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Follicular Mucinosis and Cutaneous T-Cell Lymphoma Association: A Clinicopathologic and Molecular Analysis of 19 Cases

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Follicular Mucinosis and Cutaneous T-Cell Lymphoma Association: A Clinicopathologic and Molecular Analysis of 19 Cases

Abstract

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Table 1. Mean, standard deviation, and IQR of common gastroenterological procedure costs comparing all, academic, and private hospitals.

	US Complete	CT Abdomen with Contrast	MRI Abdomen with Contrast	Liver Elastography	EGD (outpatient)	Colonoscopy Screening	Liver Biopsy IR	Liver Biopsy Percutaneous
All Hospitals								
Mean	742.13	2579.98	3494.29	172.76	3141.28	4617.89	952.68	6717.58
Standard Deviation	602.41	1772.17	1793.03	48.09	1446.46	4518.85	524.26	4298.29
IQR	513.63	1296.03	2532.48	25.74	2564.17	2280.58	454.03	5675
Academic Hospitals								
Mean	971.49	3001.66	3608.17	162.35	3665.42	4416.29	952.68	9907.5
Standard Deviation	626.50	2555.68	2093.95	53.09	1584.54	2163	524.26	2741.45
Private Hospitals								
Mean	673.69	2211.02	3394.66	204	3050.18	4994.16	N/A	5441.61
Standard Deviation	574.66	591.04	1627.06	N/A	1390.85	5594.19	N/A	4325.64

#62. The GIPP Never Lies: A Case of Refractory Ulcerative Colitis

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Mentor: Kathryn Hutchins

Program: Internal Medicine

Type: Case Report

Background: *Listeria Monocytogenes* is a rare infection with approximately 800 confirmed cases per year. The pathogen can present as a mild infection but can be more severe in immunocompromised patients. We present a case of invasive listeriosis in an ulcerative colitis patient.

Case: A 58-year-old male with a past medical history significant for refractory ulcerative colitis (UC) presented to the emergency department after 3-days of worsening

hematochezia and abdominal pain. Patient had transitioned to ustekinumab from infliximab and started a 40 mg prednisone taper two weeks prior due to refractory UC symptoms. On admission, the heart rate was 125, blood pressure was 82/55, and temperature was 102.1. Pertinent labs included white blood cell count 20.8, procalcitonin 0.37, lactic acid 1.7, and CRP <0.5. Patient was given fluids and broad-spectrum antibiotics along with methylprednisolone. CT significant for mild to moderate mural thickening in distal colon and rectum. Gastrointestinal pathogen panel (GIPP) was negative. Flexible sigmoidoscopy revealed diffuse disease consistent with active ulcerative colitis the following day. Two days after admission, 2/2 blood cultures

were positive for *Listeria Monocytogenes*. Infectious disease was consulted, and the patient was treated with ampicillin and gentamicin. Following treatment initiation, hematochezia decreased, and abdominal pain subsided. Patient was discharged on amoxicillin with a continued prednisone taper and mesalamine suppositories without recurrence of symptoms.

Conclusion: Patients on immunosuppressive therapy are at higher risk of developing listeriosis. Clinicians should be aware that listeria is not a tested pathogen on GIPP and can present as invasive diarrhea.

#63. Exploring the Efficacy of Pulmonary Artery Pressure Monitoring in Rural LVAD Patients: A Retrospective Cohort Study on Clinical Outcomes

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Program: Internal Medicine

Type: Original Research

Background: Rural healthcare challenges impede equitable care of advanced heart failure patients with left ventricular assist devices (LVAD) due to poor access to specialized resources. Remote pulmonary artery pressure (PAP) monitoring offers a potential solution to improve clinical outcomes in these patients.

Methods: A retrospective cohort study was conducted at an academic tertiary care center on adults 19 years or older implanted with LVADs between January 2015 and May 2022. Participants included resided in counties with populations under 50,000 as per the 2020 census. The study used the CardioMEMS PAP monitor by Abbott Laboratories in Abbott Park, Illinois. The primary outcome was the rate of right heart failure hospitalizations, with secondary outcomes including time-to-event of first right heart failure hospitalization post-LVAD implantation and overall survival.

Results: A total of 156 patients underwent LVAD implantation during the study period. Twenty-four patients had concurrent PAP monitors in place. The PAP monitor group showed a higher mean hospitalization rate for right heart failure (1.1383 per patient year) compared to the non-PAP monitor group (0.5024 per patient year) with a significant p-value of 0.0481. The PAP monitor group had a significantly shorter time-to-event of first right heart failure hospitalization (p=0.0162) (**Figure 1**). No statistically significant difference in survival was noted between the groups (p=0.2849).

Conclusion: PAP monitor implantation in this rural patient cohort does not demonstrate clinical benefit in heart failure outcomes. Certain statistical observations may be influenced by selection bias in this retrospective study, potentially favoring the preferential choice of more complex patients for PAP monitor implantation.

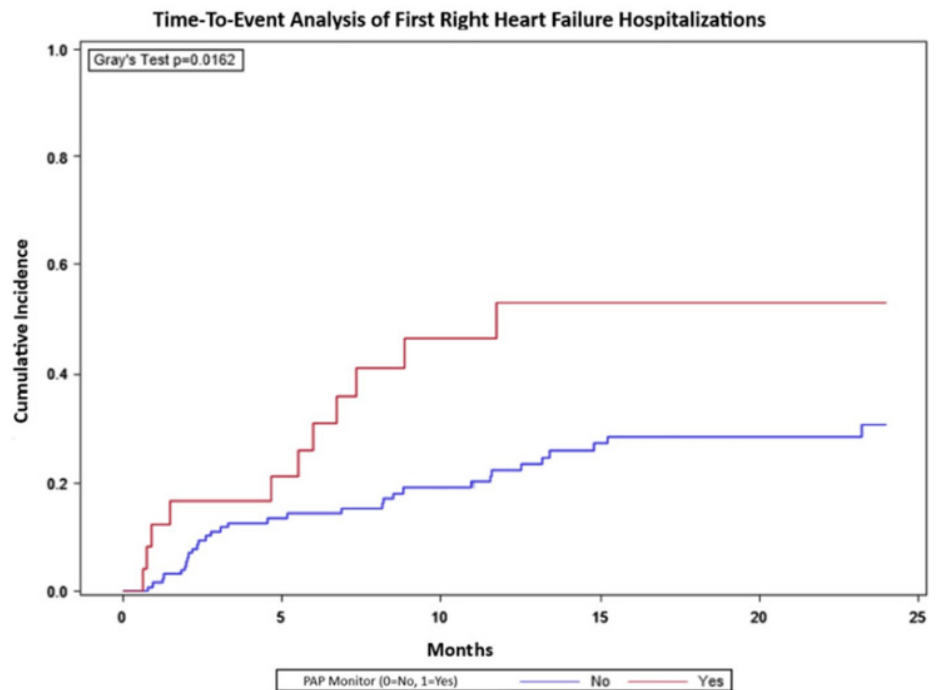


Figure 1. A visual depiction illustrating the time-to-event analysis for the occurrence of the initial hospitalization due to right heart failure after LVAD implantation.

#64. Looking at ANCA Vasculitis

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Mentor: Felipe S. Naranjo

Program: Internal Medicine – Nephrology

Type: Case Report

Background: Anti-neutrophilic cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a group of autoimmune diseases that can cause inflammation and damage to small blood vessels throughout the body. Inflammatory eye disease is described in 50% to 60% of patients with AAV, and for 8% to 16% of patients, it is an initial manifestation.

Case: A 74-year-old man with a history of chronic sinusitis and chronic kidney disease (G3a) presented to clinic with worsening

kidney function and proteinuria without hematuria. He was seen by Ophthalmology a few months prior for scleritis and was treated with a course of NSAIDs, with symptomatic improvement. Kidney biopsy showed a lesion of focal segmental sclerosis with obliteration of the capillary lumen, mild interstitial fibrosis and tubular atrophy, immunofluorescence was negative and electron microscopy with rare small dense deposits. The patient was started on SGLT-2 inhibitors. Renal function continued to deteriorate. The patient's condition was complicated by acute bronchitis, diffuse anterior scleritis, acute kidney injury, labs revealed positive ANCA antibodies with elevated PR3 antibodies. Repeat kidney biopsy revealed necrotizing crescentic

glomerulonephritis. He was treated with IV steroids, rituximab and ultimately required dialysis.

Conclusion: Renal involvement is a significant feature of AAV and can lead to acute kidney injury or chronic renal failure if left untreated. Clinicians should consider AAV in the differential diagnosis of patients presenting with scleritis, especially in those with renal dysfunction even with partial or complete resolution in ocular symptoms. Early diagnosis and treatment are essential in preventing irreversible organ damage and improving patient outcomes.

#65. Acute Gastroparesis Following Atrial Fibrillation Ablation

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Mentor: Jason Payne

Program: Internal Medicine

Type: Case Report

Background: Gastroparesis is a seldomly reported complication of atrial fibrillation

ablation but reports of less severe gastrointestinal symptoms are more common reported as high as 50% of the time. The mechanism of injury is believed to be collateral periesophageal vagal nerve injury.

Case: A 67-year-old male presented with

progressively worsening chest pain, abdominal distention and nausea six days after pulmonary vein isolation with radiofrequency ablation for atrial fibrillation (AF). Esophageal temperature monitoring was utilized throughout ablation and peak temperature was 36.2C. Prior to ablation he had no

gastrointestinal symptoms. CT abdomen and pelvis showed a massively dilated stomach (**Figure 1A**) compared to previous CT (**Figure 1B**) within a year prior to AF ablation with normal appearing stomach. A nasogastric tube was placed for decompression and gastroenterology was consulted. He underwent esophagogastroduodenoscopy (EGD), showing patent pylorus and large amount of gastric contents, which was suctioned with no evidence of mechanical obstruction. He was managed with metoclopramide, liquid diet and fully recovered clinically.

Conclusion: Given the proximity of the vagal nerve to the pulmonary vein, we believe that there was collateral insult to the vagal nerve leading to his acute gastric atony. Our case highlights a rare complication of acute gastric atony following pulmonary vein isolation and should be on the differential for patients presenting with acute gastrointestinal symptoms following AF ablation.

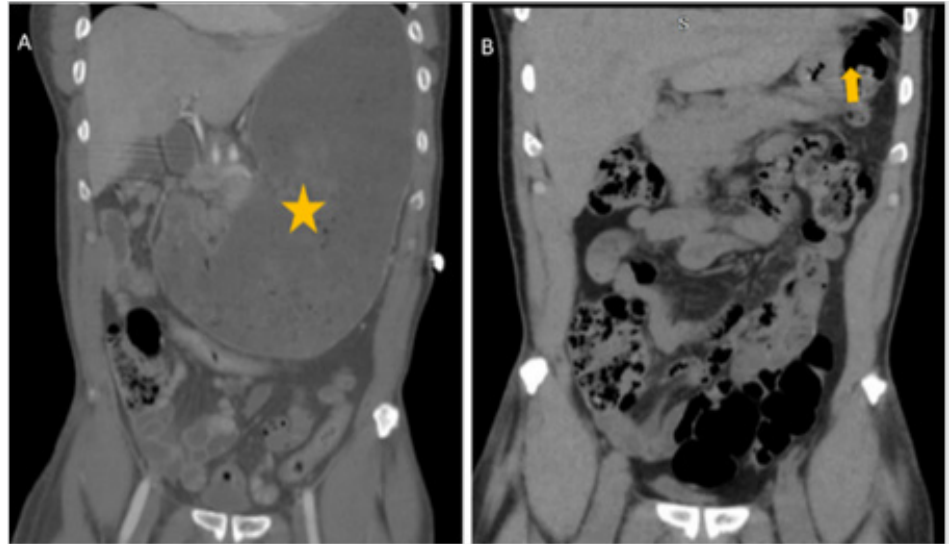


Figure 1. CT abdomen and pelvis coronal view showing large stomach (yellow star) with undigested food content (**1A**) compared to similar image a year prior to AF ablation with normal appearing stomach (yellow arrow) (**1B**).

#66. When Inflammatory Arthritis Gets Rash, Consider PsAPASH

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Mentor: Amy Dreessen

Program: Internal Medicine

Type: Case Report

Background: PsAPASH – psoriatic arthritis, acne, pyoderma gangrenosum, and hidradenitis suppurativa – is an autoinflammatory syndrome caused by innate immune system dysfunction, resulting in significant inflammatory arthritis, skin lesions, and joint pain. This rare syndrome and similar “P-syndromes” are a diagnostic puzzle and require a multidisciplinary approach for prompt recognition and treatment.

Case: A 41-year-old man presented with 6 years of chronic bilateral knee pain and

tender skin lesions. He endorsed joint pain and fatigue but denied fever, chills, and night sweats. Examination revealed tachycardia, severe cribriform scarring over his face, axillae, and groin, ulcerated and open groin lesions, diffuse muscle wasting with proximal muscle weakness, large bilateral knee effusions without warmth or tenderness, and multiple metatarsophalangeal joints with subluxation. Initial labs showed an elevated erythrocyte sedimentation rate, elevated c-reactive protein, leukocytosis, and an elevated protein gap. Bilateral knee x-rays and foot x-rays demonstrated multifocal osseous erosions with large knee joint effusions and polyarticular inflammatory arthritis in multiple metatarsophalangeal and interphalangeal joints. The patient

declined joint aspiration. Rheumatology was consulted, and additional infectious and rheumatologic workup was within normal limits. Dermatology was consulted and felt his skin manifestations were consistent with infected hidradenitis suppurativa lesions and pyoderma gangrenosum on his abdomen. Dermatology and rheumatology deemed the constellation of symptoms appeared consistent with PsAPASH syndrome.

Conclusion: He was started on steroids for his inflammatory arthritis, doxycycline for his infected hidradenitis suppurativa, and a tumor necrosis factor-alpha inhibitor for disease-modifying treatment. He was discharged with close follow-up, noting improvement in his pain and quality of life.

#70. Follicular Mucinosis and Cutaneous T-Cell Lymphoma Association: A Clinicopathologic and Molecular Analysis of 19 Cases

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Mentor: Safina Hafeez

Program: Pathology

Type: Original Research

Background: Follicular mucinosis is a rare dermatologic disorder characterized by the accumulation of mucin within hair follicles, resulting in distinct skin lesions.

It can be associated with cutaneous T-cell lymphoma (CTCL), further complicating its management.

Methods: We identified 19 cases of follicular mucinosis from 2010 to 2023. We assessed hematoxylin and eosin (H&E) stained slides for mucin presence and lymphocytic infiltrate. Immunostains and T-cell receptor (TCR) gene rearrangement analysis was performed to

investigate clonality.

Results: Patients aged 12 to 87 years (median age: 38), with 11 males and 8 females, were included. Biopsies were obtained from various anatomical sites (**Table 1**). All cases exhibited mucin accumulation within hair follicles, confirming follicular mucinosis by H&E staining. Immunostains were evaluated to rule out lymphoma, with additional staining

Table 1. Summary of the clinical and molecular features of follicular mucinosis and cutaneous T-cell lymphoma association.

Cases	Age	Gender	Diagnosis	Anatomic Site	Epidermotropism	TCR Gene Rearrangement
1	17	M	follicular mucinosis	Forehead, thigh	Not present	Positive
2	35	F	follicular mucinosis with mycosis fungoides	Back	Not present	Positive
3	66	M	follicular mucinosis	Back	Not present	Positive
4	47	M	follicular mucinosis	Temple	Not present	Negative
5	38	M	follicular mucinosis	Back	Not present	The quantity of T-cell-derived DNA was insufficient for a T-cell gene rearrangement study
6	38	M	follicular mucinosis	Back	Not present	indeterminate for a clonal T-cell population
7	13	F	follicular mucinosis	Back	Not present	Negative
8	68	M	follicular mucinosis	Elbow	Not present	Negative
9	61	M	follicular mucinosis	Cheek	Not present	Suspicious but not diagnostic (small clonal population)
10	12	F	follicular mucinosis	Eyebrow	Not present	Not performed
11	13	M	follicular mucinosis	Cheek	Not present	Not performed
12	54	M	atypical lymphoid proliferation with follicular mucinosis	Scalp	Present	Positive
13	36	F	follicular mucinosis and atypical T-cell proliferation	Cheek	Not present	Positive
14	83	M	follicular mucinosis with mycosis fungoides	Back	Present	Positive
15	87	F	follicular mucinosis	Cheek	Not present	Positive
16	15	F	follicular mucinosis	Chin	Not present	Suspicious but not diagnostic (two sub-threshold peak)
17	49	F	follicular mucinosis	Labia Majora	Not present	negative
18	21	M	follicular mucinosis and atypical T-cell proliferation	Neck	Minimal epidermotropism	Positive
19	56	F	follicular mucinosis and atypical T-cell proliferation	Leg	Not present	Positive

(colloidal iron and Alcain blue) supporting mucin deposition. Two cases were associated with mycosis fungoides, and four cases were associated with atypical lymphoid proliferation. In the majority of cases (16/19), epidermotropism was not present. However, prominent epidermotropism occurred in 1/2 cases of mycosis fungoides and 1/4 cases of

atypical lymphoid proliferation. Additionally, minimal epidermotropism was noted in one case of atypical lymphoid proliferation. TCR gene rearrangement analysis in 17 cases showed clonality in 9 cases.

Conclusion: In cases of atypical lymphoid proliferation, TCR gene rearrangement was

positive, but there was insufficient evidence to diagnose lymphoma. Close follow-up was recommended. Some cases may exhibit clonal TCR gene rearrangements without T-cell lymphoma evidence, referred to as pseudo-clonality or benign clonality. Distinguishing between true clonality and pseudo-clonality can be challenging.

#71. Challenges and Improvements of Vitreous Fluid Processing Protocol in the Diagnosis of Intraocular Lymphomas: A 13-Year Institutional Review

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Mentor: Ana Yuil-Valdes

Program: Pathology

Type: Original Research

Background: Upon notification that a vitreous specimen will be sent to our Cytology department, the specimen is deemed a high priority specimen and ran as ‘stat’. Eight cytopspins are then made; one Diff-Quik stained, one Papanicolaou stained, and six unstained for potential immunocytochemical stains. The cytopathologist will triage the material and decide whether to order immunocytochemical stains, flow cytometry, and molecular studies. We also conducted a 13-year retrospective review to demonstrate the efficacy of our vitreous fluid protocol for the workup and diagnosis of intraocular lymphomas.

Methods: Upon notification that a vitreous specimen will be sent to our Cytology department, the specimen is deemed a high priority specimen and ran as ‘stat’. Eight cytopspins are then made; one Diff-Quik stained, one Papanicolaou stained, and six unstained for potential immunocytochemical stains. The cytopathologist will triage the material and decide whether to order immunocytochemical stains, flow cytometry, and molecular studies. We also conducted a 13-year retrospective review to demonstrate the efficacy of our vitreous fluid protocol for the workup and diagnosis of intraocular lymphomas.

Results: Thirteen cases of intraocular lymphoma were identified in nine patients. We found this workflow allowed for optimal cell preservation, aiding in the diagnosis of intraocular lymphomas.

Conclusion: Cytologic analysis of vitreous specimens is a valuable diagnostic tool in cases of intraocular lymphoma. Developing a multidisciplinary approach for processing vitreous fluid is vital to producing an adequate diagnostic sample. This protocol allowed for the accurate diagnosis of intraocular lymphoma.