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Lymphangiomas in Adults: A case series

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Abstract
Lymphangiomas of the head and neck region result from abnormal growth of the lymphatic vessels and are mostly benign. They are mostly diagnosed before the age of 2 years. The treatment of choice is complete surgical removal ; however the tumor tends to spread along vital structures therefore sometimes complete surgical removal is impossible. We describe the clinical and pathological features of cystic lymphangioma diagnosed in three adults with lateral neck mass. Clinical examination , radiological assessment and fine needle aspiration cytology were performed. Confirmed diagnosis is reached only after histopathological examination of the surgical specimen posing a diagnostic difficulty.
Despite the rarity of adult lymphangiomas these cases suggest that we need to consider cystic lymphangioma as a differential diagnosis for lateral neck masses.

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I. Introduction

Lymphangiomas are benign malformations that occur most commonly in childhood. (1) The initial appearance of lymphangioma or cystic hygromain adulthood is rare with less than 100 cases reported in literature.(2) There are three theories proposed for its pathogenesis- firstly blockage or arrest of normal growth of primitive lymph channels ,secondly primitive lymph sac fails to reach the venous system and thirdly lymphatic tissue lies in the wrong place.

Lymphangiomas are thought to arise from an embryological defect that consists of an abnormal connection from the jugular sac to peripheral lymphatic system.(3)

Surgical excision is the best treatment. Sclerotherapy maybe an alternative treatment. Bleomycin, tetracycline, Doxycycline, dextrose and OK – 432 have been used for sclerotherapy. Most of the agents except OK-432 lead to extensive perilesional fibrosis which may complicate the salvage later. (4)

We discuss three cases of adult onset lymphangiomas presenting with swelling on the lateral aspect of the neck . After due clinical , cytopathological and radiological assessment the patients underwent surgery as a definite treatment of choice.

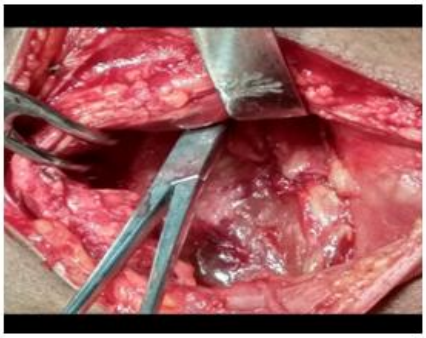
Case 1

A 28 year old female presented with a neck swelling on the right side which she noticed 6 months ago and had since then gradually progressed from the size of a lemon to a melon. On clinical examination the swelling was oval in shape , the skin was not erythematous with no engorged veins or discharging sinuses and did not cause any dyspnoea , dysphagia or restriction of neck movement. On palpation, the swelling was cystic in consistency with no local rise of temperature, fluctuant with illumination test positive and no tenderness measuring 12x15 cm.

On radiological imaging , a well marginated,unilocular cystic mass measuring 10x10 cm reaching up to the apex of axilla was seen.

After complete anaesthetic evaluation the patient was operated upon. Intraoperatively a large cystic mass was seen in the supraclavicular space displacing right sides sternocleidomastoid muscle insinuating between the clavicle and the anterior rib cage. Post operatively the patient did well with no weakness or restriction of neck movement and is on follow-up till date. The tissue sent for histopathological examination showed features consistent with lymphangioma.

Preoperative Photograph



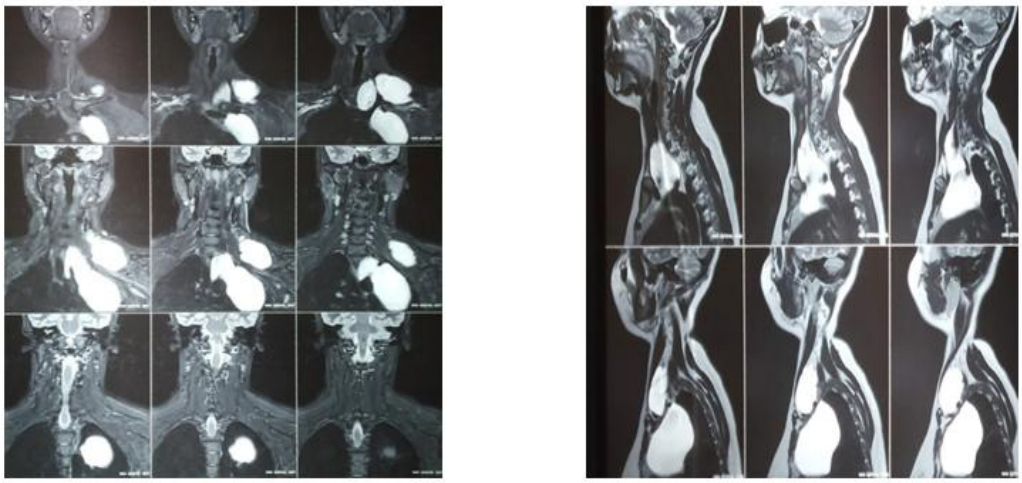
Intraoperative Photograph

Case 2

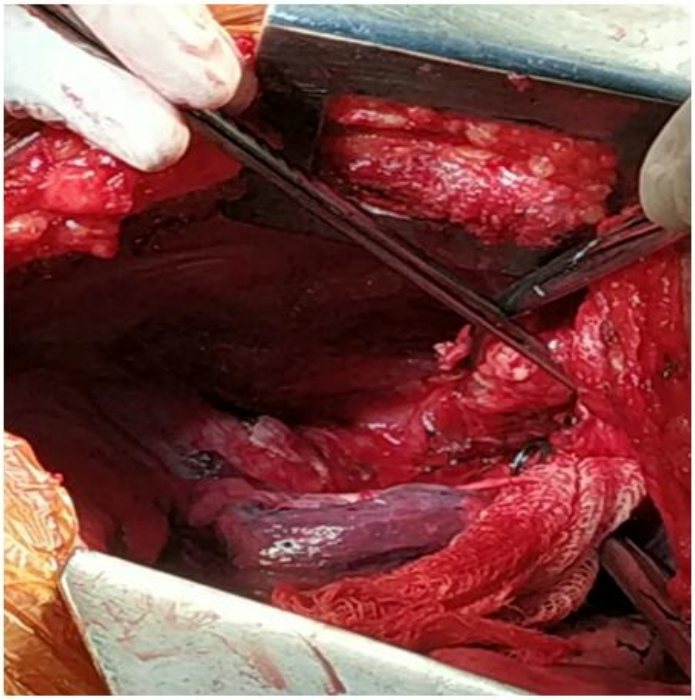
A 21 year old female came with the complaint of mild swelling over the left supraclavicular region and lateral aspect of neck since 3 weeks. She did not complain of any dyspnoea , dysphagia, pain or restriction in neck movement. On inspection there was a mild bulge on the left supraclavicular region, no erythema over the skin or discharging sinuses , pulsations or engorged veins were seen. The swelling was an approximate size of 8x6 cm. On palpation , the swelling was cystic, non tender, reducible with no fixation to skin measuring 7.5x7 cms with an indistinct margin.

MSCT neck showed a large multilocular cystic mass lesion in the left side of neck extending from the level of thyroid into the anterior and middle mediastinum , encasing the arch vessels. It was closely abutting the aortic arch and pulmonary artery and had multiple internal septations but no internal vascularity.

Fine needle aspiration cytology from the swelling revealed serous fluid suggestive of lymphangioma. After a complete anaesthetic evaluation the patient was prepared for surgical excision in conjunction with the department of cardiothoracic vascular surgery. The swelling was excised without sacrificing any major vessels or nerves via a thoracotomy and a neck incision. Post operatively the patient did well with no dyspnoea, dysphagia or weakness / restriction of neck or upper limb movements. The healing 3 weeks post operatively of the neck incision and thoracotomy was satisfactory.



Radiological Imaging of the patient

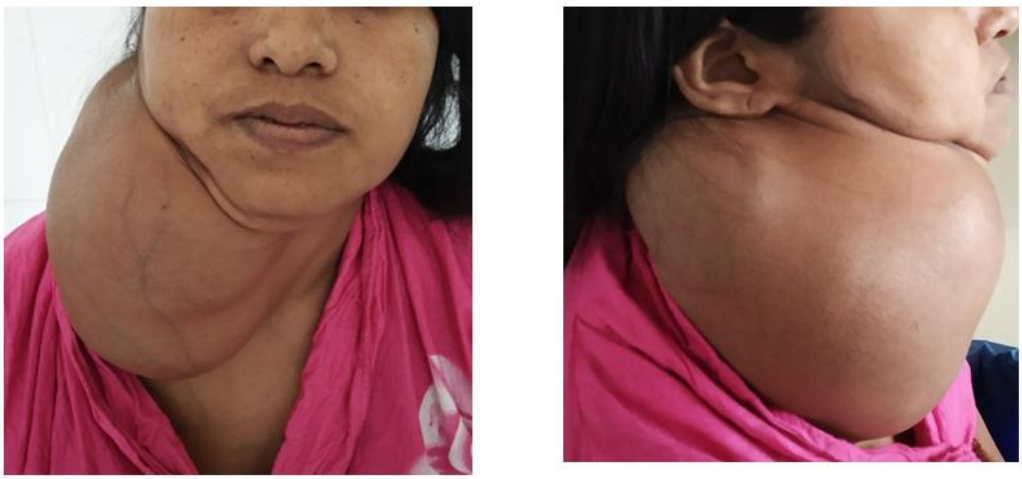


Intraoperative picture of the patient

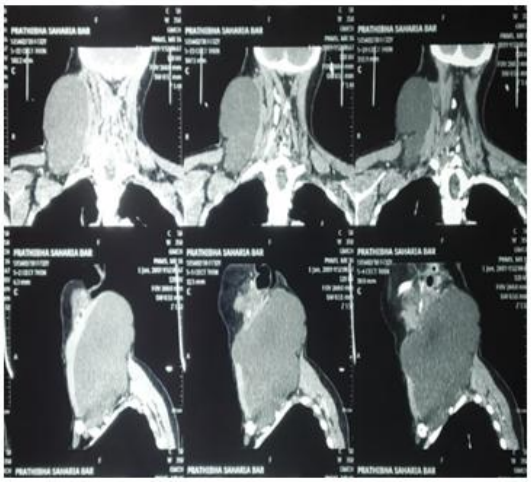
Case 3

A 33 year old female presented with rapidly increasing swelling on the right side of the neck since 9 months . The patient also complained of difficulty in neck and shoulder movement and occasional dyspnoea on lying down in the left lateral position. On inspection , the swelling extended from the angle of the mandible to the clavicle externally. There were engorged veins over the skin surface but no erythema, discharging sinuses or pulsations. On palpation there was no local rise of temperature, the swelling had indistinct margins , was cystic in consistency , fluctuant , compressible but not reducible with transillumination test positive. Needle aspiration revealed serous fluid and it was surgically excised after a thorough anaesthetic evaluation. Post operatively patient had definite improvement in the ability to shrug the shoulders and neck movement.

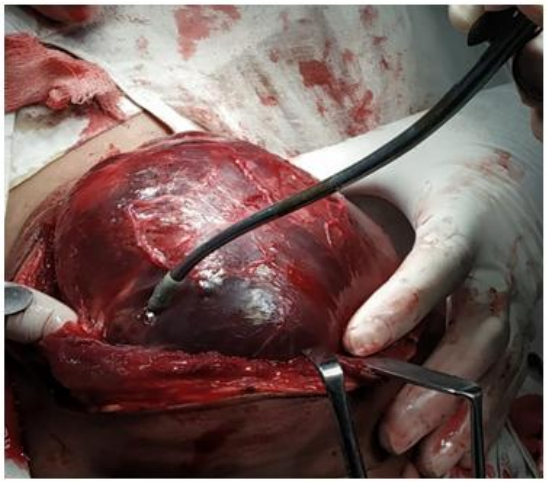
There was no post operative dyspnoea/ dysphagia. Patient was followed up 4 weeks post operatively and showed satisfactory healing.



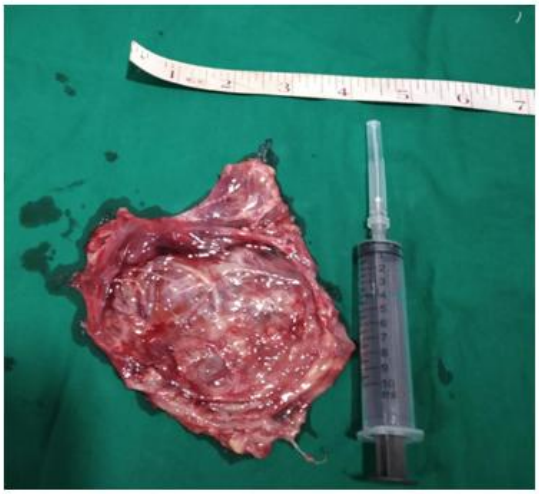
Preoperative photographs of the patient



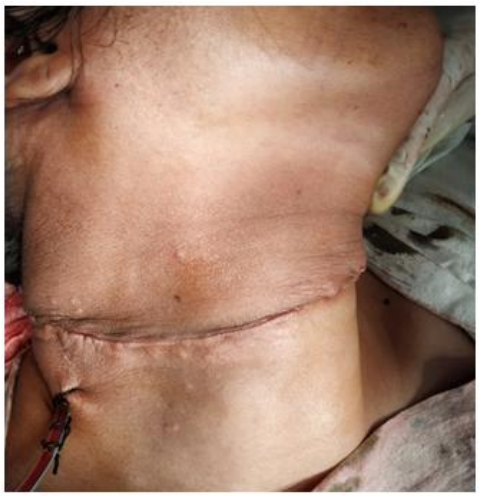
Radiological Imaging of the patient



Intraoperative photograph



Post operative specimen



Post operative scar on Post operative day 3

II. Discussion

Lymphangiomas are rare congenital malformations of the lymphatic system that may occur anywhere in the body.. They can be classified as microcystic (i.e. capillary lymphangioma), macrocystic (i.e. cavernous lymphangioma) and cystic hygroma.[5] Lymphangiomas of the mixed type contain both microcystic and macrocysticcomponents.Finally, lymphangiomas may be described in stages, which vary by location and extent of disease.[6]

Stage I- Unilateral infrahyoid;
Stage II- Unilateral suprahyoid;
Stage III- Unilateral suprahyoid and infrahyoid;
Stage IV- Bilateral suprahyoid; and
Stage V- Bilateral suprahyoid and infrahyoid.

Most lymphangiomas are asymptomatic. They usually present as a progressively enlarging painless mass. Typically, the mass is soft, non-tender, and ill-defined. Symptoms may develop due to mass effect when the lymphatic malformation enlarges and compresses surrounding vital structures. Obstructive symptoms of dysphagia, dyspnea, or airway compromise may develop.

Ultrasonography and computed tomography (CT) scanning have been used extensively to evaluate the anatomy of lymphangiomas. On ultrasound examinations, they appear as thin-walled, multiseptate, multicystic, hypoechoic masses.[7] On CT scanning lymphangiomas appear as multiloculated cystic lesions. On T2-weighted MRI, lymphangiomas appear isointense to cerebrospinal fluid, whereas their intensity varies on T1-weighted images due to variable protein content.[8] Fine-needle aspiration (FNA) is routinely used to evaluate most neck masses. FNA findings include small and round lymphocytes with intermingling histiocytes without mitoses or atypia. In adults with compromised airways, FNA may be therapeutic, as well as diagnostic.[9]

Surgery is considered to be the treatment of choice but given the close proximity of these to vital structures of neck and insinuating nature, plan for subtotal or near total excision should be tempered with symptomatic recurrence and scarring encountered with reexplorations. Hence, a pragmatic approach is advocated in treating these lesions. Balakrishnan K et al consider primary end point of surgery should be functional optimization and symptom reduction rather than aggressive excision.[10] These author argue that patients content with surgery, with lesser or none deficits are better and further justify that residual lesions might regress overtime. However, these regressing lesions are defined with respect to presentation inchildhood.[11] Complications of resection include infection, bleeding, hematoma, and postoperative seromas. Injury to facial, hypoglossal, glossopharyngeal, recurrent laryngeal, and lingual nerves has been reported.[1,3,12] Complete excision of a lymphangioma has been shown to have an 81% cure rate. When only part of the lymphatic malformation is excised, there is an 88% recurrence rate.[13] They state that non-encapsulated lesions recur more frequently because of their tendency to infiltrate major neurovascular structures.[14]

Sclerosing agents and radiation therapy have not been shown to play a role in the primary treatment of cystic hygromas. They may be indicated in lymphatic malformations that are macrocystic and not amenable to surgical resection.[6,15,16,17]Sclerotherapy with OK-432, a lyophilized, low-virulence SU strain of group A Streptococcus pyogenes, has been suggested as a possible therapy for macrocystic lesions. To date, however, the efficacy of OK-432 has not been proven in prospective, controlled trials.[17,18]

III. Conclusion

Lymphangomas presenting in adulthood are rare benign neoplasm. Though easy to diagnose clinically, it needs good radiological evaluation. Complete surgical excision is possible even in very extensive lesions. Great care must be taken during excision to avoid injury to vital structures in the neck especially the spinal accessory nerve.

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