

PEDIATRIC SURGERY Update Vol. 52 No. 02 FEBRUARY 2019

Pediatric Breast Cancer

Breast masses in female children are mainly benign with fibroadenoma being the most common diagnosis. Breast cancer in the pediatric age is very rare occurring in 0.1% of all breast cancers with an estimated incidence of one case for every 125000 females under the age of 21 years. When breast cancer occurs in the pediatric age, the child presents with late stage disease, more nodal metastasis and lymphovascular invasion and few to none hormone receptor positive tumor. Breast cancer is the most common cancer in females between 15 and 40 years of age. The median age in the pediatric group with breast cancer is 17 years with 80% of cases in nonblack patients. Common symptoms include a lump, changing size of the breast, dimple of the skin, inverted nipple, and red or swollen skin changes in the breast. US and MRI suggest the presence of a malignancy and biopsy confirms the diagnosis. Suspicious lesions of any size with atypical findings on ultrasound or MRI (cystic areas, hyperlobulation, angularity, calcifications or irregular margins) or masses above 4 cm should undergo core or excisional biopsy. The most common histology in order of frequency is breast adenocarcinoma (50%), fibroepithelial/phylloides tumors (35%) and sarcomas (15%). Most cases undergo surgery as management of the tumor including modified mastectomy. Adenocarcinomas and sarcoma carry a worse survival when compared with fibroepithelial tumor. By stage fibroepithelial tumors is almost all early stage disease, whereas adenocarcinomas and sarcomas have both early and advanced stage tumors. High grade and advanced stage disease has a worse outcome than low grade and early stage disease. Fibroepithelial tumors are nearly as twice as common in black patients when compared to nonblack patients. This is probably caused by an early menarche age in black females with longer exposure to estrogen. Fibroepithelial tumors arise from the specialized connective tissue around mammary lobules and are often large (> 6 cm) estrogen-sensitive tumors with a high risk of local recurrence. Almost 20% of pediatric breast cancer dies of their disease.

References:

1- Westfal ML, Chang DC, Kelleher CM: A population-based analysis of pediatric breast cancer. J Pediatr Surg. 2018 Oct 5. pii: S0022-3468(18)30627-4. doi:10.1016/j.jpedsurg.2018.10.009.

2- Chen HL, Zhou MQ, Tian W, Meng KX, He HF: Effect of Age on Breast Cancer Patient Prognoses: A Population-Based Study Using the SEER 18 Database. PLoS One. 2016 Oct 31;11(10):e0165409. doi: 10.1371/journal.pone.0165409. eCollection 2016.

3- Ademuyiwa FO, Gao F, Hao L, Morgensztern D, Aft RL, Ma CX, Ellis MJ: US breast cancer mortality trends in young women according to race. Cancer. 121(9):1469-76, 2015

4- Gutierrez JC, Housri N, Koniaris LG, Fischer AC, Sola JE: Malignant breast cancer in children: a review of 75 patients. J Surg Res. 147(2):182-8, 2008

5- Wang XX, Jiang YZ, Liu XY, Li JJ, Song CG, Shao ZM: Difference in characteristics and outcomes between medullary breast carcinoma and invasive ductal carcinoma: a population based study from SEER

18 database. Oncotarget. 2016 Apr 19;7(16):22665-73. doi: 10.18632/oncotarget.8142.

Dog Bites Injury

More than 4.5 million dog bites occur in the USA annually requiring hospitalization in more than 80% of them. It is estimated that 60 million's dollars are spent annually for dog-related hospitalizations. Children are a specially vulnerable population at higher risk for sustaining dog bite injuries. The most common circumstance for injury involved a child playing with or while petting a dog. This benign initial encounter should be highly monitored during early interactions between a dog and child. Food-related encounters like a dog protecting its food or a child eating near a dog are common circumstances of injury. Dog bites leads to life and limb threatening infections in children due to the ischemic nature of tissue loss and mixed bacterial contamination. Infants are more than four times likely to be bitten by the family dog and most of these occurs in the head, face and neck region. Preschool children dog bites accounts for more than half of the injured population. Most dog bites can be repaired in the emergency room with primary closure. Complex lacerations, avulsions and shearing injuries will need repair at the operating room. Pit bulls are the most common breed to inflict dog bite injury requiring direct and reconstructive repair. Children between ages nine and 12 years are the most common victims of pit bulls attacks. Following pit bulls, Labrador retriever, German Shepard and Husky follow in as much as attacking dog breed. Location of dog bites in children is usually the head and face region due to child short stature. larger head circumference and underdeveloped evasive motor skills to protect the face. Simple puncture wounds and abrasions can be managed with wound irrigation and application of nonadherent dressing. Simple lacerations can be sutured in the emergency room under local sedation and analgesics. Complex lacerations, partial flap avulsions or complete full-thickness tissue loss requires operative repair and hospitalization. Broad-spectrum antibiotic coverage is recommended for all dog bite wounds that require operative intervention. Extended hospitalization is usually associated with local wound infection. Dog owners should be held legally and financially responsible for stray dogs that place people, particularly young children at risk for body harm.

References:

1- Alizadeh K, Shayesteh A, Xu ML: An Algorithmic Approach to Operative Management of Complex Pediatric Dog Bites: 3-Year Review of a Level I Regional Referral Pediatric Trauma Hospital. Plast Reconstr Surg Glob Open. 5(10):e1431, 2017

2- Abraham JT, Czerwinski M: Pediatric Dog Bite Injuries in Central Texas. J Pediatr Surg. 2018 Oct 31. pii: S0022-3468(18)30672-9. doi:10.1016/j.jpedsurg.2018.09.022.

3- Chen Y, Tan Y, Yan S, Li L: Dog bite and injury awareness and prevention in migrant and left-behind children in China. Sci Rep. 8(1):15959, 2018

4- Pai D, Kamath AT, Panduranga KP, et al: Survey of knowledge of school children towards the prevalence, severity, management of maxillofacial injuries, and rescue skills in the event of a dog bite. J Indian Soc Pedod Prev Dent. 36(4):334-338, 2018

5- Arhant C, Beetz AM, Troxler J: Caregiver Reports of Interactions between Children up to 6 Years and Their Family Dog-Implications for Dog Bite Prevention. Front Vet Sci. 4:130, 2017

6- Bykowski MR, Shakir S, Naran S, et al: Pediatric Dog Bite Prevention: Are We Barking Up the Wrong Tree or Just Not Barking Loud Enough? Pediatr Emerg Care. 2017 Apr 11. doi: 10.1097/PEC.00000000001132.

Gilbert Syndrome

Gilbert syndrome (GS) is considered the most common inherited disorder of bilirubin metabolism due to reduction of uridine diphosphate-glucuronyl transferase 1A1 activity. The hepatic bilirubin glucurodination activity is approximately 50% lower than normal. It is characterized by asymptomatic unconjugated hyperbilirubinemia in the absence of liver disease or hemolysis. GS is mild and benign condition inherited as autosomal recessive that affect being either homozygotes or compound heterozygotes (two different recessive alleles at a particular locus). It may be precipitated by dehydration, fasting, menstruation, overexertion or stress due to intercurrent febrile illness or even vigorous exercise. Other than experiencing jaundice patients are typically asymptomatic. GS usually manifests itself after puberty, affects 3-7% of the population with male predominance. Children may report vague abdominal discomfort, general fatigue and malaise without evident cause. Caloric restriction raises the serum unconjugated bilirubin lever twofold in affected patients with Gilbert syndrome. Fasting, caloric restriction and the intravenous nicotinic acid (niacin) provocative test has been used to diagnosed GS. The oral rifampin test induces cytochrome P-450 isoenzymes and competes for the excretory pathways in the liver at the cellular level causing an exaggerated elevation in unconjugated total serum bilirubin level in GS. Liver biopsy is not indicated and is usually normal. Genetic testing for the A(TA)7TAA variant and Gly71Arg mutation is diagnostic of Gilbert syndrome. GS can present in the newborn period if there is concurrent hemolytic disease such as ABO incompatibility, hereditary spherocytosis or G6PD deficiency. GS has been found to be a precipitating factor for idiopathic cholelithiasis in children. There is no specific treatment except to avoid fasting and severe stress. The prognosis overall is excellent.

References:

1- Keren G, Mazilis A: Gilbert syndrome presenting in a young boy, confirmed by the rifampin test. Isr Med Assoc J. 9(8):626-7, 2007

2- Memon N, Weinberger BI, Hegyi T, Aleksunes LM: Inherited disorders of bilirubin clearance. Pediatr Res. 79(3):378-86, 2016

3- Lee JH, Moon KR: Coexistence of gilbert syndrome and hereditary spherocytosis in a child

presenting with extreme jaundice. Pediatr Gastroenterol Hepatol Nutr. 17(4):266-9, 2014

4- Kitsiou-Tzeli S, Kanavakis E, Tzetis M, Kavazarakis E, Galla A, Tsezou A: Gilbert's syndrome as a predisposing factor for idiopathic cholelithiasis in children. Haematologica. 88(10):1193-4, 2003

5- del Giudice EM, Perrotta S, Nobili B, Specchia C, d'Urzo G, Iolascon A: Coinheritance of Gilbert syndrome increases the risk for developing gallstones in patients with hereditary spherocytosis. Blood. 94(7):2259-62, 1999

6- Karpathios T, Moustaki M, Yiallouros P, Sharifi F, Attilakos A, Papadopoulou A, Fretzayas A: Severe jaundice in two children with Kawasaki disease: a possible association with Gilbert syndrome. J Korean Med Sci. 27(1):101-3, 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 Children's & Women Hospital. Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426. Tel (787)-999-9450 E-mail: *titolugo@coqui.net* Internet: http://home.coqui.net/titolugo

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