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RARE CASE OF CYSTIC HYGROMA IN ADOLESCENT

-Dr Virendra S Athavale, Professor, Department of General Surgery, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth,Pimpri, Pune, India 411018.

-Dr Prachi V Athavale, Assistant Professor, Department of Microbiology, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth,Pimpri, Pune, India 411018..

-Dr Rohan Jaiswal*, Resident, Department of General surgery, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, India 411018 (E-mail- jaiswalrohan0811@gmail.com)

Corresponding author- Dr Rohan Jaiswal (jaiswalrohan0811@gmail.com)

ABSTRACT

Cystic hygroma, a benign cystic congenital malformation of the lymphatic system that occurs in children younger than two years of age with an occurrence rate of 1 in 6000 new born, however is very rare in adolescence and adults. [1] Complete excision remains the treatment of choice giving an 81% cure rate. Partial excision was curative in only 12% of cases, Aspiration of the cysts has diagnostic and may provide symptomatic relief. [2] Following we demonstrate a case of rare occurrence of cystic hygroma in a 15 year old male patient and its management.

Keyword- Cystic hygroma, Cystic hygroma adolescent, Cystic hygroma excision, Cystic hygroma treatment

INTRODUCTION

As an anomaly in the growth of the lymphatic system, cystic hygroma (CH), also known as a lymphangioma, has its origins in the embryonic stage [1, 2,]. It accounts for 5% of benign congenital abnormalities in children [2].

The majority of cystic hygromas are seen in children under the age of two.[1] They are extremely uncommon in adults and are thought to be caused by the growth of lymph vessels in response to head and neck infections or trauma.[3]

The lymphatic system is derived from five basic sacs that were created from the venous system, according to research by Florence Sabin (1901). These are made up of two jugular sacs, one retroperitoneal sac, and two posterior sacs located close to the sciatic veins. The peripheral lymphatic system is formed centrifugally by endothelial outbuddings from these sacs. Secondary structures that form include the subclavian lymph nodes, thoracic duct, and cisterna chyli.[4]

The theory that cystic hygroma most likely develops from lymphatic tissue sequestrations during embryonic development was first advanced by McLure and Sylvester in 1909. He thought that these rests still had the ability to grow quickly and proliferate. The bigger lymphatic channels and these sequestrations might never form anastomoses.[4]

CH typically manifested as a soft, painless mass in the head and neck regions (75–80%), collarbone, and axilla. The posterior triangle of the neck is the most typical location for the tumor, and it may affect critical parts of the body like the sympathetic chain, carotid sheath, and branches of the hypoglossal, lingual, and face nerves [2,3]. The lesion is typically unilateral, fluctuant, and tender.

The mass effect causing compression on airway makes them potentially fatal in children.[3]

The aim of this report is to review our experience in the management of Cystic hygroma in adolescent population.

CASE PRESENTATION

-: A 15 year male presented with complaints of lump in left neck region since 1 year not associated with pain or breathlessness, Patient also had previous surgical history of excision of cystic hygroma from right side of the neck when he was 1 year of age, swelling was approximately 20x10 cm in dimension,extending from post auricular region to nape of neck, fluctuant and brilliantly transilluminating (refer to Fig 1,2,3), Similar smaller swellings were also noted on the occipital region approximately 3x3 cm in size,The remainder of his head and neck examination was entirely normal, and no significant deviation of the airway was noted on indirect laryngoscopy. A computed tomography (CT) scan of the neck with contrast was obtained; it revealed Computed tomography of neck was done which showed lobulated fluid density lesion of approximately 52x80x133mm is seen in the left side of neck deep to left sternocleidomastoid muscle extending in the posterior triangle of neck,abutting adjacent structures- left parotid gland, left common carotid artery, left internal jugular vein and left sternocleidomastoid muscle, Similar morphology smaller lesions were noted in subcutaneous plane posterior to posterior paraspinal muscle,right parieto-occipital scalp and left occipital scalp ranging approximately 30x10 mm in size (refer to fig 4,5). All above radiologically suggestive of cystic hygroma.

Decision of excision of lump in the left posterior triangle and occipital region under General anaesthesia was made. The lump was excised in toto without any harm to adjacent structures,negative suction drains were kept in situ. Histopathological examination revealed large irregular vascular spaces lined by flattened bland epithelial cells, the stroma showing fibrocollagenous and fibro adipose tissue along with lymphocytic infiltration consistent with cystic hygroma.

Postoperatively, the patient had an uncomplicated course. The suction drains placed under the skin flaps to maintain coaptation were removed on postoperative day 5. On postoperative day 7, the patient was discharged home with no obvious signs of recurrence in 5 months.

DISCUSSION

Cystic hygroma is thought to originate from a congenital lymphatic system abnormality in which lymph buildup results from a breakdown in connection between the lymphatic and venous routes. The majority of management literature takes into account paediatric cases because cystic hygromas typically present in gestation or in infancy.[5,6,7]

The preferred course of management for cystic hygromas has always been complete surgical resection. Sclerotherapy, however, may be a better suitable first-line therapy, according to a number of authors. Although the use of sclerotherapy in the treatment of neonatal and pediatric cystic hygromas is now well established, there have been comparatively fewer reports of its application to adult patients.[8]

One such case report is on a 32 years old female by S Woolley and DRK smith in 2008 in Otolaryngology Department, University Hospital of Wales, Cardiff, Wales, UK concluded that Treatment with OK-432 is useful in the management of cystic lymphangiomas in adults and should be considered as first line treatment however in their cases patient Following surgery, had a sudden growth in the size of the cystic lesion, along with an accompanying localised inflammatory reaction and fever.[9]Since the increase in size cannot be quantified or predicted one should be vary of the risk of post procedural airway obstruction

In an effort to prevent surgery, a number of sclerosing agents have been utilized, including OK-432[9], 50% dextrose[10], steroids, alcohol[11], bleomycin, cyclophosphamide, interferon alfa-2a[12].

The most effective sclerotherapy agent is alcohol, which has been used to treat varicose veins for over 100 years. However About 25% of patients experience acute blistering, and about 21% of patients experience skin necrosis with ulceration along with local inflammatory changes[13]

In the OK-432 trials by woolley a 60% reduction in size of tumor was observed when compared to complete surgical excision the results may not always be acceptable to patients and can be subjectively deemed unsuccessful in treatment outcome.[9]

In other non surgical modalities of treatment Radiation is also an option with varying degrees of success. The use of radiotherapy, which uses irradiation or the implantation of radon seeds, is restricted due to the obvious hazards involved and higher chances of recurrence[14]

Although currently Surgical excision remain the mainstay in treatment of cystic hygroma specially in adults, apprehension in dissection around the tumour still remains a bothersome point to many surgeon owing to frequently encountered thin walled swelling observed in cystic hygroma, Intra-op rupture is common causing loss of demarcating plane between tumor wall and surrounding structure leaving the possibility of residual structure post excision, One such study conducted by Reichelmann in 1999 about total,subtotal and partial excision of cervico-facial lymphangioma observed that When tiny plaques of the tumour wall were known to be remained in situ after subtotal excision, very low rates of recurrence we observed albeit still relatively more than the cases of total excision (1/9 patients).[15]

CONCLUSION

With all the above mentioned treatment modalities in consideration in our case, The treatment of choice was complete surgical excision as multiloculated cystic hygroma may not respond to sclerotherapy and radiation was completely omitted from options owing to its associated adverse effects. Patient along with his gaurdians were explained about the possible intra and post- operative complication such as bleeding, damage to neuro vascular structures,possibility of surgical site infection, Recurrence and post operative scar. After receiving consent from the patient's parents we went ahead with the surgery and to the best of our knowledge complete excision of tumor mass was achieved (refer to Fig 5,6) and very low chance of recurrence was expected considering historical evidence. No evidence of any obvious recurrence has been noted in our patient till date 6 months post-surgery and the patient still continues to be on follow up.

Further extensive studies are warranted for better understanding of cystic hygroma in adolescents and adults with a comparative analysis on efficacy between the various non-surgical and surgical treatment modalities available especially in adult demographic as owing to its rarity, well defined treatment guidelines still remains unavailable.

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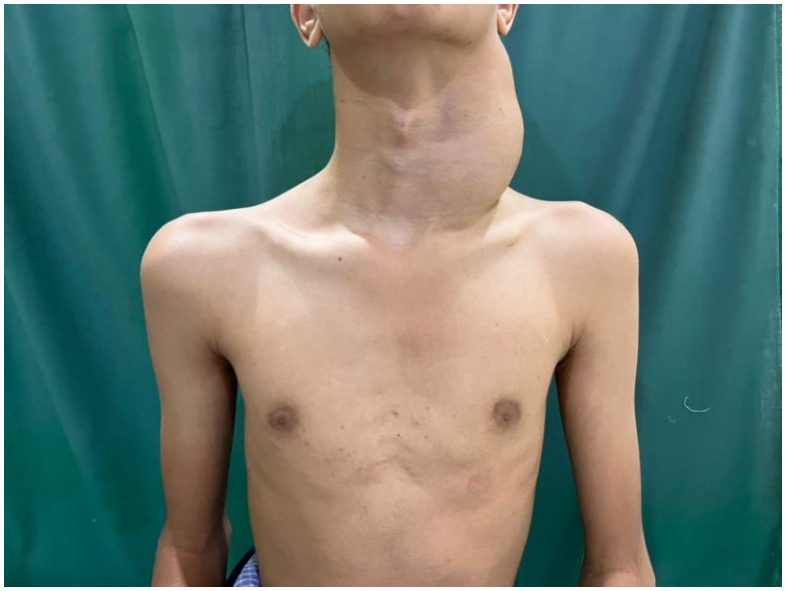
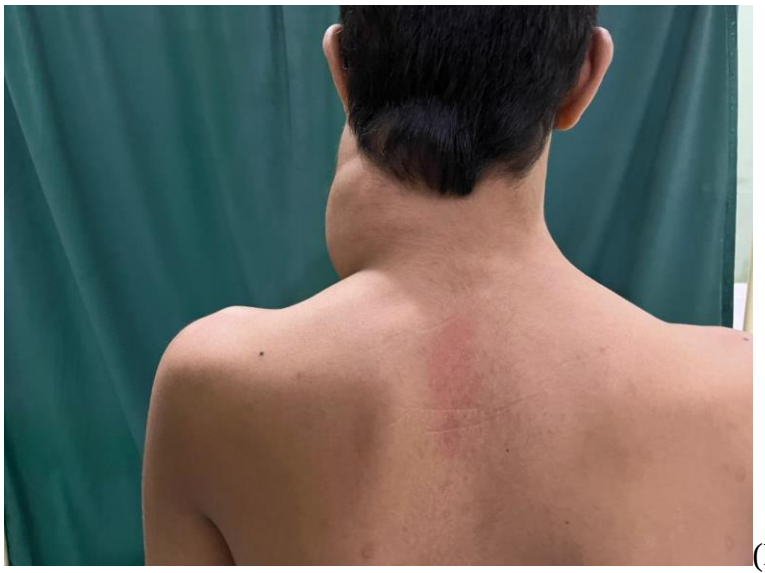

(Fig 1)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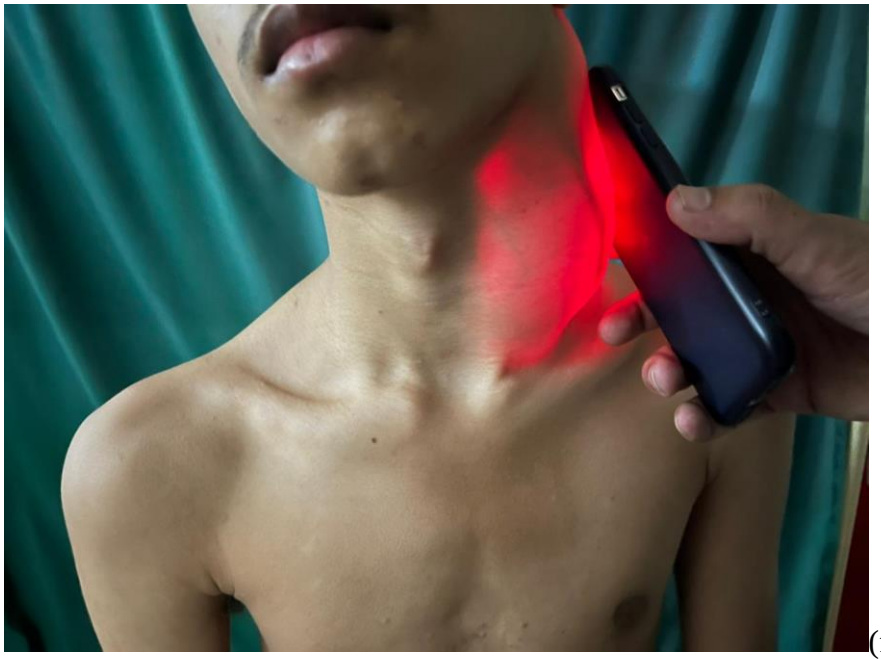
(fig 3)

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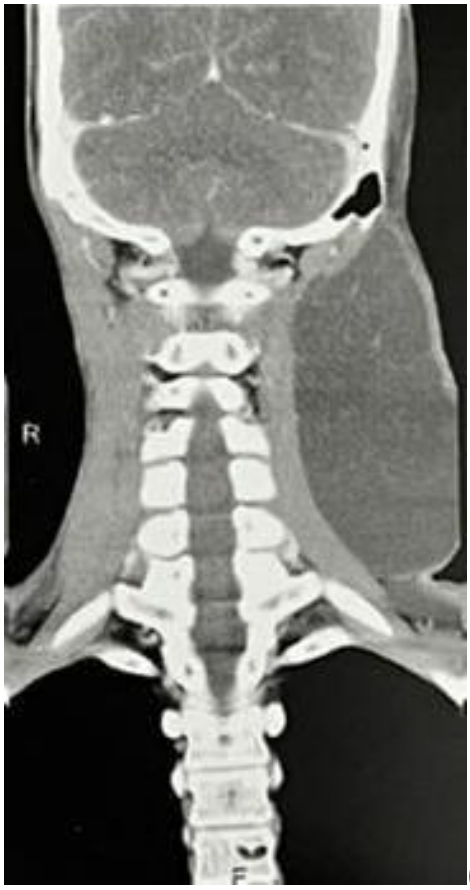
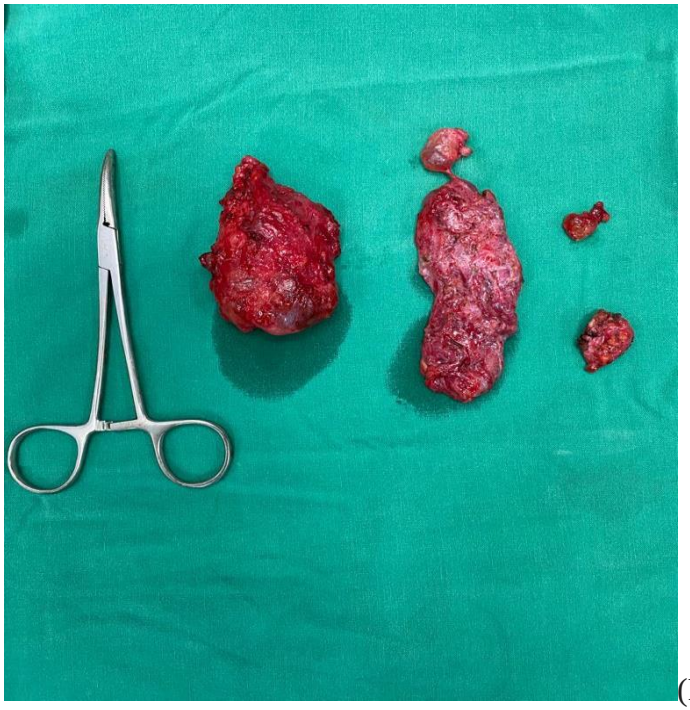
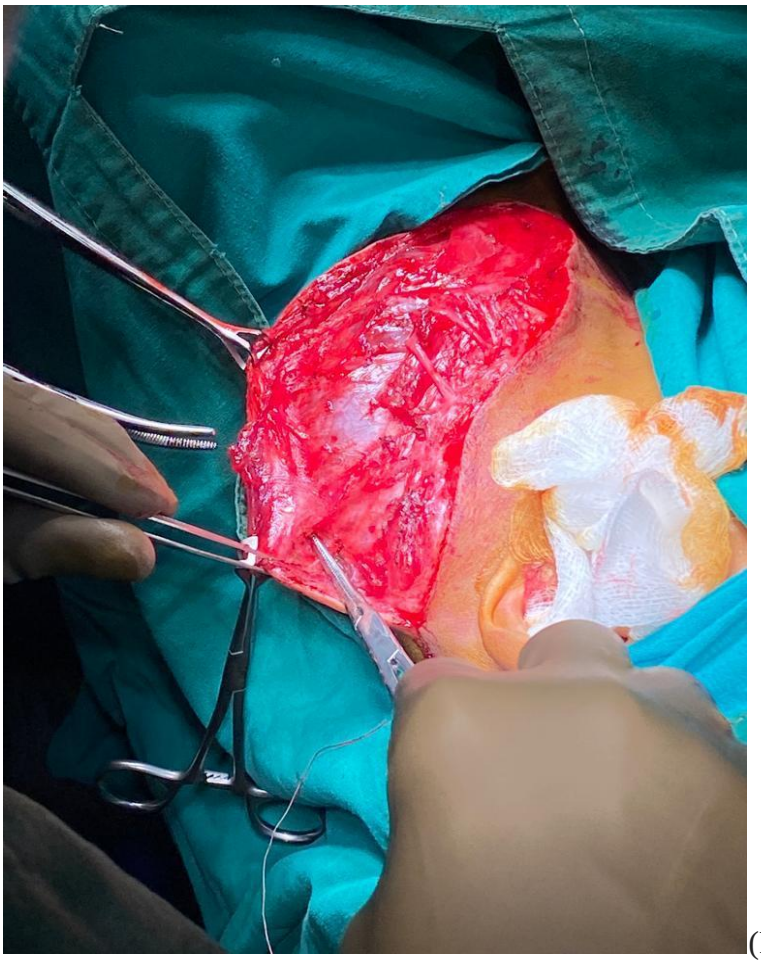
Clinical pictures of 15/Male case of left side cystic hygroma of neck



(Fig 4)



(Fig 5)Intraoperative photograph of excised specimens (Intra mass fluid evacuated)



(Fig 6)Intraoperative photograph of the tumor after elevation of skin flaps. The lesion has posterolaterally displaced the overlying sternocleidomastoid muscle