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CASE REPORT

Cervicomediastinal cavernous lymphangioma in an adult

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ABSTRACT

Background: Cavernous lymphangiomas are rare benign lymphatic tumours in the head and neck region. Cervicomediastinal extension of cavernous lymphangiomas are exceedingly rare in adults. This is probably the first published report of adult cervicomediastinal cavernous lymphangioma in India to our knowledge, highlighting its clinical presentation, diagnostic process, and management. **Case Presentation:** This case report describes a 20-year-old male with a painless neck swelling found intraoperatively to be a cervicomediastinal cystic mass, which was surgically excised and confirmed as a cavernous lymphangioma on histopathology. **Conclusion:** This case underscores the importance of considering cervicomediastinal cavernous lymphangioma in young adults presenting with neck masses. Early diagnosis and surgical intervention are crucial to prevent complications associated with mass effect on adjacent structures. The patient remains asymptomatic with no recurrence at follow-up, emphasizing the effectiveness of surgical management in such cases.

Keywords: Lymphangioma,Cavernous; Lymphangioma,Cervicomediastinal; Lymphangioma,Adult; Branchial Cyst; Vagus Nerve; Laryngoscopy

INTRODUCTION

Lymphatic malformations are congenital abnormalities of the lymphatic system characterized by the buildup of fluid in cystic spaces and channels. They are believed to result from the sequestration of lymphatic tissue during the formation of lymphaticovenous sacs, which fail to communicate with the rest of the lymphatic or venous system. These lesions later develop a cystic appearance due to the dilation of the sequestered lymphatic tissues. [2]

Lymphatic malformations are less common in adults compared to children, with an incidence of 1 in 100,000 adults admitted to hospitals versus 1 in 20,000 children. They account for less than 6% of childhood benign tumours, or 1.2 to 2.8 per 1,000 cases. Up to 75% of these malformations occur in the cervicofacial region, likely due to the predominance of the lymphatic system in this area.[3] In children older than two years, less than 10% of patients are affected, with an equal distribution in both sexes. These malformations can develop before birth, become evident at birth, or appear in the first few months of life, and may occasionally extend into the mediastinum.

Ultrasound (US) and magnetic resonance imaging (MRI) scans are commonly used to diagnose lymphatic malformations. However, complete surgical excision can be challenging due to the anatomic relationships in the neck, often leading to disease recurrence, typically in the early postoperative phase. The case report presented here discusses a case of cervicomediastinal cavernous lymphangioma in an adult and its management.

Histologically, lymphatic malformations can be categorised as being macrocystic (previously called cystic hygromas) or microcystic (cavernous lymphangiomas). Serres et al(1995) classified lymphatic malformations according to their extent.[Table 1]

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Table 1: Classification of lymphangiomas based on their location

STAGE	LOCATION
I	Unilateral infrahyoid lesion
II	Unilateral suprahyoid lesion
III	Unilateral lesion extending both above and below the hyoid
IV	Bilateral infrahyoid lesion
V	Bilateral lesion extending both above and below the hyoid

CASE REPORT

A 20-year-old male patient presented to the ENT OPD with a swelling over the left cervical region which was insidious in onset and gradually progressive since 1.5 years. The mass was slow growing without any symptoms. On examination, a painless 3x3 cm diameter smooth mass lesion was palpated and the anterior margin of sternocleidomastoid muscle in the middle 1/3rd region.

USG of the neck revealed one well demarcated elliptic shaped cystic structure (3.58 x 1.31 x 2.84 centimetres in size) is seen at the left side of the neck anterior to the sternocleidomastoid, having imperceptible walls and dense echogenic internal fluid content without any obvious increased internal vascularity most likely branchial cleft cyst (likely branchial cleft cyst). [FIG 1]

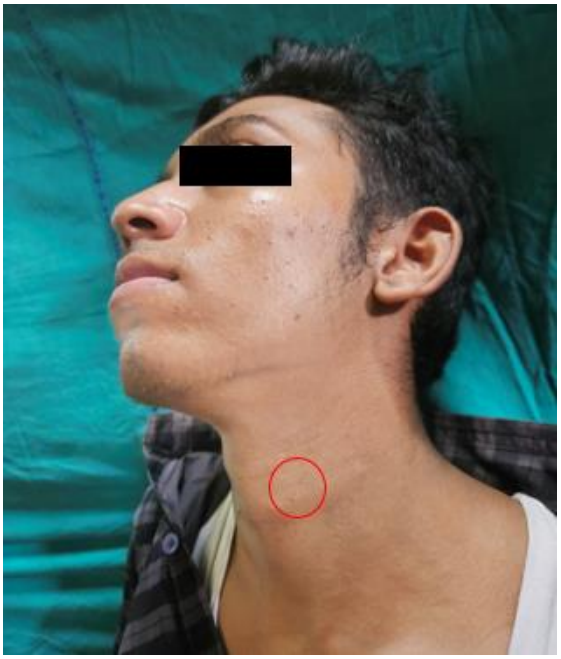

FIG 1: 20 year old patient with 3x3 cm painless left cervical swelling

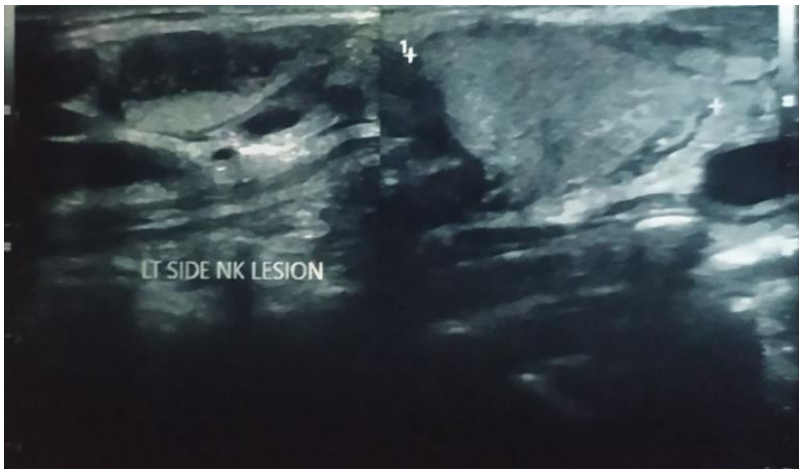

FIG 2: one well demarcated elliptic shaped cystic structure (3.58 x 1.31 x 2.84 centimetres in size) is seen at the left side of the neck anterior to the sternocleidomastoid, having imperceptible walls and dense echogenic internal fluid content without any obvious increased internal vascularity

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On fibre optic laryngoscopy, no internal communication to the tonsillar fossa base or pyriform sinus was noted. [FIG 3a]

Based on the initial assessment, the patient was planned for excision of the branchial cyst under general anaesthesia after informed consent from the patient. During surgery, an anterior neck incision was made from left anterior border of Sternocleidomastoid muscle to right anterior border. The cystic structure was dissected from strap muscles. The superior extent was upto the angle of mandible, and inferior extent was beyond the clavicle into the mediastinum adherent to the carotid sheath. The cystic mass was separated carefully from the internal carotid and vagus nerve and tied at the angle of mandible superiorly. The inferior margin was dissected and was tied by using Negus ligature slipper. [FIG 3b]

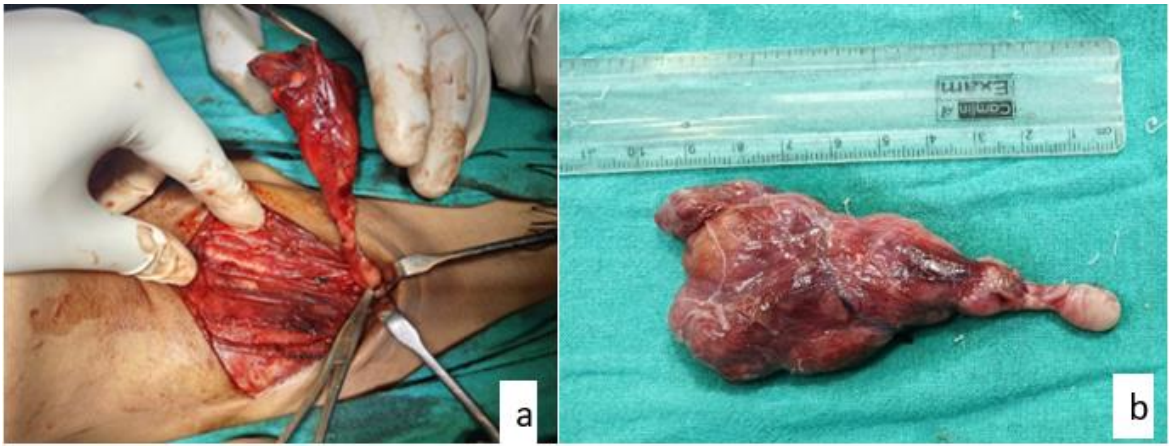

FIG 3 (a) Showing a cystic mass dissected superiorly and inferior margin going into mediastinum tied by using Negus ligature slipper(b)9.8 x 4 cm cystic specimen sent for histopathology

CHALLENGES

The inferior limit of the duct from the cystic where not identified and hence a remnant of the mass was left behind. The intra operative surgical extension of the mass did not corroborate with the initial diagnosis of branchial cyst. Careful dissection from the vagus nerve branches was done, however traction during dissection of one of the branches encasing the cyst could have led to post-operative hoarseness of the patient which improved in 3 weeks. [FIG 3b]

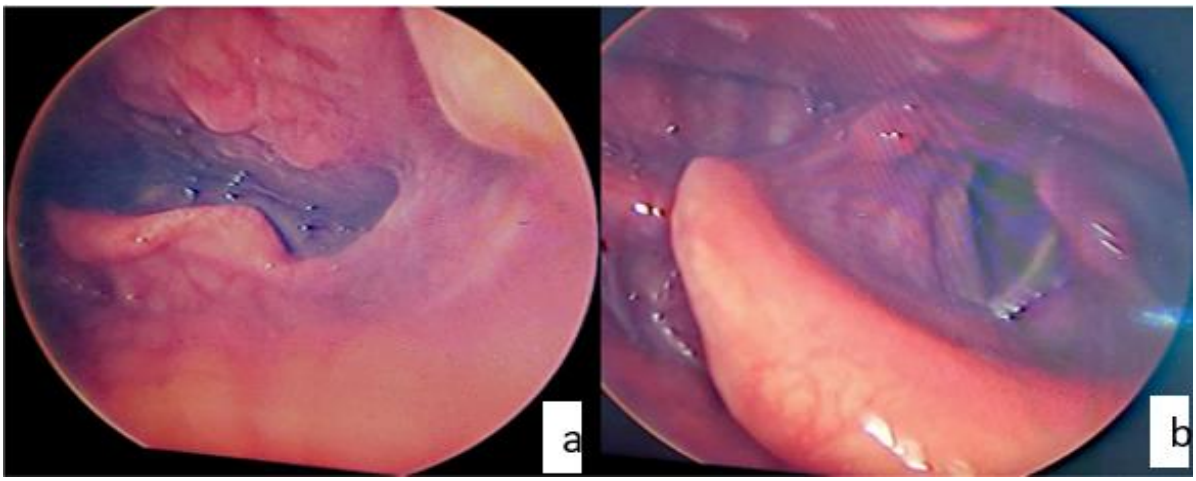

FIG 3 (a)Pre-operative laryngoscopy showing no internal communication of cystic mass to the tonsillar fossa base or pyriform sinus(b)Post-operative laryngoscopy showing hypomobility of left vocal fold

On histopathological examination, a single irregular brownish multiloculated cyst 9 x 4 x 1 cm3 with smooth, shiny, intact outer surface and yellowish thick material inside was noted. A diagnosis of cavernous hemangioma was made. [FIG 4]Currently the patient is 2 months post operative, and there is no recurrence.

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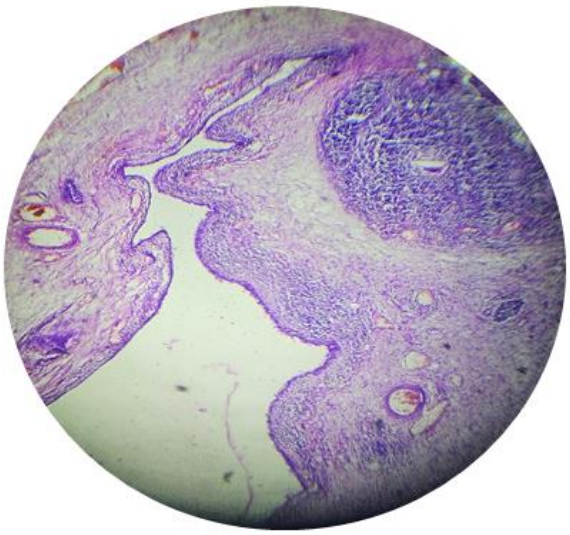

FIG 4: Dilated lymphatic vessels lined by flattened epithelium and surrounded by lymphoid follicles (H&E, x100)

DISCUSSION

Lymphangiomas are predominantly benign, non-tender, soft, compressible, and slow-growing masses that rarely cause mass effects or bruit similar to our case. [2] However, they can lead to symptoms like dysphonia, dysphagia, and airway obstruction depending on their location. In this case , the adult patient had a swelling on the left side of the neck for 1.5 years without any symptoms. Iatrogenic injuries, neoplasms, infections and trauma causes adult presentation, which were absent leading to first erroneous diagnosis. The mediastinum may occasionally be involved in their gradual evolution. [4] According to Table 1 classification of our case is Stage III lymphatic malformation.

The differential diagnosis includes cystic lymphangiomas, dermoid cysts, thyroid cysts, aneurysms, lipomas, tuberculous lymph nodes, or cervical lymph node metastases. Imaging modalities like ultrasound, CT, and MRI are useful in defining the extent and relationship of the lesion with surrounding structures.[5]

A therapy decision-making system based on five distinct stages of lymphangiomas suggests that small asymptomatic lesions may not require treatment, while larger or more functionally impaired lesions (stages II-IV) may benefit from surgery with or without sclerotherapy. However, stage V lesions with a higher risk of complications may be better suited for non-surgical treatment. [6-7] Despite surgery being the most commonly recommended approach, 50% higher recurrence rates have led to the exploration of various other treatment modalities, including aspiration, radiotherapy, laser therapy, and sclerotherapy using agents like bleomycin, OK-432, and doxycycline. [4] The location and size of the lesion can influence the recurrence risk, with suprahyoid lesions having a higher rate due to the difficulty in complete excision without damaging critical neurovascular structures.[8] Large infiltrating lesions to the local structures could result in inadequate resection and local recurrence, although Fliegelman et al.'s [9] finding of a strong association between the surgical and microscopic assessments of encapsulation did not support that claim.

There are a few case reports of adult cervicomediastinal cavernous lymphangiomas in the World such as a case report described a 35-year-old male with a large mediastinal lymphangioma extending from the neck to the mediastinum. The patient underwent complete surgical excision of the lesion. [Teramoto et al 2008] [10] However there was no cervical extension. Another case report discussed a 28-year-old male with a cervical lymphangioma involving the neck and anterior mediastinum. The patient underwent surgical excision of the lesion. [Sargunam et al 2013] [1] However there was no mediastinal involvement. Another case series reported two adult patients with cervicomediastinal lymphangiomas. One patient was a 35-year-old male and the other was a 42-year-old female. Both underwent surgical excision of the lesions. [Rasaretnam et al 1976] [11]

In this case, the patient had a cervicomediastinal cavernous lymphangioma, which is a rare presentation in adults.This makes this case probably the first published report of adult cervicomediastinal cavernous lymphangioma in India to our knowledge.

Lymphangiomas can recur following inadequate excision, 80% within the first three to five years. Recurrence should be considered in the differential diagnosis when new symptoms appear in the head and neck region. Careful surgical planning and a multidisciplinary approach are essential for managing these complex and recurrent lesions.

STATEMENTS AND DECLARATIONS

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legal guardians mentioned in the article for their images and details for publication.

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