

Extra-Intestinal Manifestations of Inflammatory Bowel Disease

Kendra Johnson July 2013

Extra-intestinal manifestations (EIM) are reported in 6-47% of patients with IBD and may be diagnosed before, after or at the same time as the IBD. The presence of one EIM increases the likelihood of developing other manifestations.

Musculoskeletal

- **Peripheral arthritis** (5-10%UC; 10-20%CD). Diagnosis is clinical. Little to no joint destruction, RF & ANA neg.
 - ****Type 1 (pauciarticular)**** <5 large joints, related to IBD activity
 - treat IBD, avoid NSAIDs (exacerbate IBD)
 - can do joint injections of corticosteroids or lidocaine
 - **Type 2 (polyarticular)** ≥5 large joints, independent of IBD activity
 - Treatment is difficult, options include: sulfasalazine, methotrexate, infliximab
- **Axial arthropathies** Do not parallel IBD activity, may progress independent of IBD activity
 - **Sacroiliitis** in IBD is usually asymptomatic and non-progressive (found on CT/XRAY in 18-32%).
 - **Ankylosing spondylitis**** (5-10% of IBD patients). Symptoms same as non-IBD ankylosing spondylitis. Tx includes steroid injections, methotrexate, azathioprine, infliximab

Skin

- ****Erythema Nodosum**** (up to 10% UC; up to 15% CD). Raised, tender, red or violet inflammatory nodules 1-5 cm in diameter, typically on anterior extensor surface of lower extremities (pretibial) but rarely face and trunk.
 - Parallels IBD flares, resolves with treatment of these flares
 - Systemic steroids are usually needed
- **Pyoderma Gangrenosum** (0.4-2%). Occur in site of trauma (can be remote) initial discrete pustules or fluctuant nodules evolve into deep excavating ulcerations with purulent material, usually sterile. Diagnosis made clinically.
 - Aggressive surgical debridement discouraged as trauma is a trigger for the lesions.
 - Treatment: usually high dose prednisone with taper and/or intra-lesional injections of corticosteroids
- **Oral aphthous ulcers*** (10% UC; 20-30% CD during active disease).
 - Managing underlying IBD is often curative (but note they can also be a side effect of methotrexate)
 - Symptomatic relief: 2% viscous lidocaine + 0.1% triamcinolone
- **Sweet's Syndrome** = acute neutrophilic dermatoses: abrupt onset of tender and red-to-purple inflammatory nodules or papules that coalesce to form plaques.
 - Usually responds well to corticosteroids

Ocular (2-5%)

- ****Episcleritis**** = acute hyperemia, irritation, burning, and tenderness to palpation. Parallels IBD activity.
 - Usually does not need specific treatment other than those for underlying IBD
 - If there is impairment of vision, scleritis must be suspected + prompt referral to ophthalmologist
- **Scleritis** = affects deeper layers of the eye, patients complain of severe pain associated with tenderness to palpation. Deep scleral vessels are hyperemic, along with the episcleral and conjunctival vessels (sclera appears red between the dilated surface vessels)
 - Can cause visual impairment if not diagnosed early -> needs aggressive treatment with systemic steroids and/or immunosuppressants
- **Uveitis** presents as ocular pain, blurred vision, photophobia, and headaches. Less correlated with GI disease
 - Diagnose with inflammatory flare in anterior chamber
 - Prompt diagnosis and treatment with topical and systemic steroids is necessary to prevent secondary glaucoma and rarely blindness

Hepatobiliary

- **Primary Sclerosing Cholangitis** (1.4-7.5%) Nearly 70-80% of PSC patients are known to have concomitant diagnosis of IBD
- **Gallstones** (10% of IBD patients compared to 7% general population)

Pulmonary:

- **bronchiectasis** and **chronic bronchitis**, possibly **asthma**, treated with inhaled steroids

Metabolic bone disease:

- **Osteoporosis and Osteopenia** (overall relative risk of fractures is 40% greater than the general population)
 - American Gastroenterology Association recommends DEXA scan in IBD patients with one or more of the following risk factors: hx vertebral fractures, postmenopausal, male >50 years of age, chronic corticosteroid use, hypogonadism

indicates an extra-intestinal manifestation that signifies IBD flare

References:

- Ardizzone, S et al. **Extraintestinal manifestations of inflammatory bowel disease.** *Digestive and Liver Disease.* 2008; 40.
- Peppercorn, Mark. **Skin and eye manifestations of inflammatory bowel disease.** *UpToDate.* June 2013.
- Trikudanathan G, et al. **Diagnosis and therapeutic management of extra-intestinal manifestations of inflammatory bowel disease.** *Drugs.* 2012 Dec 24; 72(18).