Case Report

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Surgery Section

An Unusual Case Report of Adult Cervical Cystic Hygroma

RATHNAGANPATHI THULASIKUMAR[1], THULASIKUMAR GANAPATHY[2], ABHIRAMA RAVINDRA KUMAR[3], SRIDHAR RAJAGOPAL[4]

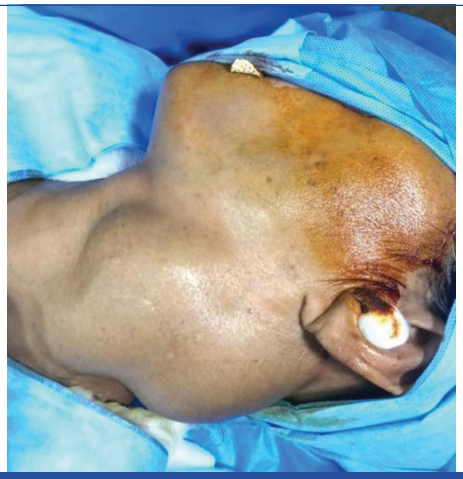
ABSTRACT

Cystic Lymphangioma (CL) is a rare condition that is typically observed as a congenital malformation in the head and neck. These congenital lesions usually appear before the age of two. Adult occurrences are extremely unusual, and only a few cases have been mentioned in the literature. CL is characterised by the collection of an epithelial lining of lymphatic fluid, which can be caused by lymphatic abnormalities, trauma, or surgery. In the present case report, the authors present the findings of a 49-year-old adult female who had painless, gradually increasing left-sided neck swelling for three years, with no other relevant medical history. The initial ultrasound revealed a large cystic lesion extending from the left sternocleidomastoid muscle to the left submandibular gland. The lesion measured 8×7×3 cm and exhibited narrow septae and loculation at the periphery. Avascularity was noted within the cyst wall and interlobular septae. The diagnosis favoured macrocytic lymphangioma/Congenital Lymphangioma (CH). During the surgical procedure, the cyst was dissected, and it was found to be located over the internal jugular vein, carotid artery, and spinal accessory nerve. The cyst was completely removed, and a surgical drain was left in place. The wound was sutured in two layers, and a sterile dressing was applied. The drain was removed on the third postoperative day, and the dressing with sutures was removed on the seventh postoperative day, revealing a healthy wound. The diagnosis was confirmed by histopathological examination {Haematoxylin and Eosin (H&E)} of the tissue specimen, which showed flattened epithelium lining the inside of the cyst. The cyst contained dilated gaps in a fibrocollagenous stroma infiltrated by lymphocytes. Therefore, CH can be considered as a differential diagnosis for head and neck masses in adults.

Keywords: Hydrocele of the neck, Lymphangioma, Lymph cyst

CASE REPORT

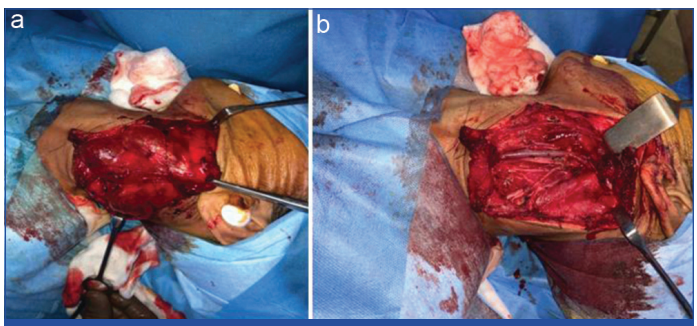
A 49-year-old North Indian woman had been experiencing painless swelling on the left side of her neck for the past three years. The swelling had progressively grown in size. She did not have any symptoms of neck compression, such as difficulty swallowing or breathing. There was no history of neck trauma, upper respiratory tract infection, previous medical conditions, or drug use. Her family members did not have similar complaints or a history of cancer. Upon examination, a solitary, non tender, brilliantly transilluminant, cystic swelling measuring approximately 10×10 cm was observed on the left side of the neck. The swelling had well-defined margins and extended 2 cm below the body of the mandible, 3 cm above the left clavicle, medially from the midline of the neck, and posteriorly up to the left mastoid process. There was no noticeable movement upon swallowing, warmth or redness of the skin, or palpable lymph nodes [Table/Fig-1].



[Table/Fig-1]: Image depicting the tumour over the left anterior and posterior triangles of the neck.

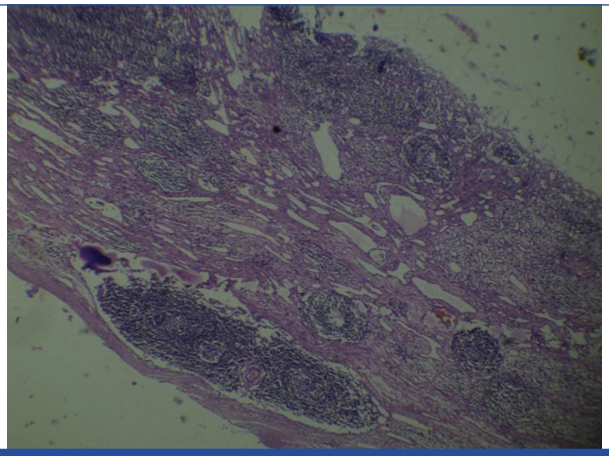
Ultrasonography (USG) revealed a large elliptical cystic lesion measuring 8×7×3 cm, extending from the left sternocleidomastoid muscle to the left submandibular gland. The cyst had narrow septae and loculations at its periphery, and it showed avascularity within the cyst wall and interlocular septae, leading to a diagnosis of macrocytic lymphangioma/CH. Surgery was performed under general anaesthesia. The skin was lifted apart with a platysmal flap, exposing the sternocleidomastoid muscle, which was horizontally sliced. The cystic mass, measuring approximately 5 cm along the widest diameter, was then removed.

The cyst was located over the internal jugular vein, carotid artery, and spinal accessory nerve [Table/ Fig-2a,b]. It was leaning on the sternocleidomastoid muscle and extended superiorly up to the submandibular gland. After complete removal of the cyst, a surgical drain was left in place, and the wound was sutured in two layers. The drain was removed on the third postoperative day, and the sutures and dressing were removed on the seventh postoperative day, revealing a healthy wound. The diagnosis was confirmed by histopathological examination of the tissue specimen [Table/Fig-3], which showed flattened epithelium lining the inside of the cyst, and the cyst contained dilated gaps in fibrocollagenous stroma infiltrated



[Table/Fig-2a,b]: Meticulous dissection of a Cystic Lymphangioma (CL) along with its excision in toto.

by lymphocytes. Lymphocytic aggregates can be seen in nearby foci. The patient has been sequentially followed-up for three years, and there has been no evidence of recurrence thus far.



[Table/Fig-3]: H&E picture of the specimen reveals flattened epithelium lines the inside of the cyst, which contains dilated gaps in fibrocollagenous stroma infiltrated by lymphocytes (4x).
Lymphocytic aggregates can be seen in nearby foci

DISCUSSION

Lymphangiomas are thin-walled cystic tumours that occur in the lymphatic system. They are similar to haemangiomas which develop in blood vessels. Lymphangiomas were first described by Redenbacher et al., in 1828 [1]. Congenital lymphatic abnormality known as CL is a benign condition, predominantly affecting children under two years old [2]. It is rare in adults [3]. The head and neck are commonly affected, and symptoms vary depending on the tumour's size and location [4].

The CL is also called macrocystic LM and has an incidence rate of 1.2-2.8 per 100,000 [5]. It can be congenital or caused by acquired factors that obstruct lymphatic channels [5,6]. Infections, trauma, and neoplasms are common causes. CL does not show gender preference and can occur anywhere on the body, with higher prevalence in the craniocervical, clavicle, and axillary regions [4]. Lymphatic malformations are classified as macrocystic, microcystic, or mixed. CL is often asymptomatic but can cause compressive symptoms such as dysphagia and dyspnoea due to compression of the esophagus, trachea, or larynx [7]. Kennedy TL et al., categorised lymphatic malformations into four types: superficial cutaneous, cavernous, CH (congenital lymphangioma), and diffuse systemic [2,6,8,9].

The CL can vary in size and occur in any part of the Head and Neck (HN) region according to physical examination. It typically presents as a lobular, soft, non tender, painless mass with unrestricted mobility. USG, Computed Tomography (CT) scan, and Magnetic Resonance Imaging (MRI) are commonly used imaging methods for assessment. The initial imaging approach should be a USG examination, which reveals a multilobulated cystic tumour with anechoic characteristics. CT or MRI, especially T1-weighted and T2-weighted MRI images, provide more accurate presurgery evaluation. Fine Needle Aspiration Cytology (FNAC) is controversial due to the risk of infection, haemorrhage, and recurrence [7,8].

From authors perspective, FNAC has limited diagnostic value as it does not impact treatment strategy. Histopathological investigation is necessary for a definitive diagnosis in adults. Sclerotherapy and laser therapy are alternative treatments used primarily in children but can also be adjuvant therapies in some adults [4]. "Watchful waiting" is recommended in certain cases to avoid surgical risks such as cranial nerve palsy, seroma, bleeding, infection, and tissue defects [9].

In authors' opinion, surgical total excision is the definitive treatment for CL in adults. Considering the size of the swelling, surgical intervention was the best option in the present case. However, other surgeons may opt for more aggressive approaches in massive encroaching swellings [10,11].

As the mass was located over the mid-neck, authors chose a mid-transverse neck incision instead of a low transverse neck incision. The former provides better exposure of both the upper and lower parts of the cyst without causing tension, retraction, or compromising flaps. However, compared to the latter, the mid-transverse incision may be less aesthetically appealing as it may not be easily concealed under clothing. Preserving the normal neovascular structures of the neck is crucial.

In a case reported by Sharma JVP et al., a Congenital Lymphangioma (CH) was observed in an adult with a lateral neck swelling, but no palpable lymph nodes were present [12]. Neck ultrasound revealed a large cystic swelling with external echoes, no vascularity, and no calcification. In another case reported by Bedir R et al., a 51-year-old male presented with hoarseness, and a cystic lesion of 0.6 cm in the right ventricle of the larynx was found to be a cystic cavernous lymphangioma on excision. Immunohistochemical staining was positive for CD31 and D2-40 [13]. However, in the present case, there was no history of hoarseness or laryngeal involvement. Another similar case reported by Yarso KY et al., described CH affecting the right lateral aspect of the neck in a 31-year-old woman, managed through USG-guided tissue biopsy and aspiration of fluid [14]. No recurrence was observed during a 6-month follow-up. In contrast, the present case underwent open and complete excision.

Incomplete excision is associated with recurrence, and the presence/absence of a well-defined capsule and the position of the lesion relative to the hyoid bone are also factors influencing recurrence. Therefore, an appropriate treatment strategy must be chosen. In present case, surgical excision was selected as the best treatment method to achieve total excision. The surgery was performed without complications, with careful preservation of vital cervical vessels and nerves, and there have been no reports of recurrence to date.

CONCLUSION(S)

Finally, lymphangiomas are commonly seen in children but are rare in adults. They are often misdiagnosed due to unclear clinical and radiographic features. Surgical excision and histopathological analysis of the specimen are necessary to confirm the diagnosis. Through describing the present case, authors aim to emphasise the rarity of lymphangiomas in adults and raise awareness that congenital lymphangioma can be considered as a differential diagnosis for head and neck masses in adults.

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PARTICULARS OF CONTRIBUTORS:
1. Senior Resident, Department of General Surgery, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.
2. Senior Consultant, Department of Vascular Surgery, Sooriya Hospital, Chennai, Tamil Nadu, India.
3. Junior Resident, Department of General Surgery, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.
4. Professor, Department of Plastic Surgery, Omandurar Government Medical College, Chennai, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Abhirama Ravindra Kumar,
No. 7, CLC Works Road, Chennai-600044, Tamil Nadu, India.
E-mail: abhiramark@gmail.com

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