PEDIATRIC SURGERY *Update* ©
Vol. 45 No. 05 NOVEMBER 2015

Macrophage Activation Syndrome

Macrophage activation syndrome (MAS) is a severe, potentially fatal condition caused by excessive activation and development of macrophages and T-cells (mainly CD8+) leading to an overwhelming inflammatory reaction in the host (cytokine storm). The main clinical manifestation of macrophage activation syndrome includes fever, hepatosplenomegaly, lymphadenopathy, severe cytopenia, liver dysfunction and coagulopathy consistent with disseminated intravascular coagulation. MAS is also associated with extreme hyperferritinemia. A pathognomonic feature of MAS is the expansion of well-differentiated macrophages exhibiting hemophagocytic activity typically found in bone marrow and lymph nodes. MAS is most strongly associated with the systemic form of juvenile idiopathic arthritis, but it can also occur in patients with systemic lupus erythematosus, Kawasaki disease juvenile dermatomyositis, antiphospholipid syndrome and mixed connective tissue disease. Diagnosis of MAS is difficult due to its resemblance to sepsis. In a child with persistently active underlying rheumatologic disease, a fall in the ESR and platelet count, particularly in a combination with persistently high CRP and increasing levels of serum D-dimer and ferritin, should raise a suspicion of impending MAS. Diagnosis of MAS is usually confirmed by the demonstration of hemophagocytosis in the bone marrow. Treatment options include a pro-apoptotic chemotherapy. Patients with evidence of continued or progressive central nervous system involvement after two weeks of systemic therapy require intrathecal therapy with methotrexate combined with corticosteroid induction followed by hematopoietic stem cell transplantation. The role of the surgeon in this rare condition includes central venous catheter placement and lymph node biopsy as warranted for the treatment and diagnosis of MAS.

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MeSS Score

Well-differentiated thyroid carcinoma is the third most common solid malignancy in children with a 2% rise in a year incidence. It is more commonly seen in females in their adolescent years with the papillary histologic variant and more than 40% of cases having cervical node disease upon presentation. Surgery (total thyroidectomy) is the mainstay method of management producing along with the use of postoperative radioiodine therapy an excellent long term survival in most cases. Many clinical variables have been utilized to predict prognosis with varying amounts of scientific certainty such as histological tumor type, age, gender, extent of primary-site disease, presence or absence of nodal disease, extent of thyroidectomy, and the use of radioactive iodine ablation. Several studies have found male gender, increasing primary disease site extension, and follicular histologic subtype all had negative prognostic influence on overall survival. The adult prognostic scales of thyroid carcinoma utilized such as AGES (age, grade, extent of disease, size), AMES (age, metastasis, extent of disease, size), MACIS (metastasis, age at presentation, completeness of surgical resection, invasion, size), and TNM (tumor, node, metastasis) are not reliable in children. Instead recent evidence has found in children that the presence of distant metastasis, tumor size and gender are independent predictors of mortality at the time of diagnosis. Based on these findings, a final prognostic scale was created and defined as distant metastasis (Me), larger primary tumor size (S), and male sex (S) (MeSS) = +5 (if distant metastasis present), +2 (if primary tumor size > 4 cm), and +3 (if male gender). MeSS score < 2 has a 0% mortality, 2-7 moderate risk (2.7% mortality) and >7 has high risk (23% mortality).

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Lymphoplasmacytic Sclerosing Pancreatitis

Lymphoplasmacytic sclerosing pancreatitis (LPSP) is a very rare cause of chronic pancreatitis occurring in the pediatric age group that can mimic a malignant pancreatic lesion. LPSP is an autoimmune from of chronic pancreatitis found most commonly in elderly men. It occurs in the absence of gallstone pancreatitis, pancreas divisum, or excess alcohol ingestion. It is a diffuse fibrosing process of the pancreas considered an autoimmune condition associated with inflammatory bowel disease, Sjögren syndrome,

primary biliary cirrhosis and atopic conditions. It has been proposed that an autoimmune mechanism against carbonic anhydrase II or lactoferrin and a Th1-type immune response may be involved in LPSP. Histologically it can be classic or intermediate in appearance. Classic LPSP has lymphoplasmacytic infiltration of the pancreas, interstitial fibrosis, periductal inflammation, and periphlebitis, while intermediate LPSP includes patients with at least two of these histological findings. Clinically the patient with LPSP presents with weight loss, jaundice, abdominal pain, elevated CEA and CA-19 levels similar to patients with pancreatic cancer. On imaging (CT, MRI, ERCP) the mass is usually in the head of the pancreas with a more diffuse appearance and irregular narrowing of the pancreatic dust as compared to the more discrete form of mass of an adenocarcinoma. LPSP produces high serum levels of IgG4, a rare gamma globulin subclass that is not elevated in pancreatic cancer. FNA biopsy of the lesion can bring conflicting results. FNA histopathological examinations of the pancreas show fibrosis and pronounced infiltration of cells, mainly lymphocytes and plasmacytes. Initial management of LPSP should consist of systemic steroid therapy which can cause remission of the mass to normal level when follow-up with imaging studies in most cases.

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***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)-999-9450 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*

Internet: http://home.coqui.net/titolugo

ISSN 1089-7739