



PEDIATRIC SURGERY Update ©

Vol 15 No 03 SEPTEMBER 2000

'Official Publication of the Puerto Rico Association of Pediatric Surgeons'

Askin Tumor

Askin tumor (synonyms are primitive neuroectodermal tumor or Ewing's sarcoma) is a malignant small round cell tumor of mesenchymal origin affecting the thoracopulmonary region of children and young adults with a tendency to recur locally. The rib is the most common site of primary tumor development. Establishing an accurate preoperative diagnosis of Askin's tumor is difficult. Microscopy and immunohistological stain of the specific marker - neuron-specific enolase, is essential. CT scan is valuable for evaluating tumor extension at diagnosis, the effects of chemotherapy and assessing recurrence after surgery, but can overestimate pleural, lung or diaphragmatic infiltration. MRI can determine chest wall muscle and marrow involvement. Neither is adequate for adjacent lung invasion. Bone, bone marrow and lung are the most frequent sites of metastasis. Treatment includes radical surgical resection (including affected lung tissue), neoadjuvant (local control of disease is critical) and adjuvant chemotherapy plus radiation. Surgical resection, with en bloc removal of involved structures and chest wall reconstruction, provides excellent local control of malignant chest wall tumors. Human dura, prosthetic material (Gortex, Marlex, Vicryl) and myocutaneous flaps have been used for reconstruction. Patients with Askin tumors treated with aggressive pre-resection chemotherapy have smaller tumors to resect (less than 100 cc by volume) with improved survival. Overall the prognosis is poor.

References:

- 1- Dang NC, Siegel SE, Phillips JD: Malignant chest wall tumors in children and young adults. *J Pediatr Surg* 34(12):1773-8, 1999
- 2- Sawin RS, Conrad EU 3rd, Park JR, Waldhausen JH: Preresection chemotherapy improves survival for children with Askin tumors. *Arch Surg* 131(8):877-80, 1996
- 3- Walton JM, Bass J, Sambey E, Rubin SZ: Use of human dura in pediatric chest wall reconstruction after tumor resection. *J Pediatr Surg* 29(9):1189-91, 1994
- 4- Shamberger RC, Tarbell NJ, Perez-Atayde AR, Grier HE: Malignant small round cell tumor (Ewing's-PNET) of the chest wall in children. *J Pediatr Surg* 29(2):179-84, 1994
- 5- Bourque MD, Di Lorenzo M, Collin PP, Russo P, Laberge JM, Moir C: Malignant small-cell tumor of the thoracopulmonary region: 'Askin tumor'. *J Pediatr Surg* 24(10):1079-83, 1989
- 6- Shamberger RC, Grier HE, Weinstein HJ, Perez-Atayde AR, Tarbell NJ: Chest wall tumors in infancy and childhood. *Cancer* 15;63(4):774-85, 1989
- 7- Parikh PM, Charak BS, Banavali SD, Advani SH, Saikia TK, Gopal R, Borges AM, Chinoy RF, Desai PB: Treatment of Askin Rosai tumor--need for a more aggressive approach. *J Surg Oncol* 39(2):126-8, 1988
- 8- Grosfeld JL, Rescorla FJ, West KW, Vane DW, DeRosa GP, Provisor AJ, Weetman R: Chest wall resection and reconstruction for malignant conditions in childhood. *J Pediatr Surg* 23(7):667-73, 1988
- 9- Askin FB, Rosai J, Sibley RK, Dehner LP, McAlister WH: Malignant small cell tumor of the thoracopulmonary region in childhood: a distinctive clinicopathologic entity of uncertain histogenesis. *Cancer* 43(6):2438-51, 1979

Gastric Duplication

Gastric duplications cysts account for less than 5% of all enteric duplications (the rarest form). As a duplication it is attached to its origin, has a well developed smooth muscle coat and gastric epithelial lining. Prenatal ultrasound finding of a cyst with peristaltic activity within the right upper quadrant of the fetal abdomen suggest the diagnosis. The most common site of origin of the duplication cyst is the greater curvature. Most are closed spherical cysts. Most cases are diagnosed during the first two years of life and are more common in females. The usual presentation is an abdominal mass with vomiting. Complications reported consist of recurrent pancreatitis, hemorrhage, perforation, peritonitis, torsion and malignant change in gastric mucosa. Contrast studies, US or CT-Scan suggests the diagnosis. Management of a gastric duplication cyst is surgical excision that can be accomplished laparoscopically. Gastric mucosal along heterotopic pancreatic tissue can be found in the cyst wall.

References:

- 1- Bidwell JK, Nelson A: Prenatal ultrasonic diagnosis of congenital duplication of the stomach. *J Ultrasound Med* 5(10):589-91, 1986
- 2- Sieunarine K, Manmohansingh E: Gastric duplication cyst presenting as an acute abdomen in a child. *J Pediatr Surg* 24(11):1152, 1989
- 3- Bajpai M, Mathur M, Duplications of the alimentary tract: clues to the missing links. *J Pediatr Surg* 29(10):1361-5, 1994
- 4- Blais C, Masse S: Preoperative ultrasound diagnosis of a gastric duplication cyst with ectopic pancreas in a child. *J Pediatr Surg* 30(9):1384-6, 1995
- 5- Koumanidou C, Montemarano H, Vakaki M, Pitsoulakis G, Savvidou D, Kakavakis K: Perforation of multiple gastric duplication cysts: diagnosis by sonography. *Eur Radiol* 9(8):1675-7, 1999

Bile Duct Rhabdomyosarcoma

The botryoid variety of embryonal Rhabdomyosarcoma (RMS) is the most common tumor of bile ducts presenting during early life. Peak incidence at three to four years with a slight female predominance. The tumor is characterized by multiple polypoid grape-like projections into the lumen of the common bile duct with plate-like thickening of the common bile duct wall. It is characterized by a high risk of local recurrence to adjacent lymph nodes and a low risk of remote metastasis. Obstructive jaundice, cachexia, pain and abdominal mass are the usual presentation, often with fever and hepatomegaly. Attribution of these symptoms to hepatitis commonly delays definitive treatment. Other times the preoperative diagnosis is mistaken for a choledochal cyst. US defines the relationship of the tumor with portal vessels and biliary tract while CT-Scan and MRI determine operability. Aggressive surgery combined with the new adjuvant therapies (chemotherapy and radiotherapy) appears to provide the best chance for a longer survival. Intra-operative cholangiography is a valuable technique in establishing the level of biliary tree obstruction and verifying a functioning drainage procedure. The prognosis is poor and death is usually due to the effects of local invasion by the tumor.

References:

- 1- Taira Y, Nakayama I, Moriuchi A, Takahara O, Ito T, Tsuchiya R, Hirano T, Matsushita T: Sarcoma botryoides arising from the biliary tract of children. A case report with review of the literature. *Acta Pathol Jpn* 26(6):709-18, 1976
- 2- Lack EE, Perez-Atayde AR, Schuster SR: Botryoid rhabdomyosarcoma of the biliary tract. *Am J Surg Pathol* 5(7):643-52, 1981
- 3- Martinez-F LA, Haase GM, Koep LJ; Akers DR: Rhabdomyosarcoma of the biliary tree: the case for aggressive surgery. *J Pediatr Surg* 17(5):508-11, 1982
- 4- Ruymann FB, Raney RB Jr, Crist WM, Lawrence W Jr: Rhabdomyosarcoma of the biliary tree in childhood. A report from the Intergroup Rhabdomyosarcoma Study. *Cancer* 56(3):575-81, 1985
- 5- Geoffray A, Couanet D, Montagne JP, Leclere J: Ultrasonography and computed tomography for diagnosis and follow-up of biliary duct rhabdomyosarcomas in children. *Pediatr Radiol* 17(2):127-31, 1987
- 6- von der Oelsnitz G, Spaar HJ, Lieber T, Munchow B, Booss D: Embryonal rhabdomyosarcoma of the common bile duct. *Eur J Pediatr Surg* 1(3):161-5, 1991
- 7- Sanz N, de Mingo L, Florez F, Rollan V: Rhabdomyosarcoma of the biliary tree. *Pediatr Surg Int* 12(2-3):200-1, 1997
- 8- Balkan E, Kiristioglu I, Gurpinar A, Sinmaz K, Ozkan T, Dogruyol H: Rhabdomyosarcoma of the biliary tree. *Turk J Pediatr* 41(2):245-8, 1999

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

Associate Professor of Pediatric Surgery, University of Puerto Rico School of Medicine and University
Pediatric Hospital, Rio Piedras, Puerto Rico.

Address - P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico 00922-0426.

Tel (787)-786-3495 Fax (787)-720-6103 E-mail: *titolugo@coqui.net*

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

© PSU 1993-2000