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A Rare Case of Cervical Cystic Lymphangioma in a 32 Year Old Lady - The Case Report

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Abstract: ***Cystic lymphangioma (lymph cyst) is rare in adults. Most cases occur in childhood in the head and neck region due to the congenital obstruction of developing lymphatic vessels. Here we report a 32 year old female patient who came to our outpatient department with a left sided painless cervical swelling for 1 year duration. Ultrasonogram and CECT neck were done. Clinical and radiological findings were equivocal and the cyst was surgically excised. Pathologically, the cyst was found to be lined with cuboidal epithelium with lymphocytic infiltration. Hence, diagnosed as cystic lymphangioma. Being a rare pathology, with this article, we document cystic lymphangioma in an adult and highlight its importance in the differential diagnosis of cervical masses in adults.***

Keywords: Lymphangioma, Branchial cyst, Lymphocele, cervical masses, adults

1. Introduction

Cystic lymphangioma is a benign cystic lesion of the lymphatic vessels (1). Though it may be congenital or acquired, more than 90% cases are congenital affecting children less than 2 years of age (2). Owing to its rich lymphatics, cystic lymphangioma are more common in the head and neck region. Congenital lymphangiomas (cystic hygroma) are often associated with chromosomal abnormalities such as Turner syndrome. Acquired lymphangiomas may result from trauma, inflammation or lymhatic obstruction. Here, we report a rare case of cystic lymphangioma in a 32 year old female, initially misdiagnosed as branchial cyst.

2. Case Report

A 32 year old female presented to our OPD with chief complaints of painless swelling over the left side of neck for 1 year duration and progressively increasing in size. There was no previous history of trauma or surgery. There were no other symptoms suggestive of compression due to swelling such as dysphagia/ dyspnoea/ hoarseness of voice.

Examination Findings

On examination, Patient is conscious, well oriented. Vitals include pulse of 100 bpm, normal in rate, rhythm and volume. BP 90/60 mm of Hg in right brachial artery, supine position. So2 -99% on RA. RR- 28 cpm. General physical examination- no pallor, icterus, cyanosis, generalized lymphadenopathy, clubbing or pedal edema, no neurocutaneous markers noted and no bruit noted over the carotids and abdomen.

Systemic Examination

CVS- S1 S2 heard, no added sounds, no murmurs. JVP- normal, no signs of pulmonary arterial hypertension. RS- Trachea central, apex beat palpable (normal), bilateral chest movements symmetrical, bilateral air entry +, no added sounds. PA- soft, non tender, no organomegaly, no bruit. CNS: HMF- normal. No sensorimotor deficits. No signs of meningeal irritation noted.

Local Examination

On inspection, a swelling of size 6*5*2cm was noted in the left lower part of neck extending medially 1cm from the midline to 1cm lateral to the left sternocleidomastoid muscle, 5cm inferior to the angle of mandible till the superior border of clavicle, with smooth surface and no visible pulsations. On palpation, the swelling was cystic in consistency, transillumination was negative with no cervical lymphadenopathy.

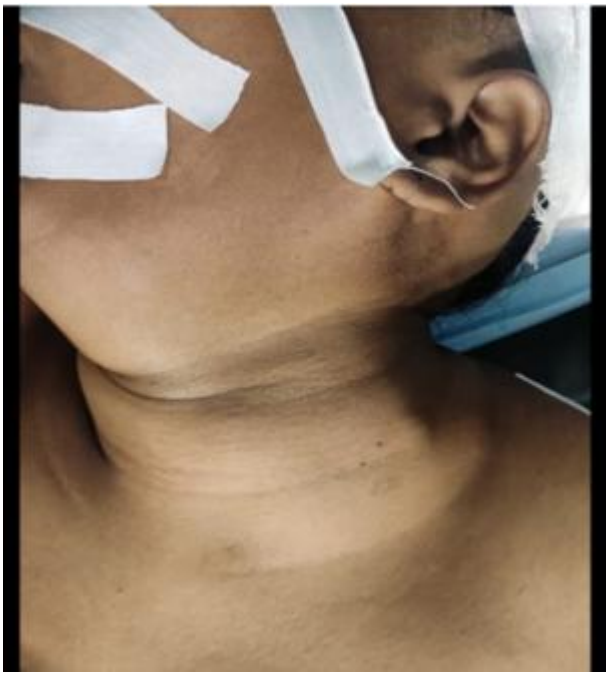

Figure 1: Preop photo of the patient showing swelling in the left lower side of neck

Radiological evaluation

USG Neck

Well defined cystic lesion of size 5.3*2.7*4.5cm in the left neck posterior to sternocleidomastoid with no solid components.

CT NECK

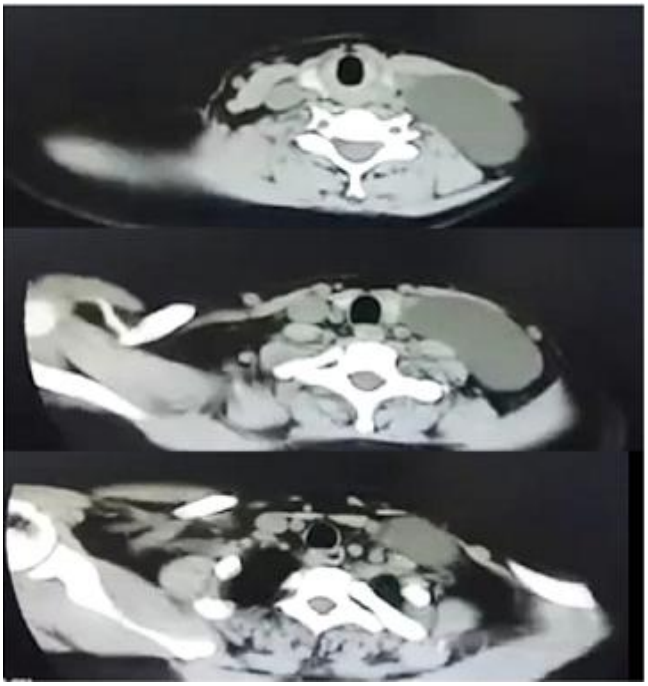

Figure 2: CT showing hypodense lesion in the left side of neck

Well defined hypodense swelling of size 6.4*3.2cm in the left side of neck lateral to carotid and posterior to sternocleidomastoid. Naso/oropharynx and larynx are normal.- possibility of left Branchial cyst

CT Contrast NECK

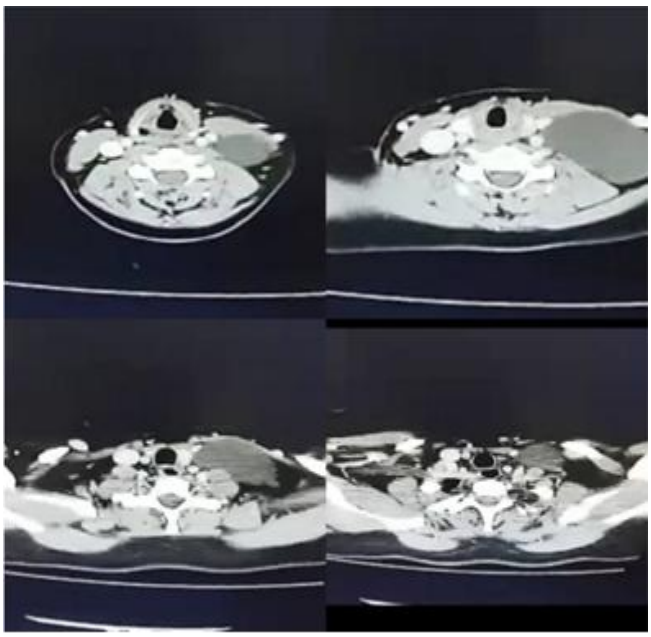

Figure 3: CECT Neck

A well defined cystic lesion with no internal septa / solid areas seen in the left side of neck, posterior and lateral to left sternocleidomastoid, splaying the jugular vein and common carotid artery, not enhancing with contrast, no lymph nodes, other structures normal- features suggestive of left branchial cyst.

Relevant blood investigations were sent

CBC		RFT	
TLC (cells/ dL)	7100	UREA	32
Hb (g/ dL)	9.6	Creatinine	0.7
Platelet count(cells/ dL)	237000		
ESR	28	TSH,T3, T4	Normal

Management

Based on the clinical examination, and radiological investigations, and considering the age, patient was tentatively diagnosed as a case of branchial cyst involving the left side of neck without any impingement on any vessels/ major structures in the neck and hence planned for surgical excision.

Intra-op findings

Deep to the deep cervical fascia, 6*6 cm swelling was noted, posterior and lateral to the left sternocleidomastoid and in close proximity to the left internal jugular vein. Carotid sheath was opened, the cyst was meticulously dissected from the adjacent structures and excised. Content- 75ml of chylous fluid. Suction DT was kept and wound closed in layers.

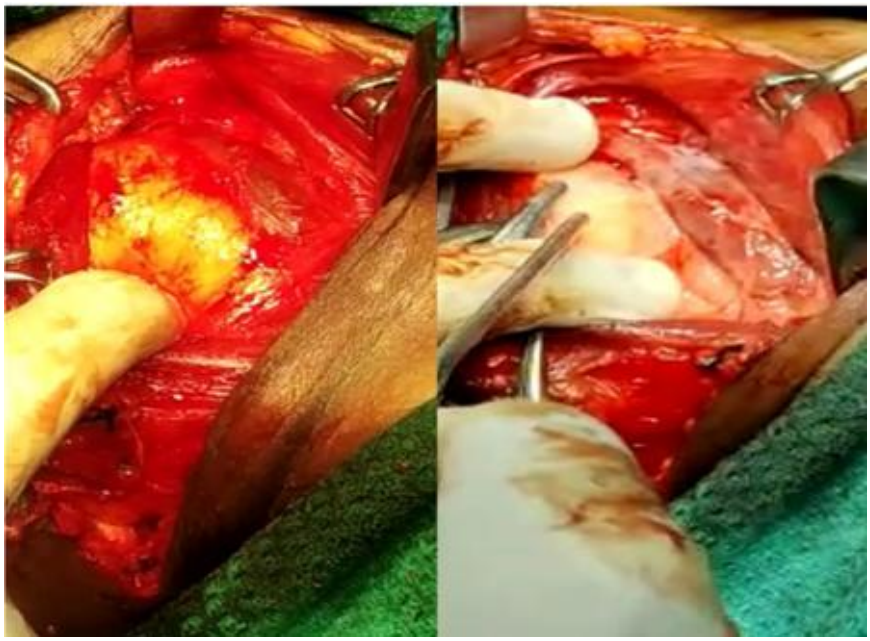

Figure 4 & 5: Intra-op findings of the cyst

Histopathological findings

Microscopic examination of the resected specimen shows fibrocollagenous cyst wall lined by flat cuboidal epithelium with lumen showing eosinophilic secretion filled with lymphocytes and wall showing lymphocytic and foamy histiocytic infiltration. Impression- features consistent with CYSTIC LYMPHANGIOMA.

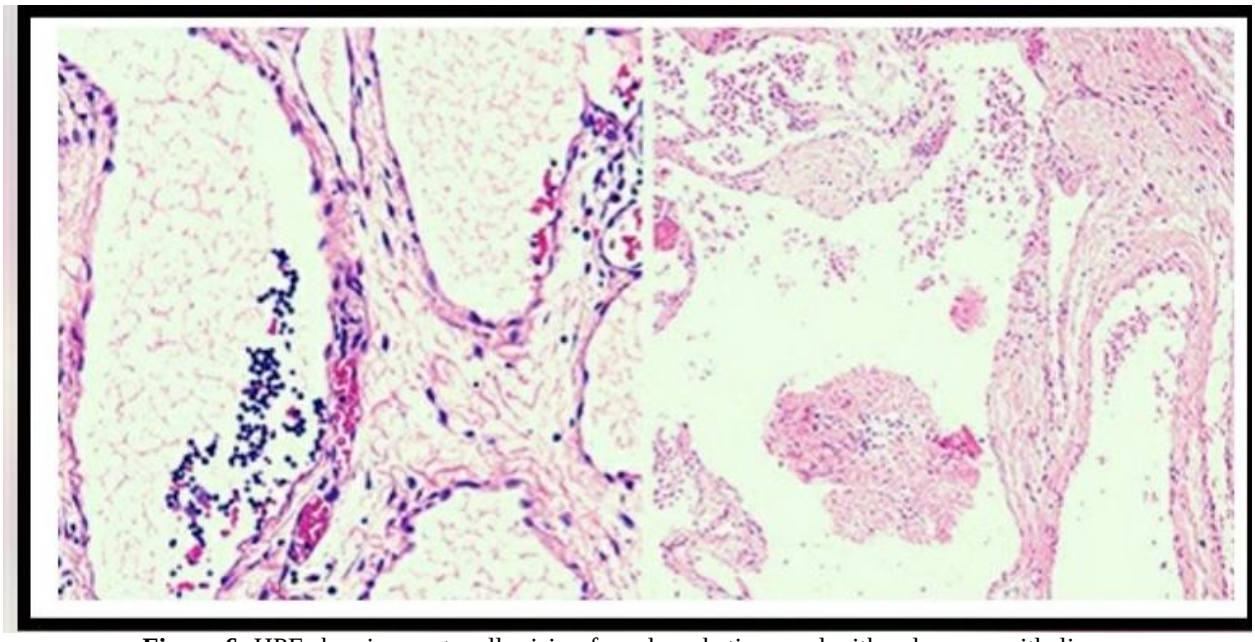

Figure 6: HPE showing cyst wall arising from lymphatic vessel with columnar epithelium

Postop management
Postop period was uneventful. DT was removed on POD-1 and the patient was discharged on POD-2

3. Discussion

Cystic lymphangioma also known as cystic hygroma is a benign congenital lymphatic malformation (1).It usually presents in the neonate or in early infancy involving parotid, submandibular,tongue and floor of mouth areas. They are brilliantly transilluminant. Cheek, axilla, groin and mediastinum are other less frequent sitea for a cystic hygroma (11).

Though rare, infection, trauma and neoplasm seem to the probable causes for cystic lymphangioma in adults. It has no gender predilection (1). Lymphatic malformations have been classified by various classification systems.Mc Gill et al. classified cystic lymphangioma based on CT scan, anatomy and histology- type 1 are macrocystic and below mylohyoid muscle whereas type 2 are microcystic and above mylohyoid muscle involving lips, tongue or oral cavity (6). Smith et al, classified lymphatic malformations as macrocystic (>2cm in diameter), microcystic and mixed variant.

Cystic lymphangioma usually present as asymptomatic mass lesion, however, huge cysts may produce symptoms due to compression of vital structures in the neck such as dyspnoea (trachea), dysphagia (oesophagus), hoarseness of voice (larynx).(1,5). Differential diagnoses of cystic lymphangioma include branchial cysts, lymphocele, salivary gland tumors, soft tissue sarcoma and thyroid masses.

Branchial cysts are the vestigial reamanants of second branchial cleft and seen in the upper neck in the early or late adulthood. They are cystic, transilluminate and lined by squamous epithelium(11).The case discussed in our article presented with cystic swelling in the lower neck and lined by columnar epithelium thus ruling out beanchial cysts.

Lymphocele is a localised collection of lymph without a distinct epithelial lining.It results from leakage of lymph into the soft tiissues or body cavity secondary to surgical disruption of lymphatic vessels. Clinically, lymphocele presents as a non-tender, non pulsatile lump(11). The case presented in our article had no previous history of surgeries and the cyst being lined by columnar epithelium, thus ruling out the possibility of a lymphocele.

Radiological evaluation includes USG, CT scan and MRI. Ultrasound remains the first line of investigation and shows anechoic, multilobulated, cystic mass. CT scan and MRI guide in a detailed evaluation when ultrasound findings remain equivocal. Role of Fine Needle Aspiration Cytology (FNAC) remains controversial (8).However, Owing to its rare presentation, histolopathological examination remains the cornerstone in the diagnosis of cystic lymphangioma in adults.

Complete surgical excision of the cyst remains the definitive treatment for cystic lymphangioma in adults. There is a reported recurrence rate of 10-15% if the cyst is not completely excised.

4. Conclusion

Cystic lymphangioma is extremely rare in adults. Surgical excision remains the primary treatment and definitive diagnosis can be done only by histopathology. Hence, cystic lymphangioma should be included in the differential diagnosis of neck mass with unusual presentation in adults.

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