Journal of Community Hospital Internal Medicine Perspectives

Volume 13 | Issue 6 Article 28

2023

Maryland ACP Chapter Winners at Mulholland Mohler Residents Meeting - 2023

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Recommended Citation
Woodward, Maryellen (2023) "Maryland ACP Chapter Winners at Mulholland Mohler Residents Meeting - 2023," *Journal of Community Hospital Internal Medicine Perspectives*: Vol. 13: Iss. 6, Article 28.
DOI: 10.55729/2000-9666.1265
Available at: https://scholarlycommons.gbmc.org/jchimp/vol13/iss6/28

JCHIMP

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Journal of Community Hospital Internal Medicine Perspectives 2023;13:129–131
https://doi.org/10.1016/j.clnesp.2023.09.009

The Not So Benign Sickle Cell Trait: A Case of Renal Medullary Carcinoma

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Introduction: Sickle cell trait (SCT) has long been considered a benign carrier state with malarial protection. However, carriers may be affected by increased venous thromboembolism, exercise-related injury, renal complications and very rarely, a fatal renal malignancy.[1,2]
Case: 32-year-old-female with history of asthma and SCT presented to the ED with one-month history of cough, hemoptysis, left flank pain and gross hematuria. She denied fevers, chills, night sweats, mouth sores, joint pain, or rashes. Vital signs were stable with physical exam notable for left flank tenderness. Admitting labs revealed WBC 7.55 x10^3/μL, Hb 11.9 g/dL, Cr 0.67 mg/dL, ESR 40mm/hr, CRP 3.58mg/dL, procalcitonin <0.09ng/mL with urinalysis notable for protein 100mg/dL, large blood, too numerous to count RBCs, negative leukocyte esterase/nitrite with few bacteria. CT chest imaging showed scattered ground glass and nodular opacities with mediastinal and hilar adenopathy. CT abdomen without contrast showed large lobulated soft tissue structures in retroperitoneum suspected to be lymph nodes. Pulmonary-renal syndrome was considered, but renal function was normal. HIV and hepatitis viral serologies were negative with normal serum complement levels. ANCA, anti-GBM Ab and aspergillus specific IgE all resulted as negative. MRI renal mass protocol revealed an 8cm mass arising from the left kidney with extensive lymphadenopathy compressing the left renal vein. Renal biopsy revealed renal medullary carcinoma with PET/CT scan showing multiple pulmonary and osseous metastases. Combination chemotherapy was initiated. Following a protracted course, she expired three months after diagnosis.
Discussion: Renal medullary carcinoma (RMC) is an aggressive form of non-clear cell renal carcinoma that is extremely rare accounting for <0.5% of all renal carcinomas such that true incidence is not well defined[2]. It was first described in 1995 with a case series of 34 patients, 33 of whom had SCT[3]. Since then, a handful of cases have been described in sickle cell disease[2]. It occurs almost exclusively in sickle cell trait such that when the diagnosis occurs without it, the term renal cell carcinoma, unclassified with medullary phenotype, is used[4]. The tumor arises from the renal papillae or epithelium of calyx. The proposed pathogenesis of transformation to RMC includes hypoxia of the renal medulla which increases expression of VEGF[2]. This hypoxia creates a favorable environment for breaks in DNA with frequent loss of transcription factor SMARCB1/INI1[2]. Median age of onset is 24 years with males being affected more frequently.[5] The common initial symptoms are hematuria and flank pain with most patients having metastatic disease by diagnosis.[2] Management options include radical nephrectomy, palliative chemotherapy and radiotherapy.[2] Survival remains poor despite chemotherapy with median survival from diagnosis of 4 months with metastases and 17 months without metastases.[2,5]
Conclusion: Sickle cell trait while largely benign; can be associated with a rare but fatal renal medullary cancer. Renal medullary carcinoma should be considered when SCT patients present with hematuria and flank pain.

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Getting the GIST: Hemoperitoneum with an Unusual Diagnosis

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Introduction: Gastrointestinal stromal tumors (GIST) account for 0.1 - 3% of all gastrointestinal malignancies.[1] We discuss an interesting case of undiagnosed GIST that presented with spontaneous hemoperitoneum.
Case presentation: A 62-year-old male presented with a 1-day history of diffuse abdominal pain and nausea leading to a syncopal event. A review of systems was notable for intentional weight loss, and night sweats but negative for abdominal trauma, or GI bleeding. On presentation, he was markedly hypotensive, and a physical exam revealed abdominal distension with diffuse rebound tenderness. Initial lab studies revealed a hemoglobin of 10.6g/dl. A CTA abdomen showed dense hemorrhagic ascites, and a peripherally enhancing exophytic

mass adjacent to the gastric fundus. The patient underwent a EUS and FNA which showed a hypoechoic irregular lesion vs hematoma in the perigastric and perihepatic peritoneal space. The patient had another syncopal event with a brief period of cardiopulmonary arrest requiring resuscitation raising concern for hemorrhagic shock. Repeat CTA revealed worsening hemoperitoneum with contrast extravasation in the left upper quadrant probably representing a pseudoaneurysm within the mass. The patient underwent emergent exploratory laparotomy with partial gastrectomy with removal of attached mass and evacuation of 1700cc of hemoperitoneum. Surgical pathology revealed a 5.1 cm GIST, spindle cell type with focally disrupted capsule, and low mitotic rate (<5/5mm2) suggestive of low risk of malignancy. The immunohistochemical stain was positive for CD34 and C117, supporting the diagnosis of GIST.

Discussion: The clinical presentation of GIST varies based on tumor location and size. The most common presentations are anemia, weight loss, and gastrointestinal bleeding. Other rare presentations include intraperitoneal hemorrhage, rupture and peritonitis.[1] Rapid tumor growth with necrosis, neovascularization due to the release of vascular growth factors, underlying coagulopathy, and thrombotic microangiopathy have all been implicated in causing spontaneous hemoperitoneum.[2] The presence of a well-rounded exophytic mass on imaging should suggest the diagnosis of GIST. Considering that only a few cases of GIST with intra-peritoneal hemorrhage as their presenting diagnosis have been reported in the literature, awareness of such an entity is crucial for prompt multidisciplinary management of these emergencies.

Conclusion: It is pertinent to keep an undiagnosed GIST in the differential while working up a case of spontaneous hemoperitoneum without an obvious underlying cause.

Assessing Rituximab Durability and Clinical Relapse in Patients with Thrombotic Thrombocytopenic Purpura

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Introduction: Immune thrombotic thrombocytopenic purpura (iTTP) is a rare disorder caused by an autoantibody mediated deficiency of ADAMTS13 and characterized by life threatening episodes of thrombocytopenia, microangiopathic hemolytic anemia and ischemic organ injury. Rituximab, an anti-CD20 monoclonal antibody is effective in reducing relapse rates and maintaining longer periods of clinical remission. This study aimed to compare durability of rituximab response (measured relapse free survival) in first versus subsequent episodes of iTTP based on the clinical observation that patients with multiply relapsing disease may be less responsive to rituximab.

Methods: We used the United States Thrombotic Microangiopathy Consortium retrospective iTTP registry that includes data from adult patients with iTTP treated at 15 high volume academic centers. iTTP episodes treated with rituximab and with at least 1 year follow up were included. We used cox regression to evaluate factors associated with relapse and Kaplan-Meier survival curves were generated to compare relapse-free survival after the first rituximab treated episode versus subsequent courses of rituximab.

Results: From 2004 – 2020, we identified 384 iTTP episodes treated with rituximab (310 first, 53 second, and 21 were 3rd to 5th courses). Median relapse free survival was higher after the first course of rituximab compared with subsequent (2nd-5th) courses of treatment (Figure 1). Overall, second or subsequent rituximab course [HR 2.8 (95% CI 1.53-5.10), first course as reference category] and race [White: HR 0.32 (95% CI 0.17-0.59), Black as reference category] were associated with relapse free survival in a Cox regression model adjusted for age, sex, overall number of iTTP episodes and lactate dehydrogenase.

CONCLUSIONS: Despite's rituximab's known benefits of prolonging relapse-free survival in iTTP, the durability of rituximab effect declines with multiple episodes/treatment courses with a pronounced 'wearing off' effect. White patients also had longer relapse free survival than Black patients. The mechanisms underlying this phenomenon need further study, but possibilities include reduced B cell dependence and evolution of plasma cells that produce anti-ADAMTS13 antibodies. Our results highlight that patients who have had 2 or more episodes may need closer monitoring, sooner retreatment, and consideration of alternative immunosuppression.

Accuracy of POCUS Jugular Venous Pressure Assessment to Predict Right Atrial Pressure Performed by Medical Students

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Purpose: Recent evidence suggests point-of-care ultrasound assessment of jugular venous pressure (uJVP) is highly accurate for estimating right atrial pressure (RAP) when performed by cardiologists. The primary objective of this study was to determine the reproducibility of these results and operating characteristics of the uJVP and ultrasound abdominojugular test (uAJT) when performed by novice POCUS users.

Methods: Two medical students completed a 12-hour POCUS training session. Inclusion criteria were adult patients (18 years or greater), referred for right heart catheterization and capable of giving informed consent. Exclusion criteria included critical illness, congenital heart disease, inotropic therapy, right internal jugular (RIJ) venous catheters, prisoners, and pregnant patients. The uJVP was measured at the RIJ vein by finding the meniscus of blood, and was a positive test if that column was greater than or equal to 8 cm above the right atrium. The top of the meniscus from sternal angle was measured and 5 cm was added to this measurement to reflect the distance from the right atrium in the supine position at 30° to 45°. The uAJT was positive if the blood column increased by 4 cm with standard abdominal pressure for 10 seconds. Significance between variables in the primary objective was assessed using Pearson's correlation. Agreement between different examiners was assessed using Cohen's kappa.

Results: A total of 107 patients were recruited and signed consent during the 7-week study period. Eight patients were removed from the analyses due to cancellation of cardiac catheterization, positive COVID-19 status, patient refusal, or difficulties with ultrasound visualization. The total number of patients included in the analyses was 99, with a mean age of 65 years, 62% were male, Black (45%) and 70% of patients had congestive heart failure. The correlation between an elevated uJVP and uAJT for RAP $\geq$ 10 was AUC 0.673 (0.563-0.783) and AUC 0.626 (0.497-0.755) respectively. The correlation for uJVP and uAJT for PCWP $\geq$18 was AUC 0.64 (0.526-0.754) and AUC 0.75 0.699 (0.583-0.815) respectively. For patients with two examiners, there was poor agreement in measurement of uJVP (Cohen's kappa 0.21) but better agreement with uAJT (Cohen's kappa of 0.74).

Conclusion: In our study, ultrasound IJ assessment was less reliable to accurately estimate RAP than in a recent study. Only 36% of patients were referred to RHC for the indication of volume assessment as

opposed to Wang et al. where 64% were referred for this indication. Limitations include a single medical system with two medical students. POCUS may prove to be a valuable tool for volume assessment but the optimal training for uJVP and uAJT has not yet been determined.

Apical Hypertrophic Cardiomyopathy with an Unclassical Variant of MYH7

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Apical Hypertrophic Cardiomyopathy (AHCM) is an atypical variant of hypertrophic cardiomyopathy with nonobstructive hypertrophy of the apex[1]. It accounts for 25% of HCM cases among the Asian population and 1-10% among the non-Asian population[2]. Like other HCM types, it is associated with an autosomal dominant mutation in sarcomere protein genes most often affecting myosin heavy chain (MYH7) or myosin binding protein C3 genes[3].

A healthy 52-year-old South Asian male presented with an incidental abnormal EKG prior to participation in a clinical trial, which revealed stable ST depressions and T wave inversions in the lateral leads and voltage criteria for Left Ventricular Hypertrophy (LVH). Transthoracic echocardiography (TTE) revealed left ventricular apical thickness without left ventricular outlet obstruction and preserved ejection fraction. Cardiac magnetic resonance (CMR) image revealed mild focal interstitial expansion, endocardial scarring, and thinning of the apex without aneurysm formation. An exercise stress test confirmed reduced tolerance with an appropriate response in heart rate but a blunted blood pressure response. Angiogram was unremarkable for significant coronary artery disease. Genetic testing revealed an unclassified variant in the MYH7 gene and screening with surveillance was recommended in first degree relatives.

Although AHCM does not have pathognomonic clinical symptoms, typically patients experience mild exercise intolerance. Typical EKG findings include symmetrical deep negative T waves ($\geq$ 1 mV) in precordial leads associated with high QRS voltage and though characteristic, are not required for diagnosis. TTE or CMR confirms diagnosis with focal apical hypertrophy greater than 15mm distal to the papillary muscles[1]. The management is similar to HCM. Our patient noted improvement in exercise tolerance with beta blockers to optimize diastolic filling and may benefit from surveillance EKG and TTE to evaluate for arrhythmias, left ventricular end-diastolic volume and apical aneurysm. AHCM is a novel diagnosis in South Asians and overall benign with a favorable prognosis. However, warrants consideration in healthy individuals with exertional dyspnea or those with arrhythmias, heart failure and sudden cardiac death. Asymptomatic screening and genetic testing in first degree relatives is recommended for early detection and consideration for medical, ablative or surgical interventions[5]. There is a need for more prospective studies to help identify markers for an early diagnosis.