Cystic Hygroma of neck in an adult female: Case report and literature review

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Citation

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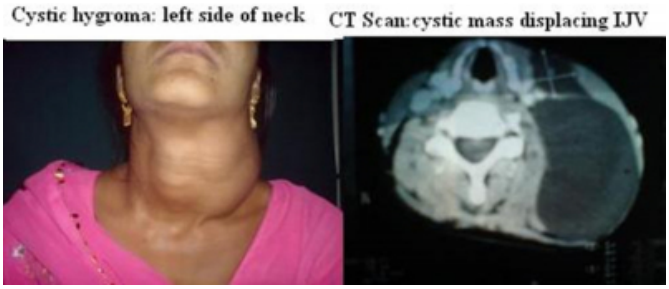
Abstract

Although cystic hygroma is well recognized in pediatric practice, it seldom presents de novo in adulthood. Cystic hygroma is very rare in adults but it should be considered in the differential diagnosis of neck swelling. Patients presenting with a painless, soft, enlarging neck mass should have a careful history and physical examination along with the appropriate radiological imaging to assist with diagnosis. Surgical intervention is the treatment of choice for this rare condition. Very few cases of cystic hygroma have been reported in adults. Here we are reporting a case of cystic hygroma in a 35-year-old female patient who was treated for tubercular cold abscess in the neck for a long duration.

CASE REPORT

A 35-year-old female patient presented in the Surgery Department of Gandhi Medical College, Bhopal, with the complaint of a painless, progressively enlarging mass in the neck which was noticed by a family member. The mass was initially of the size of a lemon situated in the root of the neck on the left side. Over a period of three years, the swelling gradually enlarged to occupy the whole neck on the left side. Her only complaints were fullness on left side of the neck and some discomfort on moving her neck on left side. There was no history of trauma in the neck region, recent or past upper respiratory infections, or any previous history of neck mass as an adult or child. She did not have any complaints of dysphagia, dyspnoea, hoarseness of voice, low-grade fever or loss of weight. She underwent FNAC at a district hospital, which revealed straw-coloured fluid and cytology showed lymphocytes suggesting chronic inflammatory exudate. The patient was put on anti-tubercular treatment for six months. She did not have any relief and the swelling gradually increased in size to occupy the whole left side of the neck. Her past medical and family history was unremarkable.

Figure 1



On examination, a soft, fluctuant, non-tender, transilluminant, well-defined, dumbbell-shaped mass measuring approximately 10x10cm could be palpated on the left side of the neck extending from the submandibular region to the supraclavicular fossa in the vertical plane, and from the paratracheal region to the posterior triangle of the neck in the horizontal plane. There was no thrill, bruit or cervical lymphadenopathy and the remaining neck and systemic examination was normal.

Ultrasonographic evaluation revealed an irregular, multilocular, cystic anechoic lesion in the posteriolateral region of the neck, anterior to major neck vessels. CECT scan of the neck and thorax showed a large loculated cystic mass lesion of near-water density with few enhancing septa within. It occupied the posterior cervical space on the left side of the neck lateral to the carotid artery. It displaced the left internal jugular vein anteriomedially and the sternocleidomastoid muscle anteriolaterally. In its cranio-caudal extent, it extended from C2 to the level of vertebra

C7. A preliminary diagnosis of cystic hygroma was made and the patient was scheduled for surgical excision.

Under general anaesthesia, through a hockey-stick incision, subplatysmal flaps were raised and the sternocleidomastoid muscle was divided to expose the multiloculated cystic lesion throughout its extent. The spinal accessory nerve was splayed anteriorly all across over the cystic mass. The spinal accessory nerve lying on the anterior aspect of the lesion and the internal jugular vein were carefully preserved, and the cystic lesion was excised.

Histopathology revealed thin connective-tissue stroma separating the cystically dilated spaces lined by a single layer of benign endothelial cells which was consistent with cystic hygroma. The postoperative course was uneventful. The patient has been symptom-free and there is no recurrence after one year.

Discussion and literature review: Cystic hygroma (lymphangioma) is a benign congenital malformation of the lymphatic system that occurs as a result of sequestration or obstruction of developing lymph vessels. Lymphangiomas usually present in infancy or early childhood. Approximately 50 to 60 percent of cystic hygromas appear before the end of the first year of life. Almost 80 to 90% of cystic hygromas present before the end of the second year of life. Cystic hygroma is an uncommon differential diagnosis of a progressively enlarging neck mass in adulthood.

In lesions diagnosed pre-natally, before 30 weeks, chromosomal abnormalities are common. When diagnosed in the prenatal period, this disease may be associated with Turner syndrome, Noonan syndrome, cardiac anomalies, trisomy syndromes and fetal hydrops. [2, 4, 7]

Approximately 75-80% of cystic hygromas appear in the neck and the lower portion of the face. Other sites are axilla [20%], mediastinum [5%], abdomen [liver, spleen and colon], pelvis, retroperitoneum [kidney], groin, scrotum and skeleton. In adults, cystic hygromas are commonly seen in the sublingual, submandibular and parotid spaces. [3]

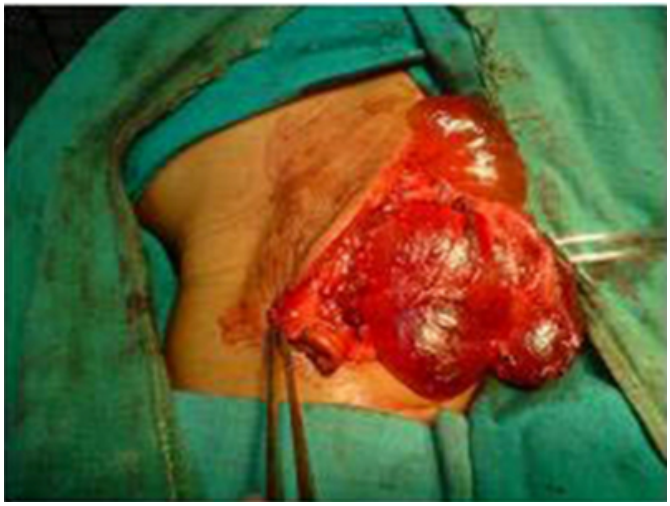
Grossly, cystic hygromas are multiloculated masses composed of many cysts. Microscopically, the cyst wall consists of a single layer of flattened epithelium and the spaces may or may not have blood-containing capillaries suggesting that there may be a combined vascular and lymphatic defect. These poorly supported blood vessels in cystic hygromas may bleed and produce rapid enlargement and discoloration of the hygroma.

Infection within the cysts (usually caused by streptococcus or staphylococcus species) may occur. This complication can also cause rapid enlargement which may result in airway obstruction. Malignant transformation has not been reported in cystic hygromas, and there have been no cases of spontaneous regression.

Three types of lymphatic malformations are described: capillary, cavernous and cystic. A classification based on CT, anatomical location, and histology was proposed by McGill and Mulliken [8] in 1993 and is clinically relevant. Type I malformations or classic cystic hygroma are macrocystic and develop below the mylohyoid muscle. They involve the anterior and posterior triangles of the neck. Type II malformations are microcystic and invasive and are found in the neck above the level of the mylohyoid. They usually involve the lip, tongue, and oral cavity and are difficult to resect.

Pre-operative imaging is essential to look for intrathoracic extension, which is present in 10% of cases. Ultrasonography or magnetic resonance imaging (MRI) is recommended. Lymphangiomas are best visualized by magnetic resonance imaging (MRI). The high water content allows lymphangiomas to appear hyperintense on T2-weighted images.

Figure 2



Doppler studies are done to evaluate the vascularity of the mass and see the relation with adjacent vessels. On CT scan, cystic hygromas appear as non-enhancing thin-walled multiloculated masses with near-water attenuation values [3].

The differential diagnosis of a neck mass in adults may

include lipoma, branchial cleft cyst, haemangioma, cold abscess, lymphoma, hamartoma, thyroid mass, thyroglossal duct cyst, dermoid cyst, metastatic disease, or other tumors.

The preferred treatment for cystic hygroma is surgical excision because of the dangers of infection and fistula formation. Sometimes, this may be impossible due to the infiltrating nature of the hygroma within and around neurovascular structures, muscles and blood vessels. In this condition, unroofing, partial cystectomy and drainage of the cystic content should be performed and all adjacent crucial structures should be preserved. In these situations, a recurrence rate of 10-15 % is reported [1]. Complications following resection are postoperative muscle weakness, nerve injuries and neural weakness, bleeding and wound infection. In our patient the spinal accessory nerve was splayed anteriorly all across over the cystic mass. On the medial aspect, the mass could be carefully separated from the internal jugular vein, carotid vessels, vagus nerve and hypoglossal nerve.

Laparoscopic techniques have been used in both diagnosis and treatment of retroperitoneal cystic hygromas. VATS has been performed in some cases for mediastinal cystic hygromas. Laser therapy for oropharyngeal extension has been used with good effect.

Percutaneous aspiration is not preferred because of the risk of bleeding, infection and recurrence. Injection of sclerosing agents like alcohol, bleomycin [5] and OK-432 (a lyophilized mixture of Streptococcus pyogenes and Penicillin G potassium) [6] have been reported with favorable results.

CONCLUSION

Cystic hygromas are a rare differential diagnosis in adult neck masses and should be remembered for the adult patients who have neck, axillary, mediastinal, groin, or retroperitoneal masses. Preoperative imaging for diagnosis and to look for intrathoracic extension is essential. Surgical excision is the treatment of choice.

References

1. Ravitch MM, Rush BF Jr. Cystic hygroma. In: Ashcraft KW and Holder TM, Editors, Paediatric Surgery, Saunders, Philadelphia, PA 1993.
2. Chappius IIP. Current aspects of cystic lymphangioma in the neck. Arch Pediatr 1994: 1(2); 186-192.
3. Mansingani S, Desai N, Pancholi N. A case of axillary cystic hygroma. Indian J Radiol Imag 2005; 15(4): 517-519.
4. Ali Güner, Akif Aydin, Faik Çelik. Cystic hygromas in adults: Reports of two cases.Bakirköy Tip Dergisi 2006; 2: 101-3.
5. Orford J, Barker A, Thonell S, King P, Murphy J. Bleomycin therapy for cystic hygroma. J Pediatr Surg. 1995; 30(9): 1282-7.
6. Smith RJ, Burke DK, Sato Y, Poust RI, Kimura K, Bauman NM. OK-432 therapy for lymphangiomas. Arch Otolaryngol Head Neck Surg 1996; 122(11): 1195-1199.
7. Morley SE, Ramesar KC, MacLeod DA. Cystic hygroma in an adult: a case report. J R Coll Surg Edinb. 1999; 44(1): 57-8.
8. Oakes MJ, Sherman BE. Cystic Hygroma in a Tactical Aviator: A Case Report. Mil Med 2004; 169(12): 985-7.

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