Recurrent cystic hygroma in an adult presenting as a neck mass - A case study

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Abstract

Cystic Hygroma although common in children, can also present in adults. It should be managed with great precision because of its proximity to vital anatomy. This case report discusses a rare presentation of a 52 years old adult presenting with cystic hygroma. It illustrates how difficult can cystic hygroma get, especially during dissection. In this case, careful delineation of structures was done to excise the mass without injury to surrounding nerves and vessels. Cystic hygromas do not resolve spontaneously and surgical excision is the treatment of choice in such cases.

Keywords: Cystic Hygroma, Adult, Posterior Triangle.

Introduction

Cystic Hygroma, also known as lymphangioma is a congenital condition of the lymphatic system. This benign malformation is generally seen in infants or children. It occurs due to sequestration or obstruction of lymphatic vessels which are in the developing stage. They were originally reported by Redenbacher in 1828 and "Cystic Hygroma", name was first given by Wernher in 1834 [1]. The embryologic development of lymphangiomas is controversial. Two predominant theories regarding the pathogenesis is either a congenital blockage of lymphatics or a true neoplasm [1, 2]. On histology, lymphangiomas are of three types- lymphangioma simplex, cavernous lymphangioma and cystic hygroma, depending on the size of vascular spaces and thickness of the adventitia.

They are rare paediatric tumors (6% of overall benign tumors and 5% of vascular tumors) and even rarely reported in adults in the literature[4]. They can involve various parts of the body including the retroperitoneum, mesentery, groin, extremities, chest wall, mediastinum and viscera [3]. Head and neck region accounts for 40% to 70% of all lesions. Despite their benign nature, surgical management is difficult, especially for the cavernous lymphangioma, because of its tendency to spread along vital structures and the subsequent high incidence of recurrence (7-40%), occurring within six months to one year postoperatively.

We present a case of a 52 years old male who presented with a left cervical mass.

Case Report

A 52-year-old male was referred to our clinic with a three year history of a slowly enlarging soft mass on the left side of his neck. He complained of mild discomfort while turning his head to the left side. Physical examination revealed a boggy mass which was mobile, measuring 09-12 cm in the left posterior triangle region of the neck (Figure 1). Fine needle aspiration cytology showed a benign cystic lesion. CT scan of the neck revealed a 07 x 08 x 06 cm lobulated cystic mass extending from the hyoid bone to left supraclavicular fossa and obliterated jugular trunk and posterior cervical triangle (Figure 2).

The mass was excised under general anesthesia. The mass was extending from mastoid tip to the upper border of clavicle lying posterior to sternoclavomastoid (SCM) and extending medially up to lateral border of thyroid cartilage and laterally up to medial border of trapezius (occupying the whole of posterior triangle. The carotid pulsations were felt separately. The modified Schobinger’s incision was given for neck dissection. Subplatysmal flaps were elevated and wide exposure was achieved. Greater Auricular nerve, External Jugular vein and SCM were delineated. A large cystic and multiloculated swelling was seen jutting out posterior to SCM and occupying mainly posterior triangle. Erb’s point was identified and Spinal Accessory nerve was dissected up to its insertion in trapezius. Dissection of mass was started medially to
The mass was adhered to the left Internal Jugular vein (IJV) sheath, at the level of the upper border of thyroid cartilage to bend of cricoid level below. IJV, Common Carotid Artery and Vagus were identified, UV was exposed in its entire cervical course and its superior and inferior ends were controlled and careful dissection was done by opening carotid sheath and separating the mass from UV sheath (Figure 3b). Post belly of digastric muscle and submandibular gland were identified and dissection proceeded in posterior triangle. SCM was retracted medially. The cyst was separated from ascending branch of transverse cervical artery. Then the cyst was separated along with its lymphatics from jugular trunk area inferiorly where the cystic duct joins right IJV bulb. The cyst was dissected from underlying scalene muscles and levator scapulae postero-superiorly. The multiloculated cyst was taken out in toto with a part of SCM fibers to which it was adherent in its posterior aspect (Figure 4). Wound was closed in layers. A drain was placed and was taken out after 48 hrs. The recovery was uneventful.

Histopathological examination revealed a cystic lesion made up of dilated lymphatic spaces. These spaces were lined by endothelial cells. Scant intervening connective tissue showing few lymphoid aggregates could be seen separating these spaces. A diagnosis of Cystic Hygroma was given.

Discussion

Cystic lymphangioma occurs approximately in 1 in 12000 births, with 95% occurring by the second year of life. This pathology has been linked with Turner syndrome, cardiac anomalies, trisomy syndromes, Noonan syndrome and fetal hydrops. Aneesh Kumar et al have suspected trauma to be a triggering cause [4]. Adult patients are usually asymptomatic and present as a soft, painless, mobile, transilluminant cystic mass on physical examination. Magnetic resonance imaging (MRI) is the imaging modality of choice. On T2-weighted images, these lesions appear hyper intense due to the high fluid content [5]. Doppler ultrasonography and computed tomography (CT) are other imaging modalities used.

Differential diagnoses comprise of hemangioma, mucocele and meningomyelocele [6]. Complications include bleeding into the cyst. Beta hemolytic streptococcus or staphylococcus aureus may cause infection within the cysts. This complication can lead to rapid enlargement resulting in airway obstruction.

These malformations do not resolve on their own accord. Percutaneous aspiration is generally not done because of the risk of haemorrhage, infection and recurrence. These lesions are not responsive to radiotherapy. Positive results have been seen in some cases with injection of sclerosing agents namely bleomycin, alcohol and OK-432 [7].

The treatment of choice is complete surgical excision under general or local anesthesia. Excision might not be possible in some cases if the hygroma infiltrates within and around neurovascular structures and muscles. Unroofing, partial cystectomy and drainage of the contents can be done in such situations. However, recurrence rate of 10-15 % has been seen with these methods [8].
Oldest case in literature of cystic hygroma is eighty nine years old [9]. Cystic hygroma is an uncommon cause of mass in cervical and axillary region in case of adults. Although this condition can be treated with different methods, surgical removal is the treatment of choice. Even though this malformation is more commonly seen in children, it should also be considered in the adults presenting with cervical or axillary masses.

Conclusion

To conclude, cystic hygroma should be considered as a differential diagnosis for lateral neck mass, even in adults as seen in our case. Imaging helps in diagnosis of this malformation. As these lesions do not resolve on their own, surgical excision is the treatment of choice. However, very careful dissection is essential to avoid injury to surrounding neurovascular structures.

Conflict of Interest

The authors have none to declare. Permission from the concerned patient has been taken for publication of photos and related data. No financial implication.

References