Protein-losing enteropathy

A 3 years old girl was admitted for weight loss, diarrhea, abdominal pain and edema.

Her past medical history was unremarkable except for recurrent upper respiratory tract infections, gastro-enteritis and an episode of periorbital edema with increased fecal calprotectin 7 months prior to admission.

The initial work-up revealed hypoalbuminemia, hypochromic microcytic anemia and absence of proteinuria consistent with Protein-losing enteropathy (PLE).

Endoscopy showed edematous, altered mucosa with signs of colitis and histology revealed duodenal lymphangiectasia (Fig A), ileitis with dystrophic epithelium, micro-abscesses, erosive lesions and focal active colitis with chronic architectural changes (Fig B).

She received supportive treatment with albumin, intravenous immunoglobulins, and red blood cell transfusion. Due to lack of response, steroids were introduced with rapid clinical and biological improvement, which were then successfully tapered over the next 4 months. She was also placed on a cow’s milk protein (CMP) free diet due to high suspicion of cow’s milk protein allergy (CMPA). 6 months later endoscopy was normal.

One year later after adequate catch-up growth and height. CMP challenge was followed by a new clinical deterioration confirming the diagnosis of CMPA.

The role of lymphangiectasia in this case remains unclear.

PLE should be considered in patients with hypoproteinemia after exclusion of malnutrition, defective synthesis or proteinuria. Defining the etiology of PLE is challenging, the prognosis of PLE is unknown and depends upon the etiology.

The most common cause in infancy is CMPA.
A. Histology: duodenal lymphangectasia
B. Histology: diffuse ileitis (dystrophic epithelium, micro-abscesses, erosive lesion)