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Cervical Lymphangioma In Adults: A Diagnostic Dilemma

Tej Pavan J[1]; Kiran M Naik[2]; Sumayya Fatima[3];
Jasoliya Arshwiben Arvindbhai[4]; Nishant Gupta[5]
(Department Of Otorhinolaryngology, Adichunchanagiri Institute Of Medical Sciences, B.G.Nagar, Mandya, Karnataka.)

Abstract:
Background: *Cervical lymphangioma in adults is a rare occurrence, with most cases seen in children under 2 years old. These benign lesions of the lymphatic system can be classified as simple, cavernous, or cystic hygromas. They may develop from the sequestration of lymphatic tissue from the primitive sac. Histopathological examination reveals massively dilated lymphatic spaces lined by epithelial cells and separated by connective tissue stroma with lymphoid aggregates.*
Case report: *A 36-year-old male presented with a swelling in the left submandibular region that gradually progressed to a size of about 5 x 7 cm over the span of 3 months. No history of trauma was reported, and the swelling was excised under general anaesthesia.*
Conclusion: *Due to their rarity, lymphangiomas in adults can be misdiagnosed, highlighting the importance of surgical intervention as the preferred treatment choice. The case report also underscores the diagnostic challenges posed by disparities between histopathological and radiological reports.*
Keywords: *cystic; neck swelling; adult cervical lymphangioma; diagnosis dilemma; surgical excision.*

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I. Introduction

Cervical lymphangioma in adults is a rare occurrence, with most cases seen in children under 2 years of age, with 2% of such cases occurring with chromosomal abnormalities (1,2). The incidence of lymphangiomas in infants has been documented to vary between 1.2 and 2.8 cases per 1000 new-borns (3). The primary sign or presentation of all lymphangiomas is the existence of a mass. They commonly present as posterior triangle masses (4). The posterior triangle is the most frequent site for the occurrence of inflammatory, metastatic adenopathy, or lymphoproliferative disorders. Furthermore, this region may exhibit congenital abnormalities such as branchial cysts, haemangiomas, and lymphangiomas, including cervical lymphangioma in adults (4). Hence, it is a dilemma for diagnosis. These benign lesions of the lymphatic system can be classified as simple, cavernous, or cystic hygromas. They may develop from the sequestration of lymphatic tissue from the primitive sac. Histopathological examination reveals massively dilated lymphatic spaces lined by epithelial cells and separated by connective tissue stroma with lymphoid aggregates (5). While various therapeutic approaches can be considered, surgical excision is still considered the best treatment option (1).

II. Case Report

A 36-year-old male presented with swelling in the left submandibular region, which was initially small in size and gradually progressed to a size of about 5 x 7 cm over the span of 3 months (Fig. 1). There was no history of trauma, shortness of breath, dysphagia, pain, or fever being reported. No personal or family history of significance in relation to the current disease was noted. On physical examination, the swelling was soft, cystic, non-tender, and mobile, extending below the angle of the mandible with no local rise in temperature, and did not move on deglutition.

Ultrasound of the neck revealed a well-defined cystic lesion with internal echoes; no vascularity or calcifications were noted, and it was reported as lymphocele. While the contrast enhanced computed tomography (CECT) of the neck reported a well-defined thin walled cystic lesion of fluid attenuation with thin internal septations of size 4.6 x 2.2 x 2.9 cm (AP x TR x CC) noted in the left submandibular space anterior to the left submandibular gland with no extension into adjacent neck spaces, there was no evidence of internal calcifications. A post-contrast study of the lesion shows mild enhancement of internal septations; it was reported as lymphangioma (Fig. 2). The Fine Needle Aspiration Cytology (FNAC) of the mass showed mucin background, foamy histiocytes, occasional signet cells, columnarization, and small clusters of epithelial cells

with mild atypia pointed towards low-grade mucoepidermoid carcinoma (MEC); but the absence of solid nests, sheets, or cords of epidermoid cells with mucous and intermediate cells steered the diagnosis away from MEC with another differential diagnosis as mucous retention cyst.

Intraoperatively, the swelling was noted in the submandibular space with extensive adhesions to the submandibular gland, creating a dilemma for its excision along with the mass. The swelling was excised after dissecting circumferentially and extracting in its entirety under general anaesthesia (Fig. 3). Transillumination of the surgical specimen revealed a thin-walled, cystic-like mass with serous content, and the tissue sample was sent for histopathological evaluation, which revealed cystic spaces containing lymph lined by flattened epithelial cells and the intervening stromal connective tissue containing lymphatic aggregates (Fig. 4). The report confirmed the diagnosis as lymphangioma.

III. Discussion

Lymphangiomas are anomalous lymphatic formations, predominantly emerge in the cervical region, while their presence in adults remains rare (1). This condition was first reported in 1828 by Redenbacher, and further study was carried out by Sabin in 1909 and 1912 (6,7). Cervical lymphangiomas are often unilateral and do not exhibit a gender or side preference in both juvenile and adult patients (8). These lesions are generally classified as lymphangioma simplex, cavernous lymphangioma, and cystic hygroma. A lymphangioma that arises in dense tissue, like the tongue, is known as a cavernous lymphangioma. However, when it forms in the loose fascia of the neck, it develops into a cystic lesion (1). The following three hypotheses have been put forward to explain its origins; The primitive lymph channels get blocked in their normal development during embryogenesis, the initial lymphatic sac fails to connect to the venous system, and in embryogenesis, lymphatic tissue is mislocated (1)(9). They are frequently confused with diverse entities like branchial, thymic, pericardial, cystic teratomas, and bronchogenic cysts. Lymphangiomas are generally asymptomatic and soft, but expansive growth can compress surrounding structures (8).

Fageeh et al. (10) suggested several methods for managing the condition, including observation, percutaneous drainage, the use of carbon dioxide lasers (CO2) and Nd-YAG lasers, diathermy, and surgical resection.

Miceli & Stewart (5) suggested that sclerotherapy with 1% or 3% sodium tetradecyl sulfate, doxycycline, ethanol, or radiation could be used as treatment alternatives. Patients under the age of 3 years with a lesion size of <4 cm and who do not have airway obstruction may be considered for observation, since they have the potential for spontaneous regression (4,10).

Surgical resection has been recommended as the most effective treatment approach. However, in situations where the lesions spread into the deep neck spaces, such as the floor of the mouth or parapharyngeal region, total removal may be challenging without causing harm to the neurological and circulatory structures. As a result of this condition, alternative methods such as drainage, sclerotherapy, and radiofrequency ablation are an alternative therapeutic approach for treating this condition.

Possible complications and their resulting morbidity may include infections, tissue necrosis, cranial nerve damage, arterial thrombosis, and even endocrine abnormalities.

This case presented a conspicuous cervical swelling devoid of trauma or infection history. Due to inconsistent radiological, FNAC, and clinical results, the case was clinically identified as a lymphatic cyst. Hence, histopathology proved to be useful in reaching a final diagnosis. With incongruent radio-clinical data and elusive radiological interpretation, the clinical diagnosis of such cases is found to be confusing in this age group.

Therapeutically, surgical resection prevails, while sclerotherapy and laser methods offer alternatives. Vigilance for recurrence, around 15%, demands judicious selection. Vigilant post-interventional monitoring ensures timely management, potentially through resection.

IV. Conclusion

Due to their rarity, lymphangiomas in adults can be misdiagnosed, highlighting the importance of surgical intervention as the preferred treatment choice. The case report also underscores the diagnostic challenges posed by disparities between histopathological and radiological reports.

Figures And Legends

Fig. 1: Left Submandibular Swelling. A) Lateral Aspect Of Neck; B) Anterior Aspect Of Neck.

Fig. 2: CECT Of The Neck Showing A Thin-Walled Cystic Lesion Of Size 4.6 X 2.2 X 2.9 Cm With No Extension Into Adjacent Neck Spaces. A) Axial View; B) Coronal View; C) Sagittal View.

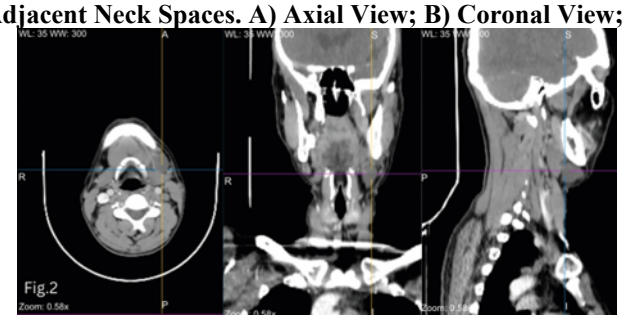


Fig. 3: Intraoperative Appearance

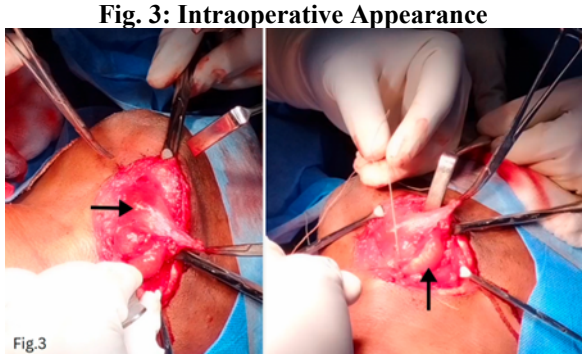
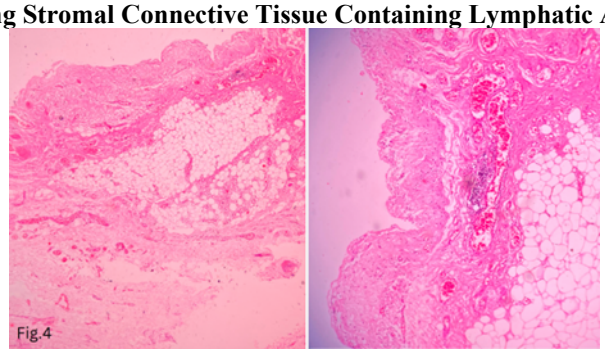


Fig. 4: HPE Showing Cystic Spaces Containing Lymph Lined By Flattened Epithelial Cells, The Intervening Stromal Connective Tissue Containing Lymphatic Aggregates



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