Case report:

Cervical Cystic Lymphangioma, Rarely Detected in Adults with Rapid Progression, 2 Case Reports

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Abstract:

Cystic Lymphangioma (CL) is rarely detected in adulthood. We presented two cases with neck swelling at an advanced age. Both of cysts were located in the cervico-mediastinally and showed rapid progression. The cysts were excised successfully and there was no recurrence in the follow-up period. Although there are many alternatives in treatment, CL, which is rare in adults, can be treated with surgical excision successfully in appropriate cases to definitively prevent recurrence and complications.

Introduction:

Cystic Lymphangioma (CL), also known as cystic hygroma, is defined congenital cystic lymphatic malformations which most commonly detected in the cervicofacial, rarely axillary and cervical mediastinal locations in infants. CL consists of cystic masses of abnormal lymphatic ducts and comprises in one in 2000-4000 live births. CL’s are characterized by single or multiple cysts within soft tissue. Cysts can be of different sizes and shapes. They are rarely detected in adulthood. Surgery is needful in the treatment of CL1-3. We report two cases of rapid progression to reveal the need to consider CL in differential diagnosis of cervical region cystic masses in adulthood.

Case 1:

A 33-year-old female patient presented with swelling on her left neck that appeared 9 months ago. On physical examination, the mass was about 5 x 6 cm non-tender, soft and fluctuant with well defined rounded borders. Cervical and chest-CT images showed a pure cystic lesion with an approximate diameter of 5 x 6 cm located in the posterior cervical triangle extending to the anterior mediastinum. Total surgical excision was performed, during the procedure, clear straw-colored cyst fluid was aspirated (Figure 1). Histopathological examination revealed that the lesion was compatible with CL. No recurrence was encountered in the 2-year follow-up.

Case 2:

A 32-year-old female patient presented with swelling on her left neck that appeared 6 months ago, showed gradually progression and sudden increase in size in the last month. On physical examination, the mass was about 6 x 7 cm non-tender, soft and fluctuant with well defined rounded borders. Cervical and chest-CT images showed a pure cystic lesion with an approximate diameter of 8 x 6 cm located in the posterior cervical triangle, extending to the thoracic outlet and causing expansion in the surrounding vascular structures. Surgical excision was performed for both diagnosis and treatment. During the procedure, purulant grey-colored cyst fluid was aspirated (Figure 2). Histopathological examination revealed that the lesion was compatible with CH (cystic lymphangioma). No recurrence was encountered in the 1 years follow-up.

Discussion:

Cystic lymphangiomas (CLs) are benign cystic tumors of lymphatic system comprising lucid or yellow colored fluid. They are form of local

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proliferations of well differentiated lymphatic tissue, complex lymph channels and cystic spaces. CL can be caused by genetic or environmental factors such as maternal virus infection, alcohol use during pregnancy, which causes abnormal development of the lymphatic vascular system during embryonic growth. The malformation may develop on its own or may be associated with some genetic syndromes such as Turner syndrome, Down syndrome or Noonan syndrome. CL is related to CL gene mutation in the PIK3CA gene. This is a non-inherited mutation isolated to the lymphatic endothelial cells lining the fluid filled channels. These factors can be counted among the reasons for the frequent occurrence of the disease in childhood. Also infection or chronic trauma to lymphatic cell proliferation can lead to CL in adulthood. Although in many cases, the cause is not clarify3,4.

CLs classified histologically as capillary, cavernous cystic and non-lymphatic lymphangioma. The most common one is cystic. They are most common in the neck and axilla. Cervical ones may have relations with the mediastinum5-6. Both of our two cases presented with neck swelling at an advanced age and both of cysts were located in the cervico-mediastinal region. Cystic subtype was detected in our two cases. CL is mostly asymptomatic due to their soft tissue. However, in some cases, symptoms such as cough, dyspnea, and chest pain may occur due to compression of the cervico-mediastinal structures. Also they can lead to complications such as infection, airway compression, cyhlothorax or chylopericardium5-7. Our cases were also asymptomatic, and they presented with a sign of increasing swelling in the neck area.

Radiological images reveals a lobulated properly bounded mass between the mediastinal structures, sometimes displacing with them. They usually have fluid and soft tissue attenuation and they may demonstrate thin septations with in the cyst. The lesions can get at large sizes rapidly within days or weeks. On MRI (Magnetic Resonance Imaging) they may have heterogenous T1 and high T2 signal intensity8,9. Our cases had radiological findings similar to the literature in direct graphy and tomography. The MRI was not used.

The main therapeutic options for treating a CL are observation, percutaneous drainage, surgery, sclerotherapy, laser therapy, radiofrequency ablation, immunootherapy or medical therapy. The type of treatment should be determined according to the patient’s age, comorbidities, location and size of the lesion. Spontaneous resolution of the cyst is uncommon. Since recurrence is expected after only percutaneous drainage, surgery or sclerotherapy should be added. Although surgical resection is the definitive treatment option, treatments with sclerotherapy after percutaneous drainage have been reported in recent years. Sclerotherapeutic substances are picibanil (OK-432), ethanol, doxycycline, tetracycline, and bleomycin. More successful results are reported with bleomycin and picibanil (OK-432). Medical therapy with the drugs sirolimus or sildenafil can be used to treat CL. This drugs allow lymphatic vessel walls to relax in connection with decreased fluid collection. The drugs are effective by inhibiting cell growth and targeting the PI3K/mTOR pathway. Targeted therapies may be developed in the future to inhibit PIK3CA gene variant activity in CL.4,8-12

Most used and most successful treatment for CL is surgically excision. The aim of surgery to remove all components of CL. The goal of surgical excision should be to eliminate the compression effect and prevent complications. The location of a CL, such as near to a vital organ, may lead only partial removal of the cyst. But, heaps of the times incomplete surgery may be sufficient to prevent complications despite the high recurrence rate4,10-13. We successfully performed total surgical excision in two cases with cervical localization.

Conclusion:

One of the rare causes of space-occupying lesions on the neck-mediastinum in adults is CL. Although there are many alternatives in treatment, surgical excision can be used successfully to definitively prevent recurrence and complications. Etiological causes and targeted therapies for CL in adults are the subject of future research.
References:


