mural enhancement, associated with edema and mass effect. A similar 3mm left parietal lobe enhancing focus was also noted. These findings were suggestive of a hemangioblastoma. Resection of the larger mass showed a benign vascular lesion with endothelial hyperplasia without the appearance of cavernous hemangioma or vascular malformation. Foci of erythroid and myeloid precursors, and megakaryocytes were seen. Immunohistochemical stains were negative for GFAP, CK AE1/AE3 and inhibin ruling out glial neoplasms, metastatic carcinoma and hemangioblastoma. Hematopoietic cells were positive for CD45, while immature red cells were positive for E-cadherin and endothelial cells were positive for CD31, consistent with EMH. Further workup was negative for any underlying hematological disorders, resulting in the diagnosis of idiopathic intracranial EMH. The patient received intensity- modulated radiation therapy (IMRT) for her remaining left parietal lesion. After 3 months of regular follow-up, her headache and malaise significantly improved.

Results (if a Case Study enter NA): NA.

Conclusion: To our knowledge, this is the first reported case of idiopathic intracranial EMH.

Immunoglobulin G4-Related Disease (IgG4-RD) Mimicking Hyaline Vascular Castleman Disease

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Introduction/Objective: Immunoglobulin G4-related disease (IgG4-RD) is a newly emerging immune-mediated fibroinflammatory disease characterized by increased IgG-4+ plasma cells. In soft tissue and visceral organs, the IgG4+ plasma cells often accompany a storiform pattern of fibrosis with obliterative phlebitis. In lymph nodes, rare cases have been described in the literature mimicking Multicentric Castleman Disease.

Methods/Case Report: Herein, we present a case of a 59-year-old male with history of lung adenocarcinoma status post resection one year prior, who presented with several months of unintentional weight loss and worsening

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