Gastrointestinal Hemangiomatosis
Joseph Junewick, MD FACR

03/06/2010

History
3 month old with protuberant abdomen and anemia.

Diagnosis
Gastrointestinal Hemangiomatosis

Discussion
Gastrointestinal hemangiomatosis is a complex vascular malformation that occurs primarily in infancy and childhood although occasionally it is not diagnosed until adulthood. The clinical presentation is variable, although most patients present with gastrointestinal bleeding. Patients may also present with intussusception, small-bowel obstruction, perforation, or malabsorption. Cutaneous hemangiomas are often present. Gastrointestinal hemangiomatosis may be associated with blue rubber bleb nevus syndrome, Olser-Weber-Rendu, Klippel-Trénaunay-Weber syndrome, Maffucci’s syndrome, Kasabach-Merritt syndrome, vonHippel-Lindau syndrome, diffuse neonatal hemangiomatosis, and Proteus syndrome. Hemangiomatosis is manifested by diffuse infiltration of the intestinal wall, the mesentery, and, occasionally, the retroperitoneum. Solid organs in the abdomen may also be involved.

The radiographic findings of hemangiomatosis include the presence of phleboliths on abdominal radiographs, scattered submucosal small-intestine nodules on barium examination, and mural thickening with phleboliths on CT. If gastrointestinal hemangiomatosis is suspected, water or fat enteric contrast should be employed instead of positive contrast.

Numerous therapies have been used in an attempt to treat hemangiomas when complications develop during the proliferative phase. The prolonged use of systemic agents during the period of endothelial proliferation is often associated with increased side effects. The current first line of treatment is systemic administration of corticosteroids. Approximately 30% of hemangiomas will respond dramatically to corticosteroids and another 40% will have some response. Side effects of steroid therapy include severe irritability, weight gain, cushingoid appearance, growth delay, hypertension, diabetes, gastroesophageal reflux, and susceptibility to infections. Other antiangiogenesis drugs such as vincristine and vincristine regimens (VAC) are also successful. Alpha-interferon therapy has been used in the past but is associated with irreversible neurologic spastic diplegia.

Findings
CT-Diffuse mesenteric and enteric hemangiomatous infiltration. Note the marked enlargement of the portal venous system and marked attenuation of the abdominal aorta distal to the superior mesenteric artery.
MR-Coronal T2 images confirm the extensive hemangiomatous involvement of the bowel and mesentery. MR arteriography and venography also confirm the vascular alterations noted on CT.

Reference
Scafidi DE, McLeary MS, Young LW. Diffuse neonatal gastrointestinal hemangiomatosis: CT findings.
Pediat Radiol (1998); 28(7):512-514.
Disclaimer
This teaching site is partially funded by an educational grant from GE Healthcare and Advanced Radiology Services, PC. The material on this site is independently controlled by Advanced Radiology Services, PC, and GE Healthcare and Spectrum Health have no influence over the content of this site.

Content Download Agreement
The cases and images on this website are owned by Spectrum Health. Permission is granted (for nonprofit educational purposes) to download and print materials to distribute for the purpose of facilitating the education of health professionals. The authors retain all rights to the material and users are requested to acknowledge the source of the material.

Site Disclaimer
This site is developed to reach healthcare professionals and medical students. Nothing this site should be considered medical advice. Only your own doctor can help you make decisions about your medical care. If you have a specific medical question or are seeking medical care, please contact your physician.

The information in this website is provided for general medical education purposes only and is not meant to substitute for the independent medical judgment of a physician relative to diagnostic and treatment options of a specific medical condition.

The viewpoints expressed in these cases are those of the authors. They do not represent an endorsement. In no event will Advanced Radiology Associates, PC, Spectrum Health Hospitals (Helen Devos Children's Hospital) or GE Healthcare be liable for any decision made or action taken in reliance upon the information provided through this website.