

Cervical Cystic Hygroma in an Adult

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Abstract

Cystic hygroma are benign tumors in the early childhood mainly manifesting in the head and neck region. Cystic hygromas manifesting in elderly patients have not been reported in the literature so far. We report a rare presentation of cervical cystic hygroma in an adult male (58 yrs.), with sudden appearance and rapid progression to a relatively big size (10 cm × 12 cm) involving superior mediastinum within a span of six months, giving a clue to malignant cystic pathology. Investigations proved the diagnosis of cystic hygroma. Transcervical excision was achieved with the help of cardiothoracic surgeons. The management of the cystic hygroma is being discussed.

Key Words

Cystic hygroma, Lymphangiomas

Introduction

Lymphangiomas, soft tissue tumors of disputed pathogenesis, 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 histogenic theories regarding the pathogenesis is either a congenital blockage of lymphatics (1,2) or a true neoplasm. Histopathologically, lymphangiomas are of three types. Lymphangioma simplex, cavernous lymphangioma and cystic hygroma, depending on the size of vascular spaces and thickness of the adventitia. Lymphangiomas are rare paediatric tumors and almost never reported in adults in the literature. These account for 6% of benign tumors in childhood and 5% of vascular tumors (4), invading various parts of the body including the retroperitoneum, mesentery, groin,

extremities, chest wall, mediastinum and within viscera (3), the head and neck region accounting 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% to 40%), occurring within 6 months to 1 year post-operatively. Also the mortality rate varies from 3-7%, the largest lesions having higher mortality, the death occurring mostly in post-operative period.

Case Report

A 58 years old male patient presented with an asymptomatic swelling involving right side of neck for past 6 months, swelling was small in size initially which

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gradually increased to size of 12 cm and 10 cm., extended from thyroid cartilage level till the clavicle (Fig. I). All the margins were well defined except the lower one which was going underneath the clavicle. Swelling was non-tender, mobile, non-pulsatile, cystic in consistency, non-compressible, fluctuant and the trans illumination test was positive. No bruit was there on auscultation. The trachea was deviated to left side & great blood vessels of the neck were normally palpable. Fine needle aspiration cytology (FNAC) revealed clear lymphoid fluid. CT scan revealed non contrast enhancing soft tissue mass extending from thyroid cartilage to sub-clavian artery with well preserved fat planes (Fig. II). It was not adherent to any vital structure in the neck. The internal jugular vein was compressed by the mass.

Transcervical excision of the mass was done under general anaesthesia. Sternal and clavicular heads of the sternomastoid muscle were separated and cystic mass (cystic hygroma) was delivered out with blunt finger dissection (Fig. III). Internal jugular vein was compressed but was patent on needle aspiration examination. Histopathologically, the diagnosis of cystic hygroma was confirmed.

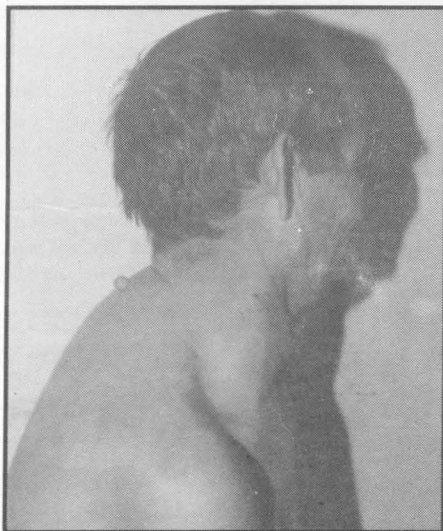


Fig. I. Showing cervical cystic hygroma extending from thyroid cartilage going deep beneath the clavicle, overlying skin is normal.



Fig. II. Non enhancing mass on (R) side extending from thyroid cartilage superiorly till sub-clavian artery inferiorly. Trachea has been pushed to (L) side.



Fig. III. Cystic hygroma being delivered by transcervical approach. Pointer is on the pedicle of cystic hygroma.

Discussion

Cystic hygromas are relatively rare lesions, approximately 65-75% of them are present at birth and 80-90% are identified by age of 3 years (4,5) and very rarely seen during adulthood. In head and neck cervical area is the predominant site for occurrence particularly the posterior triangle (75-80%), due to presence of extensive lymphatic system. Due to infiltrative nature of hygromas within the soft tissues of the neck, these may extend from posterior cervical area into the anterior compartment of neck, may cross the mid line, may reach into the cheek or down into the mediastinum and axilla. In general, symptoms vary from one mere presence of a

painless, enlarging mass to respiratory compromise, dysphagia and difficulty in feeding with regurgitation (4,5). Very rarely, massive hygroma may present with symptoms of neural encroachment (brachial plexuses and recurrent laryngeal nerve). Cystic hygroma vary from 1.0 to 30.0 cm. in size, the mean size in Stromberg's series was 8.0 cm (4).

The treatment of cervical hygromas has varied from benign neglect to complete excision. Surgical advocates (3,4), claim that these tumors may grow relentlessly, producing unacceptable cosmetic distortion of the face and neck, may compromise trachea and brachial plexus. Proponents of benign neglect cite the infiltrative hygromas as more risky while operating with the potential for incomplete excision, surgical sequel of neural injury, persistent lymphoedema, lymphocele and lymphorrhoea as being worse than the presence of the tumor itself.

The use of sclerosing agents has no role in the cure of hygromas. Aspiration of hygromas is useful only to decompress, when they are compromising the airways. Lymphangiomas being radioresistant, the radiation therapy is avoided for its ineffectiveness and potential for delayed carcinogenesis (1,4,5).

Surgical therapy, with as wide an excision as possible while preserving cosmetic function, is the best approach to these lesions (6). Staged excision may be necessary to avoid mutilating surgical procedure in some selected cases.

The surgical philosophy has been best summarised by Pott's :

“The objective in surgery of cystic hygroma is relief of obstruction upon vital structures and a good cosmetic result, good judgement must control the extent of the operation. Inadequate operation is just as inexcusable as a daring operation, and now here does this maxim apply more forcefully than in the surgical treatment of large cystic hygroma of neck”.

In conclusion, cystic hygroma are benign tumors of lymphatic system, manifesting mainly in early childhood and are best treated surgically with conservative approach.

References

1. Sabol SM, Bailey SB. Evaluation and surgical management of tumors of the neck : Benign tumors. In : Thawley SE, Panje WR, Batsake JG, Lindberg RD (eds.). *Comprehensive management of head and neck tumors*, vol. 2 W. B. Saunders company 1999 ; 1416-49.
2. McClure CRW, Sylvester CF. Sequestration of lymphatic tissue of developing jugular lymphatic sac. *Anat Rec* 1909 ; 3 : 534.
3. Van Cauwelaert PH, Gruwez JA. Experience with lymphangioma. *Lymphology* 1978 ; 11 : 43.
4. Stromberg BV, Weeks PM, Wray RC Jr. Treatment of cystic hygroma. *South Med J* 1976 ; 69 : 1333.
5. Waston WL, Mc carthy WD. Blood and lymph vessel tumors. A report of 1056 cases. *Surg Gynecol Obstet* 1940 ; 71 : 569.
6. Lynn HB. Cystic hygroma. *Surg Clin North America* 1963 ; 43 : 1157.