Chylothorax, Lymphangectasia and Inflammatory Bowel Disease

Case Report
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Signalment:
“Jesse” Munskgard, 11 yr old MN Whippet

History:
Jesse presented to the Internal Medicine Service on September 3, 2012 for evaluation of weight loss, inappetence, and intermittent vomiting. Recently, the owners have noted mild tachypnea. Previous history includes intermittent diarrhea felt to be secondary to dietary hypersensitivity. Current diet includes cooked bison, chicken, and chicken broth. Medications include famotidine (5mg orally twice daily) and Cerenia (10mg orally once daily) without any improvement of clinical signs. Bloodwork revealed a mildly elevated BUN (34 units) with a normal creatinine (1.5), hypoproteinemia (4.3), and hypoalbuminemia (2.4 units). Resting cortisol was within normal limits at 3.9 ruling out hypoadrenocorticism.

Clinical Exam:
On presentation, Jesse was quiet, alert, and hydrated with normal heart and breath sounds. He was thin with a BCS 4/9.

Diagnostic Imaging:
On 9/3/12, thoracic radiographs showed a small volume of pleural effusion with right middle lung lobe opacity and volume loss. Diagnostic thoracocentesis was performed which revealed a milky fluid consistent with chyle. Fluid analysis and cytology confirmed a chylous effusion. Bacterial culture was negative. An abdominal ultrasound was also performed on 9/3/12 which showed mild small intestinal wall thickening with an associated thickened mucosal layer. Additionally, the mucosa contained hyperechoic striations consistent with dilated lacteals. These ultrasonographic findings were suggestive of intestinal lymphangectasia and an upper gastrointestinal endoscopy was recommended.

On 9/5/12, an upper GI endoscopy was performed which showed moderate edema, hyperemia, and friability of the duodenal mucosa. Multifocal areas of raised white foci consistent with dilated lacteals were also visualized. The esophagus and stomach were unremarkable. Histopathology revealed moderate to marked multifocal to coalescing lymphoplasmacytic enteritis with random moderate to marked dilated lacteals diagnostic for intestinal lymphangiectasia and inflammatory bowel disease.
Diagnosis:
Intestinal lymphangiectasia, inflammatory bowel disease, and chylothorax.

Treatment/Management:
Jesse was transitioned to a hypoallergenic select protein diet with moderate fat restriction (Royal Canin PW moderate calorie). Jesse was started on prednisone (7.5 mg orally twice daily, 0.5mg/kg twice daily). Other medications included sucralfate (500mg orally every 8 hours), famotidine (5mg orally every 12 hours), cerenia (24mg orally once daily), and rutin (500mg orally every 8 hours). Jesse’s inappetence and weight loss continued and Mirtazepine (7.5mg PO once daily as needed) was added. Jesse's clinical signs persisted at recheck on 9/17/12 and he had mild progression of the pleural effusion. Placement of an esophageal feeding tube was discussed for nutritional support but was declined. The oral prednisone was discontinued and replaced with subcutaneous dexamethasone sodium phosphate (1.5mg subcutaneously once daily) due to the concern of inappropriate absorption of the prednisone. Vitamin B12 injections (750mcg subcutaneously once weekly for 6 weeks, then once monthly), ondansetron (0.5mg/kg subcutaneously once daily) and cyclosporine (50 mg orally once daily) were also started due to lack of response. Jesse began to improve and started gaining weight. He was transitioned back to all oral medications and slowly weaned down on the dosages. He is currently (8 months following diagnosis) on prednisone (2.5mg every other day) and cyclosporine (50mg every other day), famotidine (10mg once daily), and rutin (500mg orally once daily). Jesse is doing great at home and no longer showing any of his previous clinical signs. He has complete resolution of the pleural effusion and has gained 5.8 lbs with a BCS of 5/9.

Discussion:
Intestinal lymphangiectasia is characterized by dilatation and dysfunction of intestinal lymphatics. The abnormal lymphatics leak protein-rich lymph into the intestinal lumen, ultimately causing hypoproteinemia and protein losing enteropathy (PLE). Lymphangiectasia may be a primary disorder or secondary to lymphatic obstruction (such as infiltration or obstruction of lymphatics by an inflammatory, fibrosing, or neoplastic process; obstruction of the thoracic duct; cardiac tamponade; or right heart failure) (1). Primary lymphangiectasia is usually limited to the intestinal tract but can be part of a more systemic lymphatic abnormality involving chylothorax or chylous effusions (2).

Histopathologic confirmation of intestinal lymphangiectasia via endoscopic or surgical biopsy remains the gold standard of diagnosis. Although abdominal ultrasound can allow visualization of intestinal changes that can support lymphangiectasia, endoscopy allows for visualization of the intestinal mucosa. In one study, endoscopic duodenal mucosa appearance alone lacks specificity (42%) and has only moderate sensitivity (68%) for diagnosis of intestinal lymphangiectasia. Evaluation of biomarkers associated with PLE improved the sensitivity, but poor specificity for diagnosis of lymphangiectasia supports the need for histopathology (4).

The goal of primary lymphangiectasia treatment involves decreasing enteric loss of protein and resolve associated inflammation to stop diarrhea, while controlling any edema or effusions. Dietary changes and glucocorticoids are the most important treatments. The ideal diet is markedly fat restricted, calorie dense, and highly digestible diet. Glucocorticoid therapy (1-2mg/kg PO daily and then tapered) and a hypoallergenic or novel diet has been shown to be beneficial, especially if associated with concurrent IBD (inflammatory bowel disease). Anecdotal reports of improvement with glutamine supplementation and cyclosporine treatment have been
seen. Rutin, a benzopyrone extracted from plants, has more commonly been used in human medicine for the treatment of lymphedema following axillary lymph node excision. The exact mechanism of action is unknown; however, it has been proposed that rutin reduces leakage from blood vessels, increases proteolysis and removal of protein from tissues, and enhances macrophage phagocytosis of chyle (3). Isolated case reports have shown that it reduces the accumulation of chylous effusion. Diuretics such as furosemide and spironolactone have also been prescribed to help manage effusions (1). Overall long term prognosis is guarded to poor and many patients ultimately develop severe malnutrition, continuous effusions, or continued diarrhea.

References:
1. Ettinger SJ, Feldman EC. Textbook of Veterinary Internal Medicine, 7th edition, volume 2, pg 1566-1567