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Case Report

An Infant with Protein-Losing Enteropathy from an Unforeseen Origin

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CASE REPORT

The patient is a 9 month old full term female who initially presented to her pediatrician with failure to thrive. At her 4 month well child check, she was noted to have dropped growth percentiles (head circumference from 51st to 2nd percentile, weight from 25th to 2nd percentile, and length from 85th to 15th percentile) and physical exam revealed periorbital edema. She was exclusively breastfed but started on formula supplementation to facilitate weight gain. In spite of her weight loss, her abdomen always appeared somewhat enlarged, and her parents endorsed fussiness and poor sleep since birth. She did not have diarrhea, changes in stool, fevers, or emesis.

The patient was admitted for further evaluation, at which time she was found to have bilateral periorbital edema and non-pitting lower extremity edema. Laboratory workup revealed hypoalbuminemia (1.7 g/dL initially, <1.5 on repeat), hypoproteinemia (3.1 g/dL), hypomagnesemia (0.8-1.3 mg/dL), hypocalcemia (ical 2.9-3 mg/dL), low vitamin A, D, and E levels (16, <7, and 2.3 mg/dL, respectively), and a low IgG level (<75 mg/dL). Given her laboratory abnormalities, edema, and ascites, she was diagnosed with a protein-losing enteropathy (PLE), specifically intestinal lymphangiectasia. She underwent upper endoscopy that revealed minimal patchy dilation of the lymphatic spaces. A central venous catheter was placed and nutrition therapy was started with TPN and a PO high MCT, low LCT formula.

Two months later, her central venous catheter was found to be leaking from the line site, so she underwent catheter replacement. The patient became hypertensive post-operatively, leading to PICU admission and moderate blood pressure control with labetalol and hydralazine. She underwent an extensive hypertension workup including renal ultrasound with doppler, echocardiogram, and chest radiograph, all of which were normal. She then had an abdominal ultrasound that revealed a mass concerning for neuroblastoma. At this time, she was transferred to the hematology/oncology service and underwent abdominal MRI that showed a large midline mass replacing the bulk of the pancreas and surrounding the aorta, celiac, superior mesenteric and renal arteries (Figures 1,2). A subsequent retroperitoneal tumor biopsy confirmed the diagnosis of neuroblastoma.

VMA and HVA levels were elevated at 411.7 and 144.2mg/g Cr, respectively, and further diagnostics revealed metastasis only to her right femur along with negative N-Myc stain. Her disease was characterized as stage IV, intermediate risk neuroblastoma. Chemotherapy was initiated with carboplatin and etopside, along with daily filgrastim. Her TPN was tapered off and replaced with a PO low LCT, high MCT formula, and she began weekly intralipids and PO essential fatty acid oils. She is currently doing well on this regimen and continued chemotherapy. Repeat MRI at 9 months of age showed interval decrease in her midline mass (Figures 3,4).

PLE is a rare first manifestation of neuroblastoma, with only five previously reported cases [1-5]. In all of these children, diarrhea was one of the presenting signs. To our knowledge, our patient represents the first recorded case of neuroblastoma manifesting as PLE without diarrhea. The following mechanisms have been proposed linking neuroblastoma to PLE: inflamed or ulcerated mucosa allowing protein loss, direct tumor infiltration,

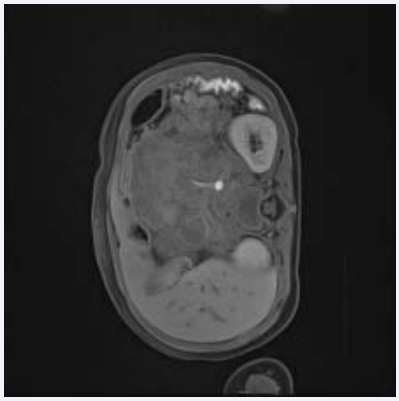

Figure 1 T1 non contrast MRI showing midline abdominal mass.

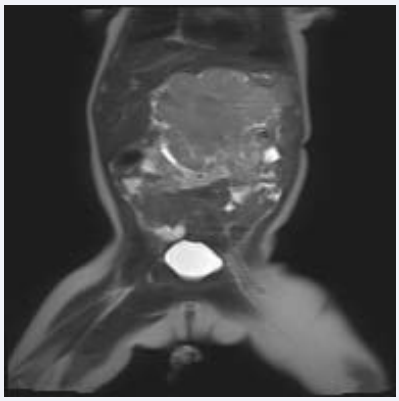

Figure 2 T2 contrast MRI showing midline abdominal mass.

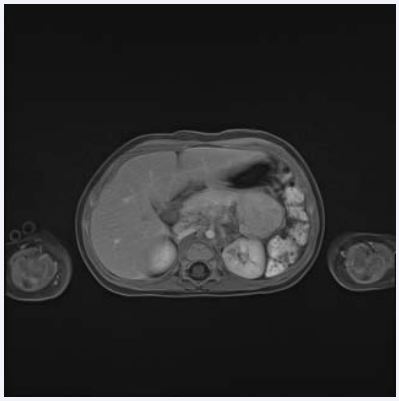

Figure 3 T1 non contrast MRI showing interval decrease in midline abdominal mass.

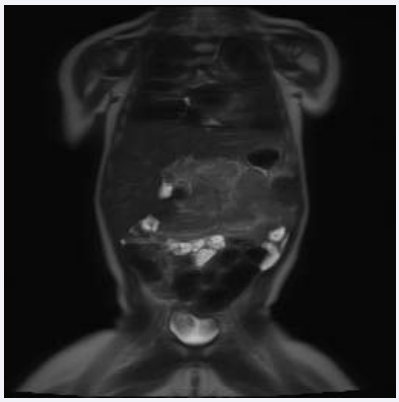

Figure 4 T2 contrast MRI showing interval decrease in midline abdominal mass.

chronically elevated right-sided venous pressure due to carcinoid syndrome, and neurohumoral effects from tumor secretions such as VIP or catecholamines, and lymphatic obstruction by tumor [1]. The latter mechanism describes intestinal lymphangiectasia, the probable etiology in this case.

When faced with a case of PLE, it is important to consider a broad range of underlying causes, including conditions leading to increased mucosal permeability such as celiac disease, hypertrophic gastropathy, or bacterial overgrowth, erosive/inflammatory disease such as inflammatory bowel disease or infection, and other sources of lymphatic fluid loss/obstruction such as congestive heart failure, sarcoidosis, or malignancy [6]. In this case, identification of a mass on ultrasound allowed for rapid narrowing of the differential and ultimately to the discovery of neuroblastoma.

In light of this and previous cases, we recommend that physicians retain a reasonable index of suspicion for secondary causes of PLE, such as neuroblastoma, in patients with failure to thrive, edema, and hypoalbuminemia, with or without diarrhea. In general, prognosis of neuroblastoma inversely correlates with age at diagnosis, underlying the importance of early disease detection [7].

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