

Panniculitis

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Panniculitis
Classification

- Based on pattern of inflammation in the subcutaneous fat on examination of the biopsy specimen.
- Septal: Inflammation primarily within septae that contain collagen and vessels between fat lobules
- Lobular: Inflammation primarily involving the fat lobules

Septal Panniculitis

- Erythema Nodosum
- Morphea / Scleroderma
- α_1 -Antitrypsin deficiency panniculitis



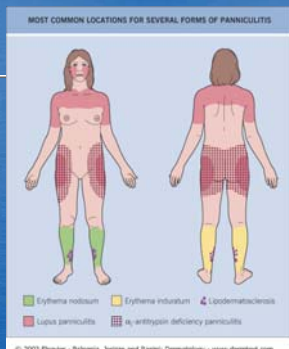
Lobular and Mixed Lobular / Septal

- Erythema induratum
- Pancreatic panniculitis
- Subcutaneous fat necrosis of the newborn
- Sclerema neonatorum
- Lupus panniculitis
- Traumatic panniculitis
- Lipodermatosclerosis
- Panniculitic like T-cell lymphoma



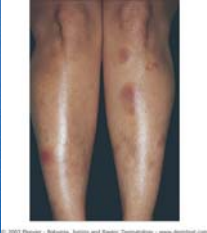
Diagnosis

- Clinical features:
 - History, review of systems, PMHx, meds
 - Distribution
 - Clinical appearance fairly non-specific
 - Histopathologic pattern:
 - Biopsy: Excision better than punch
 - Differences may be subtle
- ➔ Identify type of panniculitis
- ➔ Identify underlying etiology





Patient Presentation



- 30 year-old woman with acute onset of tender, erythematous, subcutaneous nodules
- Preceded 1 week earlier by sore throat, fever, and malaise
- History of similar lesions one year ago that resolved spontaneously after one month
- Histology: septal panniculitis

Erythema Nodosum

- Etiologies (common):
 - Idiopathic (50%)
 - Strep URI
 - Other URI (TB, mycoplasma)
 - Drugs: estrogens, OCPs, sulfas, PCN
 - Sarcoidosis
 - IBD (Crohn's > ulcerative colitis)
 - Coccidiomycosis



Erythema Nodosum

- Etiologies (uncommon)
 - Infectious: Yersinia, hep B, meningococcus, gonococcus, E. coli, syphilis, leprosy, HIV, histoplasmosis
 - Behcet's
 - Sweet's syndrome
 - Pregnancy



Finding a Systemic Cause

- >1/3 of patients have no associated systemic disease.
- Suggestive features: arthritis, diarrhea, abnormal CXR, preceding URI, elevated ASO titer, positive TB test.



- Throat culture / rapid strep test
- ASO titer
- If history unclear: CXR, cocci titer, PPD, HIV, hepatitis panel, urine pregnancy test



Treatment

- Discontinue possible causative meds
- Treat underlying condition
- Bed rest and leg elevation
- NSAIDS
- Potassium iodide
- Others: colchicine, prednisone, dapsone, hydroxychloroquine

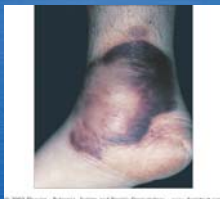


Morphea / Scleroderma

- Indurated plaques on the extremities, trunk; widespread or localized
- Primary fat involvement: deep morphea (morphea profunda)
- Secondary involvement: extension from inflammation of the dermis



Alpha₁-antitrypsin Deficiency



- Genetic protein deficiency
- Onset: infant to 8th decade
- Erythematous and purpuric, painful nodules or plaques that often ulcerate with oily discharge
- Often at sites of trauma (1/3 of cases)



Alpha₁-antitrypsin Deficiency

- Protease inhibitor deficiency: increased uninhibited degradation of tissue with secondary inflammation.
 - Patients homozygous for affected gene are more severely affected
- Associated conditions: cirrhosis, emphysema, pancreatitis, renal disease, rheumatoid arthritis, C-ANCA vasculitis, angioedema
- Treatment: alpha₁-antitrypsin replacement



Erythema Induratum



- Erythematous nodules or plaques that may ulcerate and drain, heal with scarring
- Recurrence common
- Posterior lower legs
- Women (30-40) > men
- Cause (TB vs. non-TB)



Erythema Induratum

- Term "nodular vasculitis" sometimes used to refer to non-TB cases.
- Other causes: idiopathic, infection (e.g. Nocardia), drugs (e.g. propylthiouracil)
- Differential: polyarteritis nodosa, thrombophlebitis, other panniculitis
- Workup: Biopsy, PPD, CXR, PCR of lesional specimen for TB DNA.



Pancreatic Panniculitis



- Erythematous, edematous, painful nodules; usually legs; may migrate
- Sometimes ulcerate with oily discharge
- Symptoms: fever, abdominal pain, polyarthritits

- May precede detection of pancreatic disease by 1-7 months.
- Acute or chronic pancreatitis, pancreatic carcinoma, pancreatic pseudocysts, pancreas divisum, traumatic pancreatitis
- Appearance may signal metastasis of pancreatic carcinoma



Subcutaneous Fat Necrosis of the Newborn



- Newborn full-term with one or more smooth, circumscribed, mobile, violaceous subcutaneous nodules or plaques
- Often symmetric

Arises in first 2-3 weeks; most resolve spontaneously
Possible causes (hypothermia, hypoxemia, birth trauma)
May see hypercalcemia which can be fatal if untreated (Calcium levels must be monitored for 4 months)

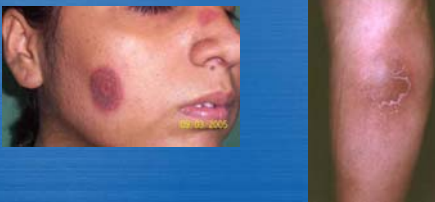


Sclerema Neonatorum

- Skin is diffusely cold, rigid, and board-like due to crystallization of fat.
- Severely ill premature infants, 1st week.
- Factors: hypothermia, asphyxia, dehydration; increased saturated:unsaturated fatty acid ratio
- Treatment is supportive only



Lupus Panniculitis



- Tender subcutaneous nodules and plaques on the face, proximal extremities, and trunk



Lupus Panniculitis

- 2-3% of lupus cases; Age: 30-40, women > men
- May underly typical discoid LE lesion (1/3 of patients have discoid LE)
- May see depression in overlying skin +/- ulceration
- Chronic, relapsing; heals with subcutaneous atrophy
- May precede cutaneous LE by many years
- Less commonly associated with systemic LE; consider dermatomyositis
- Treatment: antimalarials (hydroxychloroquine), systemic steroids (for acute phase), dapsone, cyclophosphamide, thalidomide



Traumatic Panniculitis



- Cold (e.g. popsicles, equestrians)
- Injectables (mineral oil, silicone, vitamin K, tetanus shot)
- Factitial (milk, feces)
- Blunt trauma
- Radiation therapy

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Lipodermatosclerosis



- Erythema, induration, and hyperpigmentation on one or both legs
- Setting: chronic venous insufficiency
- Acute: pain, warmth, erythema, induration
- Chronic: "inverted wine bottle"
- Tx: elevation, compression, anabolic steroid (stanozolol) - enhances fibrin breakdown



Infective Panniculitis

- Patient often immunosuppressed or has diabetes
- Causes: bacteria, mycobacteria, fungi
- May result from direct inoculation or secondary to systemic infection
- Clinical: local swelling, erythema and sometimes fluctuant nodules



Subcutaneous Panniculitis-like T-cell Lymphoma



- Subcutaneous purpuric nodules
- Signs/symptoms: fever, hepatosplenomegaly, pancytopenia, liver failure, mucosal ulcers, DIC
- Macrophages in the lesions engulf RBCs, WBCs (cytophagocytosis)
- Some cases show latent EBV
- Workup: Immunophenotyping and genotyping studies

Summary

- Panniculitis of diverse etiologies are often clinically similar.
- A thorough workup is essential to rule out potentially fatal underlying disease.
- Management is two-fold:
 - Symptomatic treatment
 - Treatment of underlying disease
- Clinical follow-up is essential, especially when the initial presentation is not classic.

TABLE 109-7

Diagnostic Tests for the Etiology of Panniculitis

CUTANEOUS BIOPSY	
Hematoxylin and eosin	See Tables 109-8 and 109-9
Special tests for microorganisms	Anti-BCG immunostaining screens for bacteria and fungi Special stains (Gomori methenamine stain, PAS stain, Gram's stain, Papanicolaou stain) Tissue culture for bacteria, fungi, and mycobacteria PCR tests for specific microorganisms (e.g., <i>Mycobacterium tuberculosis</i>)
Special tests for lymphoma/leukemia	Immunohistochemistry stains (CD43, CD3, CD4, CD8, CD20, CD36, lysozyme, myeloperoxidase, TIA, and granzyme B) Genetic studies for T and B cell gene rearrangement by PCR and Southern blot techniques
Special tests for foreign bodies	Polarizing microscopy Spectroscopic analysis
BLOOD TESTS	
Pancreatic enzymes (lipase and amylase) or α -1-Antitrypsin levels and phenotype	Antistreptolysin O antibody and other microbiological serologic tests (e.g., fungal serologies)
Antinuclear antibodies	Angiotensin-converting enzyme level
Extractable nuclear antigen antibodies	Fasting serum glucose/glycosylated hemoglobin
Anti-double-stranded DNA	Serum protein electrophoresis
Rheumatoid factor	Serum calcium, phosphorus, and PTH
Anti-neutrophilic cytoplasmic antibody	Serum uric acid
DIAGNOSTIC IMAGING	
Chest radiography	Lower extremity venous Doppler ultrasound studies
CT scan of thorax, abdomen, and pelvis	
OTHER	
Tuberculin skin test	Colonoscopy
Leprosin skin test	
