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Case Report

Surgery Section

Cystic Hygroma in Adulthood - A Case Report

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ABSTRACT

Cystic Hygroma or Cystic Lymphagioma is a rare tumour of lymphatic origin, which is considered to be a congenital malformation of the lymphatic vessels due to failure of blind clusters of lymph sacs to join the lymphatic system during development. 90% of cases are diagnosed by age of 2 years. Very rarely for unknown reasons it can present in adults (mainly in cervicofacial region). Complete excision is the treatment of choice but total removal is difficult in all cases due to its extent and involvement of vital structures. Recurrence rate of 20% is reported even after meticulous care to avoid leaving behind island of tissue which acts as foci of recurrence. A 25 year old female patient presentedto our hospital with progressive painless swelling in the left side of neck associated with discomfort while moving her neck to left side. After radiological imaging and other investigations, excision of cystic swelling was done under GA without any damage to vital structures. Post operative period was uneventful and patient was discharged 3 days after surgery.

Keywords: Lymphangioma, Recurrence, Transilluminant

CASE REPORT

A 25 year-old female complaint of a swelling in left side of neck for the past 2 years, which is progressive and painless. She also complaints of discomfort on moving her neck to the left side. No history of dysphagia / dyspnea / hoarseness of voice / low-grade fever / loss of weight. No past history of trauma neck mass.

On examination 9x10cm Swelling involving the left side of neck [Table/Fig-1], was found smelling was non-tender, cystic in consistency, with smooth surface, margins well defined, fluctuant and transillumination test was positive, (non-pulsatile, non-compressible). The trachea was central in location and carotid was palpable at normal position. No cervical lymphadenopathy.

Ultrasonography of neck showed cystic lesion with septations containing clear fluid without thoracic extension.

Under general anaesthesia, through a transverse skin incision, subplatysmal flaps were raised and the sternocleidomastoid muscle was retracted to expose the multiloculated cystic lesion throughout its extent. The Cyst got ruptured while dissecting till the margins. Complete excision of cyst was done without injuring important nerves and vessels [Table/Fig-2a,b]. The postoperative course was uneventful and patient was discharged on post operative day 3 and followed up for 3 months Gross specimen contains multiloculated masses composed of many cysts [Table/Fig-3].

Histopathology revealed connective tissue stroma filled with lymphoid cells separating the cystically dilated spaces [Table/Fig-4a,b].

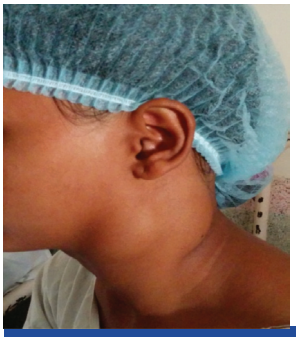
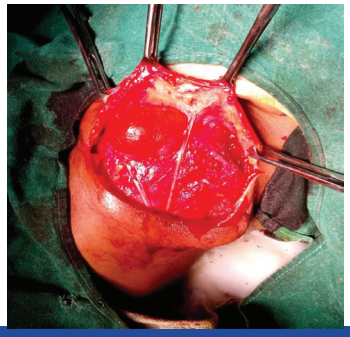
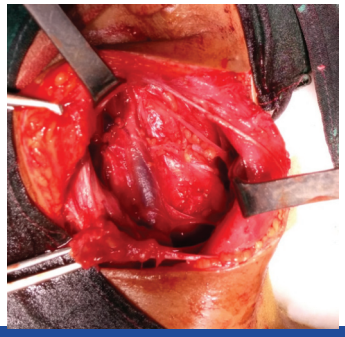
DISCUSSION

Cystic hygroma was originally reported by Redenbacker in 1828 and the name was first given by Wernher in 1834 [1]. Cystic hygroma is a benign congenital malformation of the lymphatic system, occurs as a result of sequestration of developing lymph vessels [2]. Cystic Lymphagioma occurs approximately 1 in 12000 births. Usually presents within 2 year of age, 50-60% by the end of 1st year, almost 80-90% before the end of 2nd year [3]. Prenatally diagnosed Cystic hygroma are associated with Turners syndrome, Noonan's syndrome, fetal hydrops.Very rarely for unknown reasons it can be present in adults (mainly in cervicofacial region) [4]. Trauma and infection have both been suggested as possible triggers for onset [5].

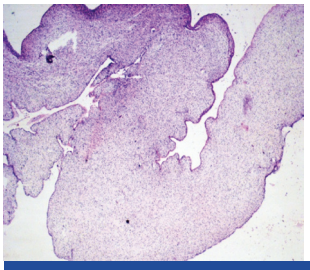
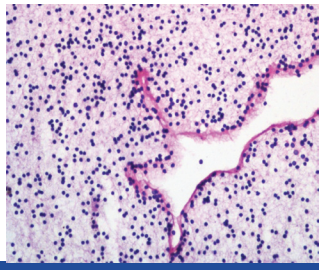
McGill and Mulliken [6] in 1993 classified Cystic hygroma based on CT, anatomical location, and histology. Type I malformations or classic cystic hygroma are macrocystic and develop below the mylohyoid muscle. They involve the anterior and posterior triangles of the neck. Type II malformations are microcystic 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.

Most common sites are posterior triangle of neck (75%), axilla (20%), mediastinum (5%), abdomen (liver, spleen, and colon), groin, pneumoperitoneum (kidney), scrotum, pelvic space, breast [7, 8].

Cystic hygroma presented in adulthood are usually asymptomatic. On physical examination Painless, Fluctuant, Tran-


[Table/Fig-1]: Cystic Transilluminant Swelling in Neck **[Table/Fig-2a]:** Cystic swelling with multiple septations containg clear fluid
[Table/Fig-2b]: - After complete excision of cyst preserving Jugular vein and accessory cranial nerve
[Table/Fig-3]: - Gross specimen of Cystic hygroma showing multiloculated masses composed of many cysts

[Table/Fig-4a]: Connective tissue stroma separating cystic spaces [H and E *10X] **[Table/Fig-4b]:** Connective Tissue Stroma contains Lymphoid cells [H and E *40X]

silluminable cystic swelling noted. Lymphangiomas are best visualized by magnetic resonance imaging (MRI); the high water content allows Lymphangiomas to appear hyperintense on T2-weighted images [9].

Histopathology shows thin connective-tissue stroma separating the cystically dilated spaces lined by a single layer of benign endothelial cells.

Differential diagnoses are Haemangioma, branchial cyst, Hamartoma, Lipoma, Cold abscess, Thyroid mass and Meningomyelocele.

Complications of cystic hygroma [are due to pressure effects (a) Neural encroachment symptoms like facial paralysis, vocal cord paralysis and shoulder weakness, (b) Dysphagia (pressure over esophagus), (c) Dyspnea (pressure over trachea), (d) Infection, (e) Sepsis and (f) Fistula formation [10, 11]. Infection within the cyst is usually caused by streptococcus or staphylococcus species [12].

Cystic Hygroma never undergo spontaneous regression. Complete surgical excision is the treatment of choice [13-15]. Care should be taken to avoid injury to important structures like Accessory cranial, Hypoglossal, Vagus, Lingual, lower branches of Facial nerves, Carotid sheath contents [13-15]. Recurrence rate after complete surgical excision is 20% [14]. Marsupialization, injection of Sclerosing agents, a steroids, diathermy and radiotherapy in unresectable or persistent cases can also be tried. Sclerosing agents used are OK432 [16] (a lyophilized mixture of group A streptococcus pyogenes of human origin), 50% dextrose, Triamcinalone, Bleomycin, Fibrin sealant and Hydrocolloid impression material.

CONCLUSION

Cystic Hygroma being a rare presentation among adults is a diagnostic and therapeutic challenge to surgeons. Surgery is the main treatment of choice. Recurrences are reported in spite of careful surgery.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interests regarding the publication of this paper.

REFERENCES

[1] Sabol SM, Bailey SB. Evaluation and surgical management of tumours of the neck: Benign tumours. In: Thawley SE.Panje WR, Batsake JG. Lindberg RD (Eds.). Comprehensive Management of head and neck tumours. vol. 2 W. B. *Saunders Company* 1999: 1416-49.
[2] Gleason T, Yuh W, Tali EA, Harris K, Mueller D authors. *Ann Otol Rhinol Laryngol.* 1993; Vol. 102:564–67.
[3] Karmody CS, Fortson JK, Calcaterra VE. Lymphangiomas of the head and neck in adults. *Otolaryngology HeadNeck Surg*. 1982; 90(3 pt 1):283- 88.
[4] Schefter RP, Oslen K, Gaffey T authors. Cervical lymphangioma in an adult. *Otolaryngology Head Neck Surg.*1985; Vol. 93:65–69.
[5] Antoniades K, Kiziridou A, Psimopoulou M authors. Traumatic cervical cystic hygroma. Int J.OralMaxillofacial. *Surg.* 2000; Vol. 29:47–48.
[6] Oakes MJ, Sherman BE. Cystic Hygroma in a Tactical Aviator: A Case Report. *MilMed* 2004; 169(12): 985-87.
[7] Nanson EM author. Lymphangioma (cystic hygroma) of the mediastinum. *J Cardiovascular surg.* 1968; Vol. 9:447–52.
[8] Chung SY, Oh KK, Kim DJ. Mammographic and sonographic findings of a breast cystic lymphangioma. *J Ultrasound Med* 2003; 22: 307-09.
[9] Terezhalmy GT, Riley CK, Moore WS. Lymphangioma (lymphaticMalformation). *Quintessence Int* 2001; 32: 495.
[10] Thawley SE, Panje WR, Batsakis JG, et al. authors. Comprehensive Management of head and neck Tumours. 1987. *Philadelphia, PA: Saunders;* p. 1350–52.

[11] Kennedy TL author. Cystic hygroma- lymphangioma: A rare and still unclear entity. *Laryngoscope.*1989; Vol. 99:1–10.
[12] Manikoth P, Mangalore GP, Megha V. Axillary cystic hygroma. *J Postgrad Med* 2004; 50:215-16.
[13] Farmand M, Kuttenberger J authors. A New therapeutic concept for the treatment of cystic hygroma.*Oral Surg*. 1996; Vol. 81:389.
[14] Chappuis JP author. Aspects actuels du lymphangiome kystique cervical. *Arch Padiatr.* 1994; Vol. 1:186.
[15] Batsakis JG author. Tumours of the head and neck. 1979. Baltimore, *MD: Williams & Wilkins;* p. 301–03.
[16] Smith MC, Zimmerman MB, Burke DK, et al. Efficacy and safety of OK-432 immunotherapy of lymphatic malformations. *Laryngoscope* 2009; 119: 107–15.

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