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## Trichodysplasia Spinulosa related to Ruxolitinib: A Case Report

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## Losing My Appetite: A Case of West Nile Encephalitis

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**Mentor:** Lisa Sieczkowski

**Program:** Pediatrics

**Type:** Case Report

**Background:** Endemic to the United States, West Nile virus is an arbovirus that is primarily transmitted through bites of the Culex mosquito. While most infections are asymptomatic, 20% - 40% of infected individuals develop flu-like symptoms and a smaller portion develop neuroinvasive disease. Neurologic involvement increases risk of mortality and can cause long-term neurologic sequela.

**Case:** This case involves a 17 y/o male with depression who presented to the emergency

department for concerns of depression and anorexia. On admission he was febrile and unable to orient to self or location. Sedated brain MRI and LP were promptly performed for evaluation of meningitis. MRI imaging was consistent with encephalitis, but vascular infarct was also considered. Cerebral spinal fluid (CSF) evaluation showed leukocytosis and elevated protein, but negative meningitis/encephalitis panel. Initial treatment included ASA, vancomycin, ceftriaxone, and acyclovir. Due to persistent fevers and new information of frequent night fly-fishing over the summer, additional workup was performed. Results were significant for positive IgM West Nile antibody in the CSF. He transitioned to doxycycline to complete an empiric 7-day course. The patient's mental status slowly

improved to baseline and he was discharged home with close follow-up.

**Conclusion:** West Nile encephalitis should be considered in cases of encephalitis, especially when risk factors, such as regional location and outdoor hobbies, are present. In areas that are high risk, prevention education is encouraged. Additionally, this case illustrates the importance of adjusting the differential with new symptomatology and patient information as the case evolves.

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## Hidden in PIAIN Sight

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**Mentor:** Prasanth Ravipati

**Program:** Nephrology

**Type:** Case Report

**Background:** Acute interstitial nephritis (AIN), a common etiology of acute kidney injury (AKI), is frequently caused by medications. Historically, a triad of fever, rash, and sterile pyuria have been used as a clinical indicator of AIN. The aim of this case report is to illuminate an uncommon presentation of AIN to emphasize the importance of kidney biopsy for definitive diagnosis.

**Case:** A 56-year-old woman with a history of cystic fibrosis (CF) presented with fatigue and myalgias who was recently started

on levofloxacin for small bowel bacterial overgrowth syndrome. On admission, she was found to have non-oliguric AKI with serum creatinine (sCr) of 1.5 mg/dl (baseline sCr 0.6-0.7 mg/dl). Urinalysis showed no pyuria, hematuria, casts or proteinuria. Urine protein creatinine ratio was normal at 0.2 g/g. Renal ultrasound unremarkable. Kidney function continued to decline despite fluid resuscitation. Repeat urine studies were again without pyuria, hematuria, casts, or proteinuria, kidney biopsy showed dense inflammatory infiltrate within the interstitium including lymphocytes and eosinophils, acute tubular injury, consistent with AIN. Levofloxacin was discontinued and she was treated with prednisone, tapered over 3 months. Patient's kidney function returned to baseline.

**Conclusion:** Levofloxacin is an uncommon cause of AIN. Patient had normal urine sediment on two occasions, lowering suspicion for inflammatory kidney injury. The triad of fever, rash, and sterile pyuria occurs in less than 10% cases of AIN. Majority of AIN cases present with sub-nephrotic proteinuria (90%) and/or pyuria (60-80%). However, it is important to recognize that AIN can present with normal urine sediment.

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## Trichodysplasia Spinulosa related to Ruxolitinib: A Case Report

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**Mentor:** Melodi Javid Whitley

**Program:** Dermatology

**Type:** Case Report

**Background:** Trichodysplasia spinulosa (TS) is a rare disease that exclusively

affects immunocompromised patients, especially those with a history of solid organ transplantation, hematologic malignancy, and disease- or drug-induced immunosuppression. Here we report a case of TS presenting after the initiation of the oral Janus-associated kinase (JAK) inhibitor ruxolitinib.

**Case:** A 67-year-old female with a history of allogeneic bone marrow transplant (ABMT) from a related donor 18 months prior for polycythemia vera and myelofibrosis, currently on ruxolitinib, presented to the dermatology clinic with a facial rash. Given the patient's history of immunosuppression and characteristic physical exam findings, a

diagnosis of TS was made. Treatment was initiated with tretinoin 0.05% cream applied nightly and oral valganciclovir, dosed 450 mg twice weekly due to reduced renal function.

**Conclusion:** Ruxolitinib (Jakavi®/Jakafi®) is an oral JAK1/2 inhibitor. We report a case of TS with a notable association with initiation

of the JAK inhibitor, ruxolitinib. As novel immunomodulatory medications come into broad clinical use, recognizing the breadth of cutaneous side effects is of importance to practicing dermatologists.

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## Isomorphic Inframammary Erosive Lichen Planus After Radiation Treatment Progressing to Squamous Cell Carcinoma

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**Mentor:** Megan Arthur

**Program:** Dermatology

**Type:** Case Report

**Background:** Erosive lichen planus commonly presents on oral and vulvar mucosal surfaces and can progress to scarring, stenosis, and squamous cell carcinoma (SCC). Diagnosis is often delayed by patient embarrassment or misdiagnosis. Here we present an unusual case appearing in the inframammary fold.

**Case:** A 71-year-old female with stage 4 CKD and breast cancer s/p lumpectomy and radiation to the right breast presented with a non-healing erosion under the right breast. She has a history of long-standing oral and vulvar

erosions. Inframammary erosions appeared following radiation treatment. Previous workup at an outside institution included biopsies from the right breast and gingiva revealing lichenoid interface dermatitis and Direct Immunofluorescence (DIF) showing lichenoid tissue reaction and no autoimmune bullous disease. Previous failed treatments included fluconazole, micafungin, hyperbaric oxygen, timolol, intralesional steroids, oral metronidazole. She improved on prednisone. The facility diagnosis was erosive lichen planus. Later, she presented to our institution with worsening erosions under the right breast. Exam revealed a 14 cm sharply demarcated erythematous friable ulceration with violaceous-lacy changes present at the medial and lateral aspects. She was then tried on topical and intralesional

steroids, hydroxychloroquine, mycophenolate mofetil, and mycophenolic acid with minimal improvement. Repeat biopsy performed revealed invasive, moderately differentiated squamous cell carcinoma, and treated with Mohs Micrographic Surgery and mastectomy.

**Conclusion:** Erosive lichen planus on the mucosal surfaces of the mouth and vulva carries an increased risk of SCC development. However, this case represents an incredibly rare instance of recalcitrant erosive cutaneous lichen planus developing within an area not normally associated with the condition.

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## Myocardial Ischemia Without Evidence of Obstruction

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**Mentor:** Trek Langenhan

**Program:** Internal Medicine

**Type:** Case Report

**Background:** Angina is a common chief complaint encountered by internists, and up to 70% of patients with evidence of myocardial ischemia do not have obstructive coronary arteries. Ischemia with non-obstructed coronary arteries (INOCA) refers to cardiac ischemia caused by vascular dysfunction without obstructive coronary artery disease.

**Case:** A 72-year-old woman presented with a one-day history of progressively worsening chest pain at rest associated with diaphoresis, nausea, and left arm heaviness. The patient's history was significant for a prior stroke with no residual deficits and two prior heart catheterizations without intervention. Furthermore, the patient had recently moved across multiple states and reported increased

levels of stress and anxiety. Cardiac troponin was 0.15 (normal <0.04) and EKG revealed T-wave inversions in the inferior leads. A diagnosis of NSTEMI was made and patient was administered aspirin, atorvastatin, and therapeutic heparin anticoagulation. Troponins and EKGs were trended with a peak troponin of 0.55 and development of new T-wave inversions into the anterior leads. Transthoracic echocardiogram was normal. Left heart catheterization revealed no angiographic evidence of obstructive coronary arteries and a diagnosis of INOCA was made. Further medical management of INOCA included initiation of ranolazine, dose increase of isosorbide mononitrate, and continuation of amlodipine. Therapies targeting coronary artery disease including aspirin, carvedilol, losartan, and high intensity statin were also continued.

with chest pain. Confirming the diagnosis is essential to allow targeted management strategies and improve patient quality of life.

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**Conclusion:** INOCA is a common diagnosis in patients presenting to the general internist

## Pediatric Acute Liver Failure During the Pandemic: A Tale of Two Viruses

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**Mentor:** Anne Marie Kennedy

**Program:** Family Medicine

**Type:** Case Report

**Background:** This is an atypical presentation of pediatric acute liver failure (PALF) in a 12-month-old female in the winter of 2022. Data from this time period has confirmed Human Adenovirus Type 41 (HAT41) infection in the majority of PALF cases. Literature review demonstrates a substantial spike in PALF cases globally in winter through spring of 2022, with clusters in the US. Preliminary data has demonstrated a distinct correlation with PALF and Adenovirus HAT41 infection and

confirmed prior SARS COV-2 infection.

Theoretical contributing factors to worsened hepatic insult in pediatric populations are the known COVID-19 utilization of Angiotensin Converting Enzyme for cellular entry, with higher gastrointestinal/hepatic expression of ACE-2 when compared to adults. HAT41 association with severe hepatic symptoms in previously healthy children is well documented.

**Case:** A previously healthy 12-month-old female presents to the Emergency Department with acute listlessness and hematemesis. Initial workup demonstrated a positive rapid COVID, hypoglycemia, hypotension, AST 8,077, ALT 6,305, with normal INR and ammonia. Imaging was noncontributory and

no lab evidence of accidental ingestion was found. The patient was admitted to the PICU where a vitamin K protocol was started. INR peaked at 3.6 on day two of hospitalization then normalized with discharge, follow up labs showing normalized liver function by two weeks.

**Conclusion:** Most cases of pediatric viral illness present initially in the primary care setting. This case highlights the importance of keeping a broad differential diagnosis and recognizing the risk of severe sequelae with what would otherwise be a benign viral illness course.

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## Giant Cell Tumor of Soft Tissue in the Dermis

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**Mentor:** Dominick DiMaio

**Program:** Pathology

**Type:** Case Report

**Background:** Giant cell tumor of soft parts (GCT-ST) is a rare benign entity that can occur anywhere but most commonly occurs on the extremities, with very few case reports in other anatomical locations. Although histologically similar to giant cell tumor of bone, GCT-ST does not harbor the same H3F3A mutation as in giant cell tumor of bone (GCT-B). Currently, there is an immunohistochemical stain (H3.3g34w) that would be positive in GCT-B. To date, there has been no distinct molecular alteration for GCT-ST.

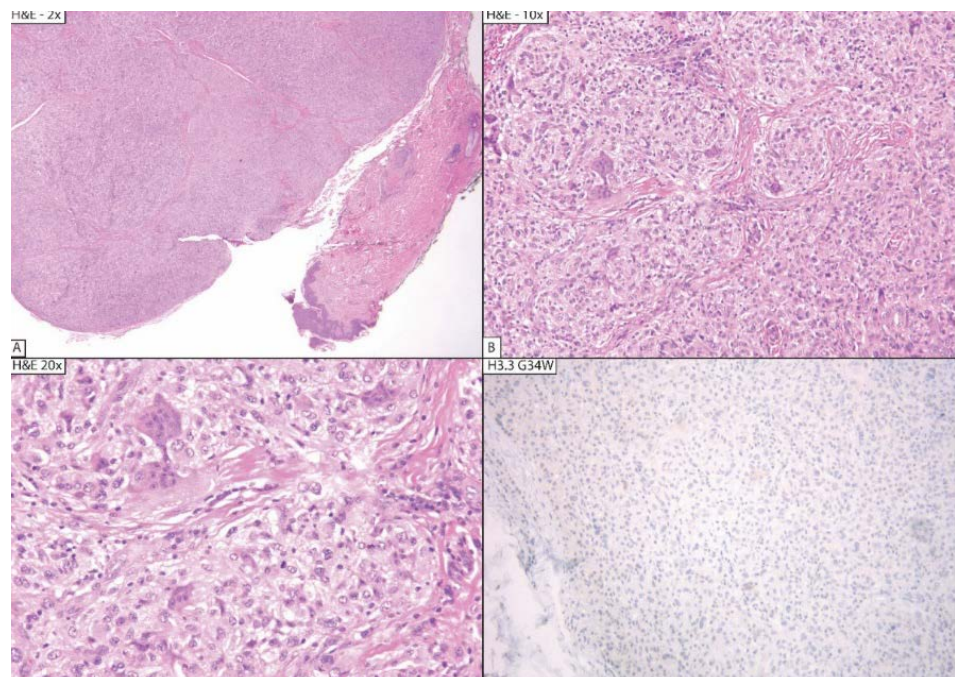
**Case:** We report on an 18-year-old woman with primary breast GCT-ST. She was referred to plastic surgery for a painless 'cyst' on the right upper breast that had been there for approximately one year. The mass was excised, and grossly had a rubbery, pale-tan multilobulated appearance that was not consistent with a sebaceous cyst. The mass is a well-defined lesion in the reticular dermis (Figure 1A), composed of sheets of histoid-appearing cells with scattered multinucleated giant cells (Figure 1B & 1C) resembling osteoclast-like giant cell tumor of bone (GCT). There are areas of cytological atypia and occasional mitotic figures. Immunohistochemical staining was diffusely

positive for CD68 and p63 with no significant staining for cytokeratins, melanocytic stains, muscle stains, CD34, CD1a, and H3.3 G34W (Figure 1D). There have been only a few case reports of GCT-ST in the breast.

**Conclusion:** We report on an extremely rare case of GCT-ST within the dermis. This

distinct entity should remain in the differential of giant cell-rich lesions as it has significant prognostic outcomes and treatment.

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**Figure 1:** Histologically the neoplasm demonstrates variable amounts of multinucleated giant cells similar to Giant Cell Tumor of Bone (GCT-B) (1A-C) but is negative for immunohistochemical staining for H3.3 G34W (1D)



## Cutaneous Involvement of IgG4 Disease

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**Mentor:** Dominick DiMaio

**Program:** Pathology

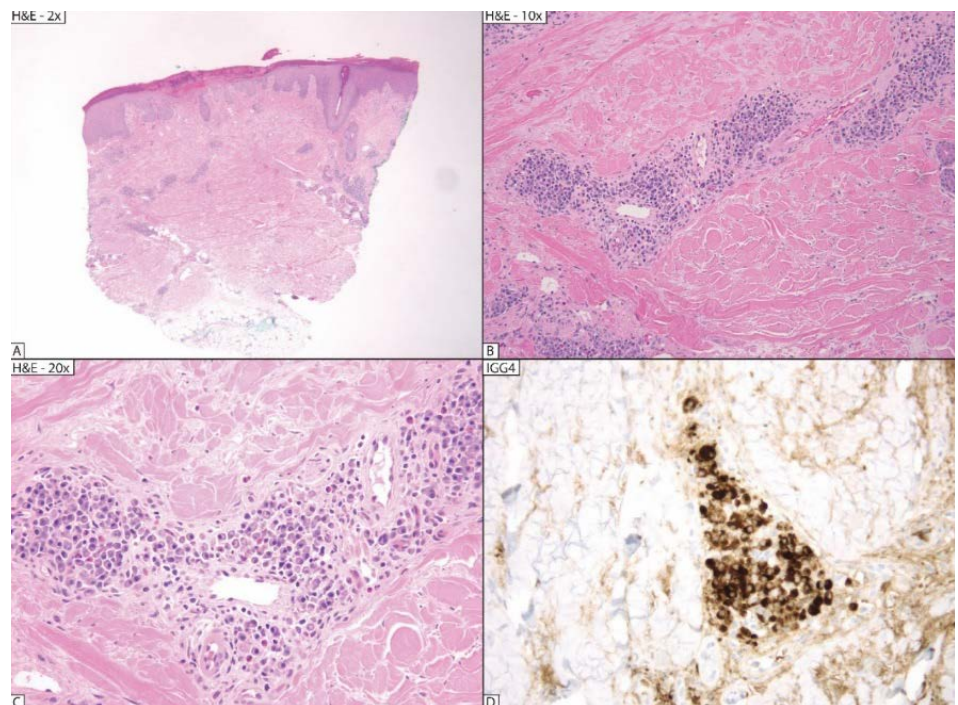
**Type:** Case Report

**Background:** IgG4 related disease is an uncommon disorder with a prevalence of 1 in 64 per million. It is an immune-mediated, fibroinflammatory condition that affects multiple organs and clinically, presents as tumor-like enlargement of organs. Histologically, it is characterized by lymphoplasmacytic infiltrate with IgG4 positive plasma cells, storiform fibrosis, and obliterative phlebitis. The current criteria include: 1) swelling or masses, 2) serum IgG4 concentration >135 mg/dL, and 3) >40% of IgG+ plasma cells being IgG4+ and >10 cells/high power field of biopsy sample.

**Case:** A 61-year-old woman presented with a 4-year history of pruritic dermatitis on her face, extremities, and back that. It appeared as pink crusted grouped papules coalescing into confluent scaly plaques with no lymphadenopathy. After undergoing numerous antibiotics and topical corticosteroid treatments, she continued to worsen. Biopsies demonstrated an epidermis excoriation (Figure 1A). Within the superficial dermis, there was a mild to moderate perivascular mixed inflammatory infiltrate composed of lymphocytes, histocytes and prominent number of eosinophils and plasma cells (Figures 1B & 1C). Plasma cells were polyclonal by kappa and lambda in-situ hybridization. Immunohistochemistry for IgG4 demonstrated increased IgG4 plasma cells (at least 50%, Figure 1D). Laboratory markers for IgE (>15 000 IU/mL) and IgG4 (515 mg/dL) were elevated.

**Conclusion:** While no one feature is diagnostic for IgG4 related disease, it is important to interpret the histologic findings in conjunction with the serologic findings to make the appropriate diagnosis. In this case, the histological, immunohistochemical, and laboratory findings support the rare diagnosis of IgG4 related disease involving the skin.

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**Figure 1:** Sections demonstrate a punch biopsy with excoriation, acanthosis, and spongiosis (1A). The superficial dermis demonstrates a perivascular inflammatory infiltrate with abundant plasma cells (1B & 1C). Immunohistochemical staining for IgG4 highlights a majority of plasma cells (at least 50%) (1D).

## Arresting Chest Pains: A Heart-Stopping Journey of VF and Vasospastic Angina

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**Mentor:** Derek Kruse

**Program:** Internal Medicine

**Type:** Case Report

**Background:** Gain knowledge and insight into the clinical manifestation, diagnostic approach, and therapeutic strategies for RCA vasospasm-induced STEMI, including the complexities and key factors in the management of these patients.

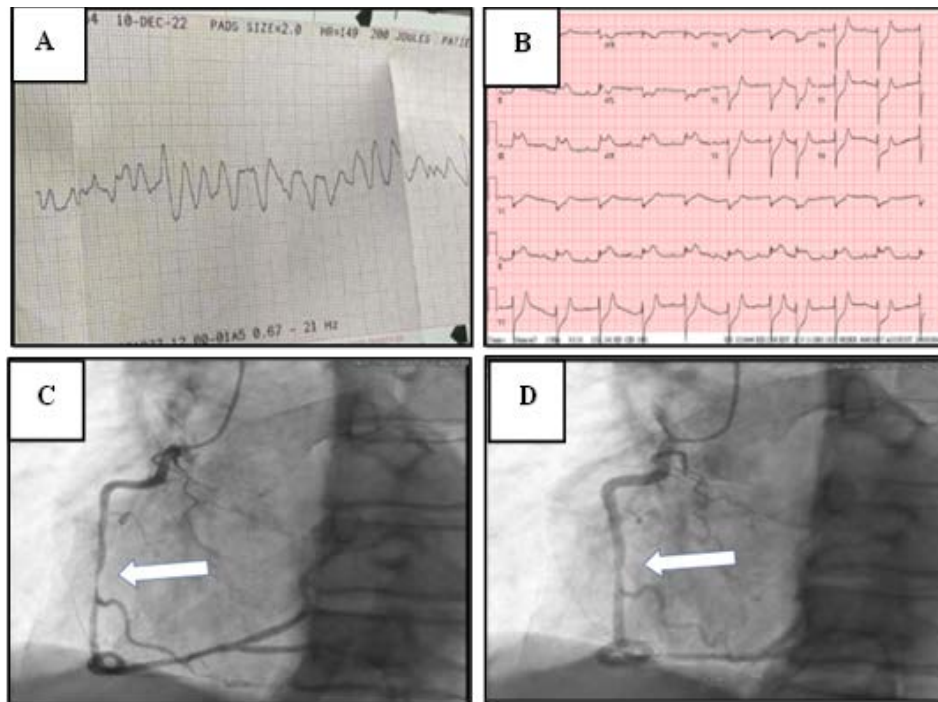
**Case:** A 59-year-old male was brought to the hospital with chest pain and diaphoresis. He went into ventricular fibrillation during transport (Figure 1A) and achieved return of spontaneous circulation following advanced cardiac life support (ACLS) in the Emergency Department. Upon evaluation by cardiology, the patient was not a qualified candidate for heart catheterization due to a poor neurological prognosis, and received TPA for STEMI treatment in the ED. The

following day the patient experienced another episode of symptoms. An EKG (Figure 1B) showed evidence of an inferior myocardial infarction. The patient deteriorated further, and he developed intermittent bradycardia and cardiogenic shock. He was taken to the Cath lab emergently, where it was discovered that the patient's RCA had severe coronary vasospasm (Figure 1C), which was treated with intracoronary nitroglycerin (Figure 1D). He was transitioned to a long-acting oral

nitroglycerin and underwent ICD placement for secondary prevention prior to discharge.

**Conclusion:** Vasospastic angina is a condition in which there is a spasm in the smooth muscle layer of a coronary artery, causing a sudden, high-grade obstruction. A recent meta-analysis of 24 studies, which included over 6500 patients suspected of having coronary disease without epicardial obstructive lesions, revealed a 40% prevalence of underlying coronary vasospasm. Myocardial infarction and life-threatening arrhythmias may occur in approximately 25% of untreated patients with vasospastic angina.

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**Figure 1:** 1A, Presenting rhythm strip from EMS, shortly prior to ROSC in the emergency department. 1B, EKG 10 minutes post-symptom onset, STEMI alert activation in hospital. PR interval 318 m/s. 1C, The arrow indicates a RCA-vasospasm causing nearly a 95% occlusion. 1D, The arrow indicates resolution of vasospasm. Non-obstructive CAD < 50% mid-RCA.

## A Small Bruise Led to a Large Discovery: Pelvic Mass Presented with Grey Turner's Sign

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**Mentor:** Stephen Cagle

**Program:** Family Medicine

**Type:** Case Report

**Background:** We present a case report that describes a patient with a large polycystic ovarian mass presenting with Grey Turner's Sign. This case highlights the importance of Grey Turner's Sign and its role in recognizing intraabdominal pathology.

**Case:** A 48-year-old female presented to an urgent care with a spontaneous bruise along her left flank and was subsequently sent to the emergency room with concern for Grey Turner's Sign. Grey Turner's Sign is an uncommon subcutaneous manifestation of intra-abdominal pathology, classically, acute necrotizing pancreatitis. The patient was otherwise asymptomatic. The initial ER evaluation included: CBC, CMP, PT/INR, PTT, UA, and Lipase which were within reference ranges. CT imaging was obtained and revealed an 8x5x4cm septate complex cystic mass with suspected right adnexal origin and confirmed by Pelvic US. Gynecology was consulted and quick outpatient follow-up was arranged. The

patient ultimately underwent laparoscopic bilateral salpingectomies with right oophorectomy. Pathology revealed benign mucinous cystadenoma.

**Conclusion:** Grey Turner's Sign is an indicator of retroperitoneal bleeding and most often the result of acute abdominal pathology. This patient was atypical in that she did not have acute pathology. She was asymptomatic and would not have pursued care for any symptoms if she had not noticed the spontaneous bruise on her flank. This case lends itself to the discussion that Grey Turner's Sign, while classically associated with severe life-threatening abdominal pathology, can be associated with any abdominal pathology that creates retroperitoneal bleeding. Grey Turner's Sign, although uncommon, is an extra-abdominal manifestation that should prompt further abdominal or pelvic work-up.

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