

A RARE CASE OF HEAD & NECK MACROCYSTIC LYMPHANGIOMA IN AN ADULT

Neha Shrivastava¹, Sharad Bhalekar², Haritosh K. Velankar³, Yogesh Dabholkar⁴, Mahak Rohmehra⁵

¹Post Graduate Student, Department of ENT, Padmashree Dr. D. Y. Patil University School of Medicine, Nerul, Navi Mumbai.

²Associate Professor, Department of ENT, Padmashree Dr. D. Y. Patil University School of Medicine, Nerul, Navi Mumbai.

³Professor, Department of ENT, Padmashree Dr. D. Y. Patil University School of Medicine, Nerul, Navi Mumbai.

⁴Professor, Department of ENT, Padmashree Dr. D. Y. Patil University School of Medicine, Nerul, Navi Mumbai.

⁵Registrar, Department of ENT, Padmashree Dr. D. Y. Patil University School of Medicine, Nerul, Navi Mumbai.

ABSTRACT

Lymphangiomas are fluid-filled thin-walled cysts formed due to malformations in the lymphatic system. It is usually seen in children and is rarely present in adults. Majority of the cases of lymphangiomas are seen in head and neck region with occasional extension in mediastinum. We present a case of an adult with macrocystic lymphangioma in the left posterior triangle of neck. FNAC and MRI are important modalities of investigation for diagnosis of lymphangioma. Surgical excision is the main modality of treatment in case of macrocystic lymphangioma. In case of a soft cystic swelling in head & neck region, a differential diagnosis of lymphangioma should be kept in mind.

KEYWORDS

Macrocystic Lymphangioma, Head & Neck Lymphangioma, Lymphangioma, FNAC, MRI, Surgical Excision.

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INTRODUCTION: Lymphatic system is a network of vessels responsible for venous return and the defence mechanism of the human body. Malformations in this lymphatic system give rise to fluid-filled thin-walled cysts known as lymphangiomas. The incidence of lymphangiomas is 1.2–2.8%. They can be found at any age of life, approximately 50% are present at birth and 90% are diagnosed before 2 years of age and are rarely seen in adults¹. Lymphangiomas can be of congenital or acquired origin. Trauma, inflammation or lymphatic obstruction are considered as the causes of lymphangiomas of acquired origin.^{2,3} Approximately, 75% of the lesions are located in the head and neck region with occasional extension in the mediastinum.^{4,5} Three distinct morphological types of lymphangiomas have been described: capillary, cavernous and cystic. Capillary lymphangiomas usually consist of small superficial lesions that are generally asymptomatic. Cavernous and cystic lymphangiomas have similar histopathological features and are differentiated on the clinical presentation. The gold standard treatment is complete surgical removal with preservation of vital structures. We present a rare case of macrocystic lymphangioma in an adult.

CASE REPORT: A 52-year-old female presented in our facility with a gradually enlarging mass on the left side of neck since 2 years with mild local discomfort and occasional pain on neck movements and without any obstructive symptoms like dysphagia or dyspnoea (Figure 1). On clinical

examination, a single, soft, cystic, fluctuant swelling measuring 4 cm X 4 cm was present in the left posterior triangle of neck. MRI showed a large well-defined thin-walled cystic lesion in the posterior triangle of neck posterior to carotid space on left side measuring 4 cm x 4 cm in size. The lesion was hypointense on T1W images and hyperintense on T2W images suggestive of lymphangioma (Figure 2). On FNAC, smears showed mature lymphocytes with few degenerated lymphocytes in a pink proteinaceous background confirming the diagnosis of lymphangioma. Surgical excision of the swelling was done under general anaesthesia with preservation of important structures in the posterior triangle of neck (Figure 3). Patient was followed up for a period of 6 months with no radiological evidence of recurrence.

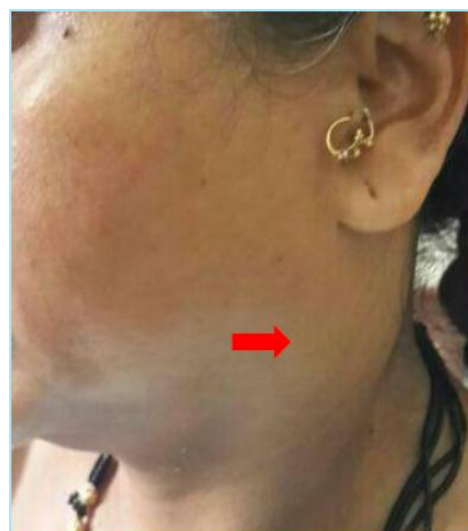


Figure 1: Macrocystic lymphangioma in left posterior triangle of neck

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Corresponding Author:

Dr. Sharad Bhalekar,

#H-7, 1/1, Paradise CHS, Sector-7, Sanpada,
Navi Mumbai-400705.

E-mail: sharadbhalekar2@gmail.com

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Figure 2: MRI T2 weighted image showing a large well-defined thin-walled cystic lesion in the posterior triangle of neck posterior to carotid space

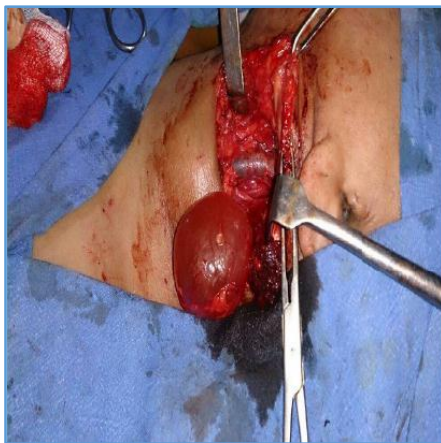


Figure 3: Surgical excision of the macrocystic lymphangioma

DISCUSSION: Lymphangiomas are benign lesions arising due to aberrations & abnormal development of the lymphatic system. They occur more commonly in children and are rarely found in adults. Lymphangiomas are usually seen in the head and neck region. The aetiology of lymphangiomas still remains unknown. It is believed that they are congenital malformations of lymphatic vessels or certain acquired factors resulting in lymphatic obstruction, lymph fluid retention, lymphangiectasia and proliferation. Three different types of lymphangiomas are described on the basis of morphologic and histological characteristics: capillary, cavernous and cystic lymphangioma.⁶ The mass can be present at birth but, if small, can remain unnoticed until an incidental trauma occurs at the site.⁷ Lymphangiomas can also be classified into microcystic, macrocystic, and mixed subtypes, according to the size of their cysts. Microcystic lymphangiomas are composed of cysts, each of which measures less than 2 cm³ in volume. Macrocystic lymphangiomas contain cysts measuring more than 2 cm³ in volume. Mixed type contain both microcystic and macrocystic components. Lymphangiomas may be described in stages, which vary by location and extent of disease. The stage depends on whether lymphangiomas are present above hyoid bone (suprahyoid) or below hyoid bone

(infrahyoid), and whether the lymphangiomas are on one side of the body (unilateral) or both (bilateral) with suprahyoid lesions having poor prognosis as compared to infrahyoid lesions.⁸ The different modalities of treatment available are surgical excision, laser excision, sclerotherapy, radiotherapy, electrocoagulation, cryotherapy & embolization. Surgical resection of lymphangiomas still remains the main modality of treatment. In our case, a horizontal incision was taken over the swelling at the junction of upper one third & lower two thirds of sternocleidomastoid (SCM). After reflecting the upper & lower flaps, the lesion was seen posterior to carotid space & deep to SCM. The thin-walled cystic lesion was carefully dissected & removed after retracting SCM like a sling. In case of ill-defined lesions, alternative modalities of treatments should be considered. The various classifications & staging system of lymphangiomas offer little help in their management.⁹

CONCLUSION: Although lymphangiomas are rare in adults, in case of a soft cystic swelling in neck a differential diagnosis of lymphangioma should also be considered for early recognition. Surgical excision has been the cornerstone of treatment for lymphangiomas of the head and neck particularly in case of well-defined macrocystic lymphangiomas. Macrocystic lymphangiomas are thin-walled cysts; hence utmost care should be taken while dissecting for their complete removal.

REFERENCES:

1. Bailey CM. Cystic hygroma. Lancet 1990;335(8688):511-2.
2. Gleason TJ, Yuh WT, Tali ET, et al. Traumatic cervical cystic lymphangioma in an adult. Ann Otol Rhinol Laryngol 1993;102(7):564-566.
3. Aneesh kumar MK, Kale S, Kabbani M, et al. Cystic lymphangioma in adults: can trauma be the trigger? Eur Arch Otorhinolaryngol 2005;262(4):335-337.
4. Pia F, Aluffi P, Olina M. Cystic lymphangioma in the head and neck region. Acta Otorhinolaryngol Ital 1999;19(2):87-90.
5. Curran AJ, Malik N, McShane D, et al. Surgical management of lymphangiomas in adults. J Laryngol Otol 1996;110(6):586-589.
6. Riechelmann H, Muehlhaff G, Keck T, et al. Total, subtotal, and partial surgical removal of cervicofacial lymphangiomas. Arch Otolaryngol Head Neck Surg 1999;125(6):643-648.
7. Roisman I, Manny J, Fields S, et al. Intra-abdominal lymphangioma. Br J Surg 1989;76(5):485-489.
8. De Serres LM, Sie KC, Richardson MA. Lymphatic malformations of the head and neck. A proposal for staging. Arch Otolaryngol Head Neck Surg 1995;121(5):577-82.
9. Bhalekar SB, Shetty Y, Velankar HK, et al. Multi-disciplinary approach towards lymphangiomas of the head and neck in adults. Int Surg J 2016;3(1):245-248.