

Mesenteric Panniculitis: An Unusual Initial Presentation Of Systemic Lupus Erythematosus (SLE)

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Citation

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Abstract

Introduction: Patients with SLE are subject to a myriad of symptoms and complaints, and the inflammatory process can affect virtually every organ. The most common pattern of disease is a mixture of constitutional complaints with skin, musculoskeletal, hematological and serologic involvement. Mesenteric panniculitis is a rare disease affecting adipose tissue of the mesentery that may result in the development of large masses in the abdomen. Diffuse chronic or intermittent abdominal pain is the most frequent symptom.

Literature Search: We performed a Medline search using panniculitis as subject heading and peritoneal and mesenteric panniculitis as additional keywords. The search yielded 142 articles. Combining this search with our second search on Systemic Lupus Erythematosus yielded only one result in a French journal of a 10 year girl with mesenteric panniculitis as initial presentation of acute lupus erythematosus.

Case: We describe a 44 year old female with one year of recurrent abdominal pain and an otherwise negative past medical history. She underwent a laparoscopy for definite diagnosis of her abdominal pain. At surgery, chylous ascites was noted and a subsequent open laparotomy showed necrotic areas in the mesentery. Biopsy results revealed fat necrosis and an abdominal CT scan showed classic features of panniculitis. This patient later presented with night sweats, fever, lower extremity rash, pain in the proximal interphalangeal joints of her left hand with swelling and decrease in range of motion and thrombocytopenia. She also complained of dryness in her eyes and ears. Subsequent workup revealed she was ANA positive with a titer of 1:1280 in a speckled pattern and positive SSA and SSB antibody. Skin biopsy showed leukocytoclastic vasculitis. In addition she had a history of two miscarriages. She was diagnosed as having systemic lupus erythematosus manifesting with secondary sjogren's and idiopathic thrombocytopenia, and was started on oral corticosteroid treatment with excellent response.

Conclusion: Systemic lupus erythematosus must be included among the etiologies of intraabdominal panniculitis.

INTRODUCTION

We describe a 44yr old white female with one year history of recurrent abdominal pain. Her surgeon performed a laparoscopic repair of an umbilical hernia and to rule out adhesions. During the procedure they found large amount of chylous fluid in the cul de sac. Similar fluid was found under the diaphragm between the liver and chest wall. An exploratory laparotomy was performed which showed thickened small bowel mesentery extending from ligament of Treitz area almost down to the distal ileum. The tissue in the mesentery was also thickened and inflamed. The microscopic diagnosis of the resected portion of the mesenteric fibroadipose tissue showed prominent fat necrosis with acute and chronic inflammation, cholesterol cleft formation and reactive fibrosis. A CAT scan of the

abdomen without and with contrast was performed from the level of diaphragmatic leaves to the iliac crest was reported to have small amount of right pleural effusion with infiltrate in the right lower lobe. Liver, kidneys and pancreas were normal. There was diffuse enhancement of fat within the mesentery with the intraperitoneal space. No discrete abscess was identified and there was diffuse enhancement of the omentum. She was diagnosed with mesenteric panniculitis and closely followed.

Figure 1



The patient presented 2 years later with a lower extremity rash on the anterior aspect of her legs. It was non pruritic and associated with intermittent swelling of her legs. She was also experiencing night sweats and low grade fever. She denied any chills or myalgia. She had episodes of epistaxis but no recurrent oral lesions. She did report dryness in her eyes and corneal irritation. She reported pain in the proximal interphalangeal joint number three in her left hand with some swelling and decrease in range of motion. She denied any joint symptoms elsewhere. Review of system was unremarkable except for palpitations. She reported no pleuritic chest pain. Her past medical history was notable for hypothyroidism, 3 pregnancies and 2 miscarriages. Her past surgeries included tubal ligation, tonsillectomy and hernia repair. She is intolerant to penicillin, codeine and vibramycin. Her only medication was Synthroid 200mg daily. She is divorced, works as an accountant and has no history of smoking, alcohol or tobacco dependence. Family history is significant for arthritis and myocardial infarction in her father. Her mother had diabetes and “cirrhosis”. A paternal aunt had rheumatoid arthritis. She has two brothers who are doing well.

She was not in any distress on examination. Head and neck examination revealed no facial or scalp lesions and her mucosal membranes were intact. Schirmer test was positive with less than 5mm in 5 minutes in each eye. There was no conjunctival erythema. Her neck was supple and there was no parotid gland enlargement or cervical nodes. Lungs were clear to auscultation and there were bilateral breath sounds. Cardiovascular examination was normal with normal heart sounds without murmurs, rubs or gallops and regular rate

and rhythm. The abdomen was soft, non-tender and she had a midline scar below the umbilicus. There were no masses, bruits or palpable lymph nodes. Her musculoskeletal system was notable for left proximal interphalangeal joint number 3 having decreased range of motion. There was no significant erythema, warmth or swelling on examination however she does report this a few weeks earlier. She regularly experiences an eruption of several purplish skin lesions in her feet, which last for 2 –3 days. They generally do not leave any scars and are not associated with any pain or pruritis. She reports no arthralgia.

Workup revealed thrombocytopenia with platelet count of 79,000. Her platelets dropped to as low as 30,000 and rose to 179,000 on treatment with prednisone. Subsequent workup also revealed that she was ANA positive with titers of 1280 in a speckled pattern. She was anti SSA and anti SSB antibody positive, SCL-70 antibody negative, Jo-2 negative and anti DNA negative. Her C3 complement was 96 and C4 complement was 9. Her antiphospholipid antibodies are negative. Her cryoglobulins is negative and serologies for Hepatitis C are also negative. Skin biopsy showed leukocytoclastic vasculitis and bone marrow biopsy was non specific. CAT scan of chest, abdomen and pelvis showed bullous lesions in her lungs and calcifications in her spleen. It suggested mesenteric panniculitis which was unchanged from her previous study.

Impression is a 46-year-old white woman with probable autoimmune thrombocytopenia, idiopathic thrombocytopenic purpura, leukocytoclastic vasculitis, sicca syndrome, history of pleurisy and mesenteric panniculitis. She is noted to be ANA positive, anti SSA and anti SSB antibody positive. The patient has a history of recurrent miscarriages. The constellation of signs and symptoms are compatible with an underlying autoimmune process such as systemic lupus erythematosus. She was diagnosed as having systemic lupus erythematosus manifesting with secondary Sjogren's and idiopathic thrombocytopenia, and was started on oral corticosteroid treatment with excellent response.

DISCUSSION

Mesenteric panniculitis is a non-neoplastic inflammatory process that constitutes the second stage in a rare, progressive disease involving the adipose tissue of the mesentery. This pathologic condition was first described by Jura in 1924 as sclerosing mesenteritis. In the 1960s, Ogden et al used the term mesenteric panniculitis to describe this syndrome. Other terms that have been used include retractile

mesenteritis, lipogranuloma of the mesentery, isolated lipodystrophy, and retroperitoneal xanthogranuloma. The disease has a 2–3:1 male predilection and is seen more frequently in patients over 50 years old. Children are rarely affected, possibly because they have less mesenteric fat. The cause of this rare disease remains unclear. Durst et al found certain predisposing factors such as recent surgery (mainly cholecystectomy and appendectomy) in 17% of cases and intercurrent disease (cholelithiasis, cirrhosis, abdominal aortic aneurysm, peptic ulceration, and gastric carcinoma) in 25%. Prior abdominal trauma, autoimmune disease, drug use, and retained suture material have also been implicated. We performed a Medline search using panniculitis as subject heading and peritoneal and mesenteric panniculitis as additional keywords. The search yielded 142 articles. Combining this search with our second search on Systemic Lupus Erythematosus yielded only one result in a French journal of a 10 year girl with mesenteric panniculitis as initial presentation of acute lupus erythematosus. We conclude from this case discussion is Systemic lupus erythematosus must be included among the etiologies of intraabdominal panniculitis.

Figure 2

Table 1: The 1982 Criteria for Classification of Systemic Lupus Erythematosus, Updated 1997

1. Malar rash	Fixed erythema, flat or raised, over the malar eminences
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur
3. Photosensitivity	Exposure to UV light causes rash
4. Oral ulcers	Includes oral and nasopharyngeal, observed by physician
5. Arthritis	Nonerosive arthritis involving two or more peripheral joints, characterized by tenderness, swelling, or effusion
6. Serositis	Pleuritis or pericarditis documented by ECG or rub or evidence of pericardial effusion
7. Renal disorder	Proteinuria > 0.5 g/d or > 3+, or cellular casts
8. Neurologic disorder	Seizures without other cause or psychosis without other cause
9. Hematologic disorder	Hemolytic anemia or leukopenia (< 4000/ ⁴ L) or lymphopenia (< 1500/ ⁴ L) or thrombocytopenia (< 100,000/ ⁴ L) in the absence of offending drugs
10. Immunologic disorder	Anti-dsDNA, anti-Sm, and/or anti-phospholipid
11. Antinuclear antibodies	An abnormal titer of ANAs by immunofluorescence or an equivalent assay at any point in time in the absence of drugs known to induce ANAs
If four of these criteria are present at any time during the course of disease, a diagnosis of systemic lupus can be made with 98% specificity and 97% sensitivity.	
Criteria published by EM Tan et al, Arthritis Rheum 25:1271, 1982; updated by MC Hochberg, Arthritis Rheum 40:1725, 1997.	

References

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