

Cystic Hygroma in Adults: The Forgotten Entity

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ABSTRACT

Cystic hygroma also known as lymphangioma are rare benign congenital malformation of the lymphatic system. With advancement of medical facilities and awareness about medical problems congenital defects present usually after birth or in childhood, however still some cases are missed and present late. Cystic hygroma presenting in adults is quite rare with just around 100 cases of adult lymphangioma been reported in the literature. In this case report we present our experience of managing an adult patient and have also briefly reviewed the literature.

Our patient was a young female with huge cystic swelling in right side of neck and upper chest wall with no intra thoracic extension. Our patient underwent complete excision and has no recurrence till yet. Though surgery forms the cornerstone of therapy, at times it is not possible or is dangerous to attempt surgery. So in these situations newer modalities like sclerosant injection, CO₂ laser therapy or interferon alpha therapy have a role to play. Total excision is the gold standard and wherever

possible should be attempted, however in inaccessible areas newer modalities can be tried.

Keywords: Cystic Hygroma in Adult, Lymphangioma.

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INTRODUCTION

Cystic hygroma also known as lymphangioma are rare benign congenital malformation of the lymphatic system that occurs as a result of development arrest of embryological connection between lymphoid vessels and venous system.¹ These lesions are usually discovered in infants or children less than two years of age, with head and neck being the favoured sites.² Other common sites of occurrence include trunk, axilla, and extremities. In the head, the oral cavity and face area are the most common sites. In the neck, the posterior triangle is the most common site. Cystic hygromas have also been reported from unusual sites like Larynx, retroperitoneum, adrenal, pancreas, liver and mesentery.²⁻⁷ The site of the lesion determines the symptomology, however most of the patients are asymptomatic other than a soft tumor mass. Oral cavity and laryngeal involvement can produce stridor, dyspnea, and feeding difficulties. Cystic hygroma presenting in adults is quite rare with just around 100 cases of adult lymphangioma been reported in the literature.⁸ Surgical excision has been the gold standard with acceptable recurrence rates, however it can be challenging if it has intrathoracic extension or situated in unusual sites like oral cavity or retroperitoneum. Few non-invasive modalities for management of cystic hygroma like laser photocoagulation, OK -432 injections, interferon alfa therapy have

also come up and are especially handy in management of cystic hygromas at surgically difficult sites.

CASE DESCRIPTION

A 23 year-old female was referred to our clinic with a 5 year history of gradually enlarging mass on the right side of her neck. Initially it was asymptomatic, however she had started having discomfort and mild pain while turning her head for the past one year. Physical examination revealed a cystic, smooth and mobile 10* 15 cm mass in the right supraclavicular fossa and anterior triangle of neck and extending down to lower border of clavicle (figure 1). The swelling was fluctuation positive and brilliantly transilluminant (figure 2). Fine needle aspiration biopsy (FNAB) showed yellow colored, liquid with mature lymphocytes. Computed tomography of the neck revealed that 12x15x9 (CC*lateral*AP) cm lobulated cystic mass extending from level of hyoid bone to inferior border of clavicle and laterally to medial two thirds of clavicle (figure 3). Excision of the mass was performed under general anesthesia and it was removed entirely (figure 4). Pathologic diagnosis was cystic lymphangioma which was supported with a thick fibrous wall. The patient has recovered well and has no recurrence.



Fig 1: Cystic lobulated swelling in anterior part of neck



Fig 2: Transillumination positive swelling

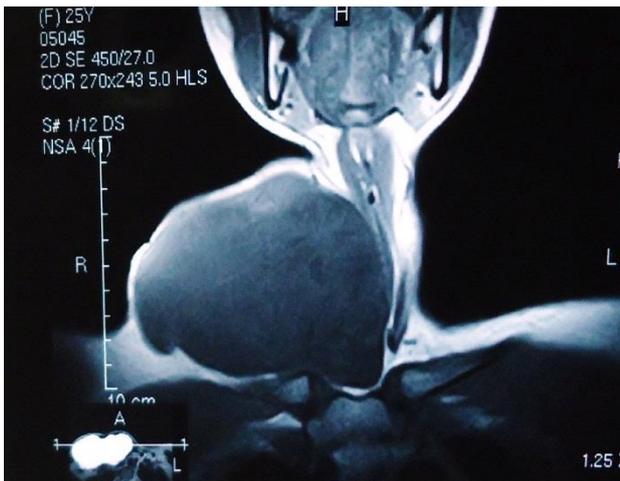


Fig 3: CT (computed tomography) image of non-enhancing cystic lesion in anterior part of neck

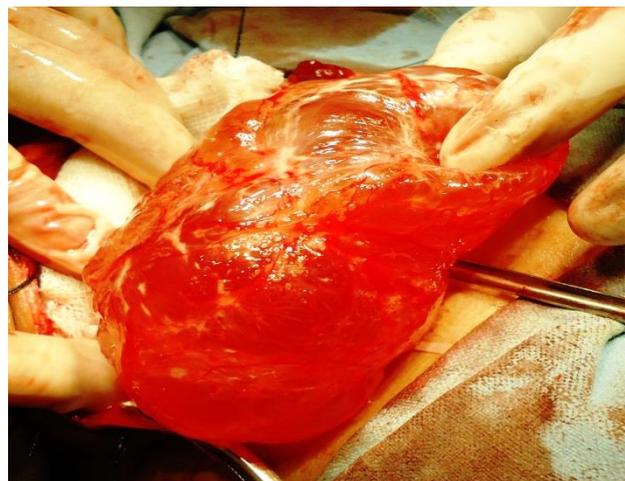


Fig 4: Intra operative image of swelling being excised completely

DISCUSSION

Cystic hygromas also known as lymphangiomas are thought to be developmental abnormalities associated with a failure of embryological connection between lymphoid vessels and venous system and generally not accepted as true tumors.⁹ However, there is a case report which suggests trauma could trigger formation of lymphangioma.¹⁰

There are three histological subtypes: Capillary lymphangioma (composed of small lymphatics), cavernous lymphangioma (composed of larger lymphatics), cystic lymphangioma (cystic hygroma- composed of large macroscopic lymphatic spaces with collagen and smooth muscle). Cavernous lymphangioma is the most common subtype.¹¹ The incidence of cystic hygroma is approximately 1/6000 live births.¹² 70–80% of cystic hygromas occur in the neck, usually in the posterior cervical triangle.¹³ The remainder 20–30% occurs in the axilla, superior mediastinum, chest wall, mesentery, retro-peritoneal region, pelvis and lower limbs.¹⁴ Cystic hygromas presenting in adulthood is rare and only few cases have been reported as usually these swellings present in infancy or in children less than two years of age. Most of these swellings are asymptomatic, however at times they present with compressive symptoms.

Lymphangiomas are best visualized by magnetic resonance imaging (MRI); the high water content allows lymphangiomas to appear hyperintense on T2-weighted images.¹⁵ The other imaging methods are Doppler ultrasonography and computed tomography

(CT). Cystic hygromas can be associated with chromosomal malformations like Turner syndrome, Noonan syndrome and trisomy syndromes.^{16,17} Usually cystic hygromas are slow growing tumors except when the cysts get infected or when there is bleeding inside the cyst. They are not known to resolve spontaneously.

Complete surgical excision is still the gold standard of management of cystic hygroma. However, this may not be possible sometimes due to the infiltrating nature of the hygroma within and around neurovascular structures. In this condition, unroofing, partial cystectomy and drainage of the cystic content can be performed, which has a recurrence rate of 10-15 %.¹⁸

Injection of sclerosing agents like alcohol, bleomycin and OK-432 (a lyophilized mixture of streptococcus pyogenes and penicillin G potassium), with favorable results have been reported.¹⁹ OK 432 is one of the most commonly used sclerosing agents. It is produced by group A *Streptococcus pyogenes*. It produces an inflammatory reaction when applied with intracystic injection. Although success rate is variable, there are promising results. Fever and local inflammatory reaction are the common side effects of OK 432.²⁰ But hypopharyngeal edema was reported especially on administration for cysts located near the airway.²¹

CO₂ laser photocoagulation has been successfully employed in managing superficial lymphangioma of oral cavity.²² Laser therapy reduces cyst size but it has significant risk of damage to the overlying skin.^{23,24} Interferon alpha has also been used in

management of lymphangioma and haemangioma because of its antiangiogenic effect. The most common side effects of interferon alpha are fever, neutropenia, and diarrhea.²⁵ Percutaneous aspiration is not preferred modality of management because of the risk of bleeding, infection and recurrence, however it can be attempted to buy time till proper surgical excision can be done.²⁶ These tumors are insensitive to radiotherapy.²⁷

CONCLUSION

Cystic hygromas are benign developmental anomalies which usually presents in young children and most of them are asymptomatic. It most commonly occurs in head and neck region, although it can occur at other places. Surgical removal is the procedure of choice, which is at times not possible due to proximity to vital neurovascular structures. In these situation new modalities can be tried like intralesional sclerosant injection, interferon alfa therapy and laser photocoagulation have a role to play.

CLINICAL SIGNIFICANCE

Cystic hygroma presenting in adulthood is rare in these days of Medical advancement. It is most commonly present in head and neck region, though it has been reported in retroperitoneum, mesentery and others sites. Total excision is the gold standard and wherever possible should be attempted, however in inaccessible areas newer modalities can be tried.

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