**GI Vascular Anomalies: Diagnosis and Management**

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NASPGHAN Annual Meeting

Atlanta 2014

Gastrointestinal vascular anomalies are a morphologically and biologically diverse group of abnormalities of vascular channels that are often congenital but may grow or change with time. They may involve single vessel types or combinations of capillaries, veins, arteries, and lymphatics. They can be isolated lesions or part of a complex multisystem disorder.

Vascular anomalies are clinically challenging for several reasons. Lesions are rare and may not be easily recognized. Non-standardized terminology leads to miscommunication between medical providers. Poorly understood pathobiology and lack of coordinated multidisciplinary leads to ineffective therapy.

Vascular anomalies are primarily classified as either tumors or malformations based on their biological and clinical behavior and their physical endothelial characteristics. This classification predicts the best avenues for therapy such as antiproliferative drug therapy versus surgical or other ablative interventions. More detailed classification splits tumors into types that are either benign, e.g. infantile hemangioma, locally aggressive, e.g. kaposiform hemangioendothelioma, or malignant, e.g. angiosarcoma. Malformations are subclassified as either simple (single channel type), e.g. venous malfomation, combined (2 or more channel types), e.g. arteriovenous malformation, specific vessel, e.g. aortic aneurism, or complex conditions associated with other anomalies, e.g. Klippel-Trenaunay VAC) tract. This may represent a referral bias due to a historically surgery-oriented center.

Typical clinical GI presentations include overt and occult GI bleeding, abdominal pain and intestinal obstruction, and malabsorption when associated with diffuse lymphangiectasia. Infantile hemangioma is the most common tumor of infancy and childhood, 1 to 2.6% neonates. The ratio of female to male gender is 3:1, multifocal distribution is seen in 20%, and extracutaneous sites (especially liver, lung, brain, and intestine) are more common if > 5 skin lesions are seen. Intestinal hemangioma is very rare: 14/16 (88%) cases in 14 year review of BCH VAC database presented with GI bleeding in first 4 months of life, 9/16 had cutaneous lesions, 7/16 had associated congenital abnormalities. Preferred therapy is propranolol or corticosteroids. Venous malformations are among the most common vascular malformations of the GI tract and are represented by focal segmental transmural lesions, multifocal polypoid lesions (blue rubber bleb nevus syndrome), and multifocal or diffuse reticular lesions (varices and telangiectasias). Therapies include surgical excision, bowel resection, sclerotherapy, and thermal cautery or ablation. Lymphatic anomalies and intestinal lymphangiectasia are among the most difficult to treat and poorly understood lesions. Some conditions such as cutaneovisceral angiomatosis with thrombocytopenia (CAT), also called multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT), share features of both tumors and malformations and remain in a category of provisionally unclassified vascular anomalies. CAT clinically improves with antiproliferative therapy but resolution is incomplete and long-term outcome remains poorly defined.

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