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Phlegmasia Cerulea Dolens

Phlegmasia cerulea dolens (PCD) is a severe form of venous thrombosis presenting as an edematous, painful, purplish-blue lower extremity. PCD is characterized by tense swelling of the lower extremity with tenderness of the thigh over the femoral vein, mottling of the limb, and absent

distal pulses. It is more commonly seen in adults than children. It is extremely important to recognize this venous thrombosis as opposed to an arterial insufficiency to avoid gangrene and amputation. Malignant disease is the most common underlying condition found in phlegmasia cerulea dolens (PCD). Unsuccessfully managed iliofemoral venous thrombosis can result in pulmonary embolism, phlegmasia cerulea dolens, and post-thrombotic syndrome. Other predisposing conditions include diabetes, hypercoagulation and previous DVT. PCD can also be seen after percutaneous insertion of a vena caval filter. PCD is evaluated using radionuclide or standard venography. Several treatment methods are used alone or in combination, namely intravenous heparin, thrombolytic therapy and venous thrombectomy. Nongangrenous forms of PCD respond well to systemic anticoagulation. Combination therapy using venous thrombectomy and heparin are indicated for severe ischemia, early venous gangrene, or failure of PCD to improve after six to twelve hours of heparin therapy. Phlegmasia cerulea dolens with venous gangrene is the lethal form of the entity and respond poorly to established therapy.

References:

- 1- Weaver FA, Meacham PW, Adkins RB, Dean RH: Phlegmasia cerulea dolens: therapeutic considerations. South Med J. 81(3):306-12, 1988
- 2- Hood DB, Weaver FA, Modrall JG, Yellin AE: Advances in the treatment of phlegmasia cerulea dolens. Am J Surg. 166(2):206-10, 1993
- 3- Patel KR, Paidas CN: Phlegmasia cerulea dolens: the role of non-operative therapy. Cardiovasc Surg. 1(5):518-23, 1993
- 4- Gutt CN, Oniu T, Wolkener F, Mehrabi A, Mistry S, Buchler MW: Prophylaxis and treatment of deep vein thrombosis in general surgery. Am J Surg. 189(1):14-22, 2005
- 5- Mousa A, Henderson P, Dayal R, Bernheim J, Kent KC, Faries PL: Endoluminal recanalization in a patient with phlegmasia cerulea dolens using a multimodality approach. Vascular. 13(5):313-7, 2005

Diaphragmatic Tumors

Tumors arising from the muscle or elements of the diaphragm are very rare in occurrence. The small published series has shown that the incidence is similar between boys and girls along with left or right involvement. Most primary tumors arising from the diaphragm are malignant, with rhabdomyosarcoma the most commonly encountered followed by sarcomas, yolk sac tumors and extraosseous Ewing sarcoma. Lymphangiomas and

hemangiomas are the most common benign tumors found in the diaphragm. The clinical presentation in children varies, with predominantly chest symptoms (chest pain, shortness of breath, cough, chest asymmetry or hemothorax). Identifying the site of origin of the tumor to the diaphragm is difficult even after using CT, MRI and ultrasound. Exploratory laparotomy with biopsy is the best tool to assign location to the tumor. Management of primary diaphragmatic tumors encompasses wide local resection with reconstruction, chemotherapy and radiotherapy. To obtain cure, a tumor free resection margin must be obtain initially or after chemotherapy shrinkage of the tumor. Reconstruction of the diaphragm at the time of resection can be accomplished with a muscle flap or prosthetic graft (PTFE or Goretex).

References:

- 1- Raney RB, Anderson JR, Andrassy RJ, Crist WM, Donaldson SS, Maurer HM; Intergroup Rhabdomyosarcoma Study Group: Soft-tissue sarcomas of the diaphragm: a report from the Intergroup Rhabdomyosarcoma Study Group from 1972 to 1997. J Pediatr Hematol Oncol. 22(6):510-4, 2000
- 2- Hagr A, Laberge JM, Nguyen LT, Emil S, Bernard C, Patenaude Y: Laparoscopic excision of subdiaphragmatic epidermoid cyst: a case report. J Pediatr Surg. Jun;36(6):E8, 2001
- 3- Akinci D, Akhan O, Ozmen M, Ozkan OS, Karcaaltincaba M: Diaphragmatic mesothelial cysts in children: radiologic findings and percutaneous ethanol sclerotherapy. AJR Am J Roentgenol. 185(4):873-7, 2005
- 4- Belaabidia B, Sellami S, Benelkhayat R, Elattar H, Elidrissi Dafali A: Leiomyosarcoma of the diaphragm: a case report and review of the literature. Cancer Radiother. 10(3):137-41, 2006
- 5- Cheon JS, You YK, Kim JG, Lee DH, Park K, Ahn CJ: Diaphragmatic lipoma in a 4-year-old girl: a case report. J Pediatr Surg. 41(1):e37-9, 2006
- 6- Cada M, Gerstle JT, Traubici J, Ngan BY, Capra ML: Approach to diagnosis and treatment of pediatric primary tumors of the diaphragm. J Pediatr Surg 41(10): 1722-1726, 2006

Breast Cysts

Breast cysts are very common in the adolescent female, while not so common in males. Rapid cyst growth causes pain and a palpable mass in the breast that brings the child to seek medical help. Other times breast cysts get infested and are managed as a breast abscess with antibiotics and drainage. In males solitary, large male breast cysts are extremely rare. They occur mainly in children up to the age of seven years and should be removed under surgery. Whenever a breast mass develops the next step in diagnosis is an ultrasound-mammography studies. This will corroborate the cystic or solid nature of the cyst along with its size. Radiation exposure in children using mammography is not necessary or warrant. Management of breast cysts in females consist of observation or aspiration of the cyst with cytological exam. The overwhelming majority of breast cyst in children are benign and will go spontaneously with time. Follow-up breast ultrasound of the child for persistent of the mass or bloodstained aspirate should be done.

References:

- 1- Jones BM, Bradbeer JW: The presentation and progress of macroscopic breast cysts. Br J Surg. 67(9):669-71, 1980
- 2- Hamed H, Coady A, Chaudary MA, Fentiman IS: Follow-up of patients with aspirated breast cysts is necessary. Arch Surg. 124(2):253-5, 1989
- 3- Devitt JE, To T, Miller AB: Risk of breast cancer in women with breast cysts. CMAJ. 147(1):45-9, 1992

- 4- Hindle WH, Arias RD, Florentine B, Whang J: Lack of utility in clinical practice of cytologic examination of nonbloody cyst fluid from palpable breast cysts. Am J Obstet Gynecol. 182(6):1300-5, 2000
- 5- Bock K, Duda VF, Hadji P, Ramaswamy A, Schulz-Wendtland R, Klose KJ, Wagner U: Pathologic breast conditions in childhood and adolescence: evaluation by sonographic diagnosis. J Ultrasound Med. 24(10):1347-54, 2005
- 6- Vargas HI, Vargas MP, Eldrageely K, Gonzalez KD, Burla ML, Venegas R, Khalkhali I: Outcomes of surgical and sonographic assessment of breast masses in women younger than 30. Am Surg. 71(9):716-9, 2005

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