



## **Masquerade Party: Case Studies in Systemic Disease Causes of Abdominal Pain**

Many disease processes produce symptoms remote from their primary target, causing abdominal pain. The differential of such diseases is broad and includes seemingly disparate causes such as metabolic, hematologic, and infectious disease. This course will help you develop an approach to abdominal pain when the cause is not abdominal.

- Develop a differential diagnosis of causes of abdominal pain due to non-abdominal disease processes.
- Develop an efficient plan for the workup of abdominal pain of non-abdominal origin.

WE-181  
Wednesday, October 13, 1999  
5:00 PM - 5:55 PM  
Room # N223  
Las Vegas Convention Center

### **FACULTY**

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# **1999 SCIENTIFIC ASSEMBLY**

## **Masquerade Party: Case Studies in Systemic Disease Causes of Abdominal Pain**

**Course # WE-181**

**Instructor:**

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**University of North Carolina, Chapel Hill**

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## Case studies in systemic disease causes of abdominal pain

### COURSE DESCRIPTION

This course will present an approach to abdominal pain from extraperitoneal, systemic causes. A classification of systemic diseases causing abdominal pain will be presented and illustrated in a case format.

### OBJECTIVES

- **Expand the differential of abdominal pain to include systemic disease processes**
- **Identify characteristics of abdominal pain suggesting a systemic cause.**
- **Develop a classification and approach to systemic causes of abdominal pain.**

### COURSE OUTLINE

#### **Differential diagnosis**

Systemic causes  
Types of abdominal pain  
Making the diagnosis

#### **Clues to suggest a systemic cause**

#### **Cases 1-6:**

Hematologic  
Metabolic  
Infectious  
Inflammatory  
Toxicologic  
Functional

## **The Challenge of Abdominal Pain**

- a common complaint in the emergency department
- frequently remains undiagnosed at discharge
- huge spectrum of disease and acuity
- a common source of misdiagnosis with risk management issues
- even common causes don't follow the rules

### **Types of Abdominal Pain**

- location, course, description, and associated symptoms may suggest certain etiologies but the overlap is extensive
- visceral pain can be caused by tension in the wall or organ capsule, ischemia, or inflammation of the serosa (peritoneum)
- in general serosal inflammation is more localizing however may be secondary to a remote cause – i.e. the perforation is painless, the subsequent peritonitis is painful but delayed, remote, or diffuse
- even in common surgical abdomen diagnoses, pattern recognition results in a 30-40% rate of misdiagnoses
- pain may also be referred to and from remote sites due to the embryonic origins of the structure or referred from contiguous, extra-abdominal structures

### **Making the Diagnosis**

- 75% of abdominal pain presenting to the emergency is non-surgical
- 40 to 50% remain undiagnosed at the point of disposition from the ED (nonspecific or undifferentiated abdominal pain)
- 80% of these undifferentiated patients remain undiagnosed or have a benign, self-limited course
- What about the other 20%?

### **Expansion and contraction of the differential diagnosis**

- Most physicians follow a step-wise approach to narrowing the broad differential diagnosis
- The “missed” 10% might be reduced if a few questions are entertained before labeling as “undifferentiated abdominal pain”

Abdominal pain

Rule out acute surgical pathology

Consider non-surgical intra-abdominal processes

- 1. Is this an atypical presentation of a common intra-abdominal disease?**
- 2. Is this a typical presentation of an uncommon (rare) intra-abdominal disease?**
- 3. Is this an extra-abdominal process causing referred pain?**
- 4. Is this a symptom of a systemic disease?**

“Undifferentiated abdominal pain”

**Clues to suggest a systemic cause of abdominal pain**

- 1. Diffuse pain**
- 2. Pain out of proportion to exam**
- 3. Patient is sick, exam is benign**
- 4. Constitutional symptoms**
- 5. Immunocompromised, Elderly**
- 6. Multiple return visits**
- 7. The patient tells you it's systemic**

**Table 1: Systemic causes of abdominal pain**

<p><b>1. Hematologic</b></p> <ul style="list-style-type: none"> <li>• Sickle cell disease</li> <li>• Leukemia</li> <li>• Lymphoma</li> <li>• Thrombocytosis</li> <li>• Cyclic neutropenia</li> <li>• Acute hemolytic states</li> <li>• Coagulopathies</li> </ul> <p><b>2. Metabolic</b></p> <ul style="list-style-type: none"> <li>• DKA</li> <li>• Uremia</li> <li>• Hypercalcemia</li> <li>• Pheochromocytoma</li> <li>• Addisonian crisis</li> <li>• Porphyria</li> <li>• Angioedema</li> <li>• Thyrotoxicosis</li> <li>• Familial Mediterranean fever</li> <li>• Hemochromatosis</li> </ul> <p><b>3. Infectious</b></p> <ul style="list-style-type: none"> <li>• Meningococcemia</li> <li>• Rocky mountain spotted fever</li> <li>• Lyme disease</li> <li>• Toxic Shock Syndrome</li> <li>• Tuberculosis</li> <li>• Varicella</li> <li>• Mononucleosis</li> </ul>	<p><b>4. Inflammatory</b></p> <ul style="list-style-type: none"> <li>• Systemic lupus erythematosus</li> <li>• Polyarteritis nodosa</li> <li>• Acute rheumatic fever</li> <li>• Henoch-Schonlein purpura</li> <li>• Rheumatoid vasculitis</li> <li>• Eosinophilic enteritis</li> </ul> <p><b>5. Toxins</b></p> <ul style="list-style-type: none"> <li>• Heavy metals</li> <li>• Iron, mercury</li> <li>• Mushroom poisoning</li> <li>• Black widow spider</li> <li>• Sympathomimetics</li> <li>• Salicylates</li> </ul> <p><b>6. Functional</b></p> <ul style="list-style-type: none"> <li>• Munchausen's</li> <li>• Malingering</li> <li>• Somatization disorder</li> </ul>
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## **Hematologic**

### **Common mechanisms for pain:**

- Localized thrombosis
- Vaso-occlusive ischemia
- Organomegaly due to sequestration or infiltrate
- Mucosal hemorrhage

### ***Sickle cell disease***

- Recurrent episodes, often precipitated by acute stressors
- Severe, diffuse, continuous pain
- Pain secondary to hypoxemia from abnormal erythrocytes obstructing arterioles
- May be associated with elevated LFTs and bilirubin
- High incidence of cholecystitis
- Consider acute splenic sequestration in very young
- Can cause ischemic bowel injury
- Patient is often helpful in suggesting whether this is a “typical” crisis

### ***Leukemia***

- May represent ischemic pain due to hyperviscosity
- *Pseudoacute abdomen* refers to a functional obstruction of the bowel of unclear etiology – autonomic dysfunction vs. local vascular compromise
- Can cause vaso-occlusive disease of the liver in graft vs host disease
- Typhlitis is a necrotizing colitis of the cecum occurring at the nadir of WBC depression
- Distinction between those requiring surgery and not can be difficult but is critical – misdiagnosis associated with a high mortality
- Admit these patients if pain is acute and cause is unknown

### ***Thrombocytosis***

- Thrombosis of the microvasculature secondary to elevated platelets
- Usually occurs at counts in excess of 1,000, but can occur if greater than  $500 \times 10^9/L$
- Can also cause hemorrhage

### ***Cyclic neutropenia***

- Rare blood disorder of periodic episodes of severe neutropenia
- Associated with spontaneous necrosis of the cecum

**Take home points:**

1. Many systemic causes are recurrent
2. An intra-abdominal process may be what triggered the systemic problem.
3. CBC is helpful in identifying hematologic causes

**Metabolic**

***Addisonian crisis***

- Infectious causes are becoming more common secondary to AIDS and immunosuppression
- Classically have hyponatremia, hyperkalemia, hypoglycemia, hypercalcemia
- Acute crisis may not reflect changes in K<sup>+</sup> and Na<sup>+</sup>
- Ask about steroids
- Remember diagnosis in undifferentiated shock states - consider steroid replacement

***DKA***

- Diffuse abdominal pain and tenderness
- May have decreased bowel sounds and guarding
- Can be the first presentation of juvenile onset DM
- Usually resolves with rehydration
- Can be precipitated by surgical pathologies, but DKA should be corrected before OR – high intra-operative mortality

***Hypercalcemia***

- Vague, diffuse, abdominal pain
- Fatigue, anorexia, constipation
- Can be associated with pancreatitis, renal calculi, peptic ulcer disease

***Porphyria***

- Rare hereditary disorder of heme synthesis
- Due to over-production porphobilinogen
- Precipitated by drugs or stressors which interfere with heme synthesis
- Severe abdominal pain, persistent or colicky, with very benign exam



- Similar episodes in past
- Associated with CNS changes
- Dx by elevated urinary porphobilinogen

***Thyrotoxicosis***

- Once thought to be a common cause of abdominal pain
- Mechanism unclear
- Associated with other symptoms suggestive of hyperthyroidism

***Angioedema***

- C1-inhibitor deficiency leads to excess active C1
- Increased submucosal permeability and edema
- Causes pseudo-obstruction
- May see “coin-stacking” on plain film

***Familial Mediterranean fever***

- Recurrent attacks of fever and severe diffuse abdominal pain
- Caused by serosal inflammation of unknown etiology
- associated with arthritis, familial

***Pheochromocytoma***

***Uremia***

***Hemochromatosis***

**Metabolic take home:**

- Look at your chemistry panel - are the changes really “just from the vomiting”
- Consider adding TSH
- Metabolic crisis usually precipitated by a stressor - look for concomitant disease

## **Infectious**

### ***Rocky mountain spotted (or spotless) fever***

- Characteristic rash, macular, begins on wrists and ankles, then palms and soles
- Spreads, becomes palpable and petechial
- Onset usually after 3-5 days
- 10-15% no rash at all
- Diagnostic serology is retrospective
- Mortality 5% in treated, 25% in untreated
- Death rare if treated within 6 days of symptom onset
- Don't wait for rash or serology, treat on suspicion
- Recall tick or in endemic area 2/3 of time
- Doxycycline 100mg BID X 7 days

### ***Tuberculosis***

- Peritoneal involvement rare but more frequent in AIDS patients and immunosuppressed
- Associated constitutional symptoms
- Pain often subacute, diffuse

### ***Varicella***

- Zoster can present with the prodrome of painful cutaneous hyperesthesia, usually in the distribution of thoracic nerve roots
- Vesicular rash appears several days later
- Primary varicella in adults and immune-compromised can cause hepatitis, pancreatitis, and enteric ulcerations

### ***Meningococemia***

### ***Lyme disease***

### ***Toxic Shock Syndrome***

### ***Mononucleosis***

### **Infectious Rashes:**

- Meningococemia - petechial
- Rocky mountain spotted fever - above
- Lyme disease - erythema migrans
- Toxic Shock Syndrome - desquamation
- Varicella - vesicular, dermatomal

**Take home points:**

- Look for characteristic rashes
- Abdominal pain follows significant constitutional symptoms
- Fever, vomiting, abdominal pain, but no diarrhea - beware the “gastro label”

**Inflammatory**

***Henoch-Schonlein purpura (HSP)***

- Self-limited generalized vasculitis
- Involves dermis, glomeruli, bowel wall
- Children 8-12
- Often follows viral RTI, in Spring
- 8% get GI bleed
- Abdominal pain responds to steroids

***Polyarteritis nodosa***

- Inflammation of medium and small arteries
- Usually 40-60 years of age
- Subacute, associated systemic signs such as arthritis
- GI bleeding common but not acutely

***Rheumatoid vasculitis***

- Necrotizing arteritis in subset of patients with RA
- Associated with skin infarction and ulceration

***Systemic lupus erythematosus***

- Abdominal complaints present in up to 50% of SLE
- GI vasculitis complicates 10%, can lead to perforation

***Eosinophilic enteritis***

- Rare cause of chronic, recurrent RLQ pain
- Uncertain etiology, may be linked to parasitic infections
- Granulomas present throughout the GI tract, associated with blood eosinophilia

### **Inflammatory Rashes:**

- Systemic lupus erythematosus: malar “butterfly” rash, sun-exposed areas
- Polyarteritis nodosa: subcutaneous red tender nodules
- Acute rheumatic fever: painless subcutaneous nodules, erythema marginatum
- HSP: palpable purpura
- Rheumatoid vasculitis: skin infarction and ulceration

### **Take home points:**

- Look for characteristic rashes
- Recurrent, multiple visits
- Other systems with signs of inflammatory disease, such as arthritis

## **Toxins**

### ***Heavy metals: lead***

- “Lead colic” described by Hippocrates in 370 B.C.
- Most common in children < 6 y.o.
- Binds to RBCs and stored in bone and teeth
- Neuropsychiatric problems, neuropathy
- Serum lead level diagnostic
- Basophilic stippling on smear
- Chelation therapy

### ***Arsenic***

- Pesticides, industrial exposures
- Metallic taste, garlic-odor of the breath
- Tissue hypoxia causes abdominal pain, N&V, diarrhea and bowel wall sloughing
- Associated CNS depression

### ***Black widow spider***

- Stimulates the release of neurotransmitters, primarily acetylcholine, causing painful muscle spasms
- Prominent over the abdomen, may have board-like rigidity
- Antivenin available

*Iron, mercury*  
*Mushroom poisoning*  
*Sympathomimetics*  
*Salicylates*

**Take home points:**

- Most of these toxins are not identified on a toxicology screen
- Need to ask about possible toxins and in particular, occupational exposure
- Associated with encephalopathy

**Functional**

***Munchausen's***

- Patient consciously fabricates symptoms or self mutilates to gain admission to hospital or medical care

***Malingering***

- Patient consciously fabricates symptoms for more obvious secondary gain – drugs, time off work, etc.

***Somatization disorder***

- Patient unconsciously experiences symptoms without organic etiology
- Functional abdominal pain common in children
- Multiple, recurrent, vague abdominal pain
- Rarely free of symptoms
- Extensive negative work-ups, tends to be chronic with poor response to psychotherapy

**Take home points:**

- Diagnoses of exclusion
- Think about systemic causes of abdominal pain first

**Suggested readings**

1. Pearigan PD. Unusual causes of abdominal pain. Emerg Med Clin North Am. 14:593-613, 1996.
2. Lukens TW, Emermen C, Effron D. The natural history and clinical findings in undifferentiated abdominal pain. Ann Emerg Med. 22:690-

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  5. Purcell TB. Nonsurgical and extraperitoneal causes of abdominal pain. Emerg Med Clin North Am. 7:721-40, 1989.
  6. Drossman DA. Patients with psychogenic abdominal pain: Six years observation in the medical setting. Am J Psychiatry. 139:1549-57, 1982.