



Gliomatosis Peritonei

Gliomatosis peritonei (GP) refers to a very rare entity characterized by miliary disseminated implants composed of mature glial (fibrous astrocytes) tissue throughout the serosal surface of the abdominal peritoneal cavity including recesses, cul-de-sac and intestinal surfaces. Though the picture is of an advance stage of neoplasia, its behavior is almost invariably benign since its differentiated cells lack proliferative activity. The majority of cases of GP are associated with an immature ovarian teratoma, though only rarely with mature teratoma. Almost 25% of immature teratoma can develop GP. Hypothesis for development of GP includes implantation and subsequent maturation of neural precursor cells detached from the primary tumor or peritoneal metaplasia from stem cells induced by growths factors. Peak incidence of GP is in the second decade with an average age of 15 years. GP can occur after capsular rupture during surgery or spontaneously. The overall prognosis of mature GP is excellent and chemotherapeutic management is rarely necessary except with high-grade or immature deposits. Implants can grow rapidly. Serum markers such as CA125 and alpha fetoprotein are elevated in GP. Histologically GP demonstrates large amounts of well-differentiated glial tissue with extensive endothelial proliferation of vessels similar to brain tumors, minimal atypia and only rare mitosis. Surgical sampling should be as extensive as possible to evaluate the maturity of the glial deposits to establish a diagnosis of GP. Immature glial tissue or other teratomatous component heralds metastatic disease and should be managed aggressively. GP does not adversely affect the prognosis of ovarian teratoma. A conservative surgical approach without adjuvant therapy or chemotherapy is recommended.

References:

- 1- Harms D, et al: Gliomatosis peritonei in childhood and adolescence. Clinicopathological study of 13 cases including immunohistochemical findings. *Pathol Res Pract.* 184(4):422-30, 1989
- 2- Chaung JH, Chen L: Ovarian teratoma with gliomatosis peritonei. *J Pediatr Surg.* 27(5):662-4, 1992
- 3- Hamada Y(1), Tanano A, Sato M, Tsuji M, Sakaida N, Okamura A, Hioki K: Ovarian teratoma with gliomatosis peritonei: report of two cases. *Surg Today.* 28(2):223-6, 1998
- 4- Hill DA(1), Dehner LP, White FV, Langer JC: Gliomatosis peritonei as a complication of a ventriculoperitoneal shunt: case report and review of the literature. *J Pediatr Surg.* 35(3):497-9, 2000
- 5- Hsieh YL, Liu CS: Progression from an immature teratoma with miliary gliomatosis peritonei to growing teratoma syndrome with nodular gliomatosis peritonei. *Pediatr Neonatol.* 50(2):78-81, 2009
- 6- Yoon NR, Lee JW, Kim BG, Bae DS, Sohn I, Sung CO, Song SY: Gliomatosis peritonei is associated with frequent recurrence, but does not affect overall survival in patients with ovarian immature teratoma. *Virchows Arch.* 461(3):299-304, 2012

Neutropenic Colitis

Neutropenic colitis (NC) refers to a potentially life-threatening necrotizing inflammation of the cecum and colon the result of a chemotherapeutic complication of leukemia treatment, acquired immunodeficiency syndrome or as a complication of bone marrow transplantation. The clinical triad of neutropenic colitis includes neutropenia, abdominal pain/tenderness and fever. The integrity of the bowel wall is compromised due to factors including mucosal injury by cytotoxic drugs, neutropenia, and impaired host defense to intestinal organisms. This leaves the bowel vulnerable to bacterial invasion, necrosis and perforation. Most cases involve the cecum and ascending colon. It is sometimes difficult to differentiate NC from appendicitis in the early onset. Critical key points for diagnosis of NC are neutropenia (absolute neutrophil counts below 500 cells/ μ L), high fever, bloody stool and aggressive progress. Diagnostic accuracy is increased with the use of US or CT-Scans when there is a bowel wall thickening above 3 mm. Factors associated with development of NC include age greater than 16 years, mucositis, stem cell transplantation and chemotherapy within the prior two weeks of symptoms. Initial management should consist of aggressive hemodynamic support, bowel rest/decompression, supplemental nutrition, broad-spectrum antibiotic therapy and G- CSF. Measurements of C-reactive protein in blood may be of benefit when assessing the clinical course. Indications for surgical intervention include diffuse peritonitis, pneumoperitoneum, persistent gastrointestinal bleeding and continued clinical deterioration despite medical therapy. The most common surgical indication is bowel perforation. Recommended surgical procedures include hemicolectomy, ileostomy and secondary anastomosis or drainage. Primary bowel anastomosis is not a good choice due to the high risk of leakage, intraperitoneal infection, abscess, intestinal adhesion and obstruction. Most children do not require surgery. Overall mortality is lower in children (5%).

References:

- 1- Moran H, Yaniv I, Ashkenazi S, Schwartz M, Fisher S, Levy I: Risk factors for typhlitis in pediatric patients with cancer. *J Pediatr Hematol Oncol.* 31(9):630-4, 2009
- 2- Sundell N, Bostram H, Edenholm M, Abrahamsson J: Management of neutropenic enterocolitis in children with cancer. *Acta Paediatr.* 101(3):308-12, 2012
- 3- McCarville MB(1), Adelman CS, Li C, Xiong X, Furman WL, Razzouk BI, Pui CH, Sandlund JT: Typhlitis in childhood cancer. *Cancer.* 104(2):380-7, 2005
- 4- Schlatter M(1), Snyder K, Freyer D: Successful nonoperative management of typhlitis in pediatric oncology patients. *J Pediatr Surg.* 37(8):1151-5, 2002
- 5- Fike FB(1), Mortellaro V, Juang D, St Peter SD, Andrews WS, Snyder CL: Neutropenic colitis in children. *J Surg Res.* 170(1):73-6, 2011
- 6-Li K, Zheng S, Dong K, Gao Y, Wang H, Liu G, Gao J, Xiao X: Diagnosis and outcome of neutropenic enterocolitis: experience in a single tertiary pediatric surgical center in China. *Pediatr Surg Int.* 27(11):1191-5, 2011

MACIS

Thyroid carcinoma is the third most common solid tumor in children and adolescent. The incidence is rising. Most cases of thyroid carcinoma are well differentiated papillary or follicular tumors. As previously stated they are best managed with total

thyroidectomy and postoperative radioiodine therapy in most cases. There are several scoring systems used to classify thyroid cancer. MACIS refer to a recently developed prognostic scoring system utilized for differentiated thyroid carcinoma in children and adults that has resulted in recognition of low- and high risk patient categories allowing meaningful comparison of a variety of treatment approaches. MACIS stand for the presence of Metastasis, Age, Completeness of resection, local Invasion and tumor Size. MACIS was designed in 1993 to predict disease specific survival in patients with differentiated thyroid carcinoma. The cutoff score has been set at a score above four heralding a poor prognosis for children less than 21 years of age. MACIS score correlates well with the response to initial therapy in children and is useful in predicting outcome. MACIS appear the most useful prognostic system taking completeness of resection into account.

References:

- 1- Palme CE(1), Waseem Z, Raza SN, Eski S, Walfish P, Freeman JL: Management and outcome of recurrent well-differentiated thyroid carcinoma. Arch Otolaryngol Head Neck Surg. 130(7):819-24, 2004
- 2- Voutilainen PE(1), Siironen P, Franssila KO, Sivula A, Haapiainen RK, Haglund CH: AMES, MACIS and TNM prognostic classifications in papillary thyroid carcinoma. Anticancer Res. 23(5b):4283-8, 2003
- 3- Powers PA(1), Dinauer CA, Tuttle RM, Francis GL: The MACIS score predicts the clinical course of papillary thyroid carcinoma in children and adolescents. J Pediatr Endocrinol Metab. 17(3):339-43, 2004
- 4- Kjellman P(1), Zedenius J, Lundell G, Bäckdahl M, Farnebo LO, Hamberger B, Larsson C, Wallin G: Predictors of outcome in patients with papillary thyroid carcinoma. Eur J Surg Oncol. 32(3):345-52, 2006
- 5- Jang HW(1), Lee JI, Kim HK, Oh YL, Choi YL, Jin DK, Kim JH, Chung JH, Kim SW: Identification of a cut-off for the MACIS score to predict the prognosis of differentiated thyroid carcinoma in children and young adults. Head Neck. 34(5):696-701, 2012

* Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP

Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,
 Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Childrens Hospital.
 Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
 Tel (787)-786-3495 Fax (787)-720-6103 E-mail: titolugo@coqui.net
 Internet: <http://home.coqui.net/titolugo>

©PSU 1993-2014
 ISSN 1089-7739