PEDIATRIC SURGERY *Update**
Vol. 54 No. 06 JUNE 2020

Thyroglossal Duct Cyst Carcinoma

Thyroglossal duct cyst (TDC) is the second most common neck mass in a child occurring in 7% of the population. It's a benign cystic bump in the middle of the neck near the hyoid bone that moves with tongue protrusion. The diagnosis can be corroborated with ultrasound. Excision of the cyst and duct along with the central portion of the hyoid bone is curative (Sistrunk's procedure). A papillary carcinoma (CA) can arise from a preexisting TDC from the thyro-embryonic follicular thyroid remnant cells. The incidence of papillary carcinoma is 1% in surgically removed TDC. 90% of TDC carcinomas are papillary or follicular in origin, 5% are squamous cell and the rest is anaplastic, Hurthle cell or adenocarcinoma. Median age of diagnosis of TDC carcinoma is 40 years. TDC-CA occurs de novo arising from ectopic thyroid gland tissue. They are not a metastasis from an occult thyroid primary and any lesion found in the thyroid gland represents a multifocal independent primary cancer. Females are more commonly affected. Cervical node metastasis from papillary TDC carcinoma occurs in 10-25% of cases. After finding a papillary carcinoma in the specimen of the excised TDC in a child there exists controversy whether performing a total thyroidectomy or not. Most cases are found incidentally after examining the specimen histologically. Once the diagnosis is established an ultrasound of the neck and thyroid gland should be performed along with FNA biopsy of any suspicious nodule in the thyroid gland or lymph node. Genetic testing for BRAF, N-RAS, and H-RAS should be performed. Low risk patients should be managed with only Sistrunk procedure and include those with less than 45 years of age, small tumors (< 1 cm), classic histology, no extracapsular spread, no vascular invasion, negative margins, no nodal or distant metastasis and a normal thyroid gland and neck by imaging studies. Without these criteria they are categorized as high risk and managed with total thyroidectomy with or without lymph node dissection if they are found to be FNA-positive, and radioiodine ablation therapy. Long-term follow-up is mandatory after primary Sistrunk procedure without total thyroidectomy. Prognosis of TDC carcinoma is excellent with five ad 10-year overall survivals of 100% and 96% respectively.

References:

1- Tahir A, Sankar V, Makura Z: Thyroglossal duct cyst carcinoma in children. J Surg Case Rep. 2015 Apr 15;2015(4).
2- Seow-En I, Loh AH, Lian DW, Nah SA: Thyroglossal duct cyst carcinoma: diagnostic and management considerations in a 15-year-old with a large submental mass. BMJ Case Rep. 2015 Jul 6;2015. pii: bcr2015210923. doi: 10.1136/bcr-2015-210923.

*3- Thompson LDR, Herrera HB, Lau SK: Thyroglossal Duct Cyst Carcinomas: A Clinicopathologic Series of 22 Cases with Staging Recommendations. Head Neck Pathol. 11(2):175-185, 2017
*4- Thompson LDR, Herrera HB, Lau SK: Thyroglossal Duct Cyst Carcinomas in Pediatric Patients: Report of Two Cases with a Comprehensive Literature Review. Head Neck Pathol. 11(4):442-449, 2017
5- Mimery A, Al-Askari M: Occult papillary thyroid carcinoma with cystic nodal metastasis mimicking a thyroglossal duct cyst: A case report. Int J Surg Case Rep. 66:76-79, 2020
6- Boyanov MA, Tabakov DA, Ivanova RS, Vidinov KN: Thyroglossal duct cyst carcinoma. Endokrynol Pol. 2020 Mar 10. doi: 10.5603/EP.a2020.0010.
7- Bahar A, Torabizadeh Z, Movahedi Rad M, Kashi Z: Papillary carcinoma in correlation to thyroidal duct cyst: A case series. Caspian J Intern Med. 11(1):110-115, 2020

Thoracic Outlet Syndrome

Thoracic outlet syndrome (TOS) is the constellation of neurologic and vascular symptoms caused by mechanical compression and entrapment of the subclavian vessels and brachial plexus within a space delimited by the scalene muscle, clavicle and first rib. As these structures pass into the upper extremity, they run through three important spaces: the interscalene triangle, the costoclavicular space and the subpectoral space. Compression can occur in any of these spaces secondary to trauma or a structural malformation. A bone abnormality or soft tissue problem usually plays the etiological role. These could be a cervical rib, abnormal first rib, long transverse process of the 7th cervical vertebra or fracture of the clavicle. Soft tissue pathology associated with TOS includes abnormal fibrous bands and ligaments or congenital/acquired alteration of scalenus anterior muscle. In adults more than 90% of TOS cases are neurogenic in origin, with venous compression comprising 5% and arterial compression in 2%. In children arterio-venous ischemic symptoms predominate in 62% with 38% presenting with neurologic symptoms. It is more commonly seen in females. The neurogenic variety of TOS manifests clinically with pain, weakness, cold intolerance, numbness of the hand and occasional loss of muscle at the base of the thumb. The venous TOS manifest with swelling, pain and bluish discoloration of the arm. The arterial TOS shows pain, coldness and paleness of the arm. Pediatric cases present with neck discomfort, upper limb numbness, weakness and sensory loss. The anatomy of neurogenic TOS is complex and is probably best determine by a combination of plain x-ray, MRI, CT-Scan, duplex scanning, nerve conduction studies and electromyography. TOS remain a diagnosis of exclusion. Differential diagnosis includes cervical disk herniation, distal compression neuropathy, syringomyelia, Pancoast tumor and brachial inflammation. Most pediatric patients are managed conservatively correcting posture, exercises and nerve block rather than with surgical intervention. Scalenectomy is suitable for all TOS patients who did not have bony compression. With cervical ribs, the rib is removed after excision the scalenus anterior, medius and minimus muscle. If the transverse process of the 7th cervical vertebra is longer that the first thoracic vertebra, the former should be removed. Surgical complications include brachial plexus injury, pneumothorax, chylous leakage, lymph effusion and hematomas. Postop rehabilitation is imperative.

References:

1- Vercellio G, Gatti BC, Coletti M, Cipolat L: Thoracic Outlet Syndrome in Paediatrics: Clinical Presentation, Surgical Treatment, and Outcome in a Series of Eight Children. J Pediatr Surg. 38(1): 58-61, 2003

2- Arthur LG, Teich S, Hogan M, Caniano DA, Smead W: Pediatric thoracic outlet syndrome: a disorder with serious vascular complications. J Pediatr Surg 43: 1089-1094, 2008
3- Maru S, Dosluoglu H, Dryjski M, Cherr G, Curl GR, Harris LM: Thoracic outlet syndrome in children and young adults. Eur J Vasc Endovasc Surg. 38(5):560-4, 2009
4- Khan A, Rattihalli RR, Hussain N, Sridhar A: Bilateral thoracic outlet syndrome: An uncommon presentation of a rare condition in children. Ann Indian Acad Neurol. 15(4):323-5, 2012
5- Rehemutula A, Zhang L, Chen L, Chen D, Gu Y: Managing pediatric thoracic outlet syndrome. Ital J Pediatr.27;41:22. doi: 10.1186/s13052-015-0128-4.
6- Chavhan GB, Batmanabane V, Muthusami P, Towbin AJ, Borschel GH: MRI of thoracic outlet syndrome in children. Pediatr Radiol. 47(10):1222-1234, 2017

Torsion Undescended Testis

Undescended testis (UT) is absence of the testis in the scrotum. Occurs in 2% of the male population. It is more commonly found in babies born prematurely. When diagnosed is done after birth the UT is termed congenital. Acquired UT can occur later in life. 80% of UT are palpable within the inguinal canal, and 20% are non-palpable. Palpable UT are managed with orchiopexy before the age of one year. Imaging studies are not sufficiently reliable to determine presence or absence of a non-palpable UT. Non-palpable UT should undergo laparoscopy early in life to determine if the testis is viable and within an intraabdominal position. UT has a higher incidence than normal testes of infertility, cancer development (seminoma), atrophy, trauma and torsion. The most serious complications of UT are a high rate of infertility and high incidence of testicular cancer. Testicular torsion is 10 times more common in UT than normal positioned testis and 10% of all testicular torsion occur in UT. Most cases of UT torsion occur at an average age of 10 months. Abnormal contractions or spasms of the cremasteric muscle and adduction contractures of the hip called scissor-leg deformity that block entrance of the normal scrotum or forces the testes out are theories of why torsion in UT occurs. This is why is seen a higher incidence of UT torsion in children with cerebral palsy. Diagnosis of UT torsion is more difficult. The clinical symptoms of UT torsion include abdominal pain, groin pain, poor oral intake, vomiting and restlessness. Physical exam might include inguinal swelling and redness if the UT was in an inguinal position, with a painful mass in the inguinal region. Rapid diagnosis of UT torsion or any other gonad torsion is critical to preserve fertility. Doppler ultrasound and technetium scrotal scintigraphy scan studies can be diagnostic with the latter being preferred. US could find decrease or absent flow to the affected testis. Testicular scintigraphy with diffuse increased activity without any photopenic area can be the only sign of torsion in an UT. CT shows a well-circumscribed isodense or heterogenous mass and has fine anatomic detail in locating the affected UT with torsion. The treatment of choice for suspected acute UT torsion is immediate surgical exploration. The rate of testicular loss with UT torsion is very high along with the rate of developing an atrophic testis. With symptoms > 24 hours, no flow and no bleeding of the tunica albuginea orchiectomy is performed.

References:

1- Artul S, Artoul F, Fahoum B, Nseir W, Nasrallah N, Habib G: Torsion of undescended third testis, as rare cause of painful inguinal mass. Case Rep Urol. 2015;2015:273508. doi: 10.1155/2015/273508. Epub 2015 Jan 26.

2- Shayegani H, Divband G, Tavakkoli M, Banihasan M, Sadeghi R: Torsion of the undescended testis detected by 99mTc testicular scintigraphy: a case report. Nucl Med Rev Cent East Eur. 2016;19(B):24-25. doi: 10.5603/NMR.2016.0034.
3- Naouar S, Braiek S, El Kamel R: Testicular torsion in undescended testis: A persistent challenge. Asian J Urol. 4(2):111-115, 2017
4- Iio K, Nomura O, Kinumaki A, Aoki Y, Satoh H, Sakakibara H, Hataya H: Testicular Torsion in an Infant with Undescended Testis. J Pediatr. 197:312-312, 2018
5- Kargl S, Haid B: Torsion of an undescended testis - A surgical pediatric emergency. J Pediatr Surg. 55(4):660-664, 2020
6- Shin J, Jeon GW: Comparison of diagnostic and treatment guidelines for undescended testis. Clin Exp Pediatr. 2020 Mar 23. doi: 10.3345/cep.2019.01438.

***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 & Woman Hospital.
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
Tel (787) 340-1868 E-mail: *titolugo@coqui.net*
Internet: http://home.coqui.net/titolugo

**** PSU 1993-2020***
ISSN 1089-7739