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Case Report

Cervical cystic lymphangioma in an adult: a rare entity

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ABSTRACT

Lymphangiomas are benign malformations of the lymphatic system, commonly arising from sequestered lymphatic vessels. While typically found in infancy or childhood, their occurrence in adults is rare. This case details a 44-year-old female with a progressively enlarging, painless neck mass. Imaging revealed a cystic lesion near major neck vessels. Fine-needle aspiration cytology initially suggested lymphocele, while ultrasonography and computed tomography angiography provided differential diagnoses, including lymphangioma. Surgical excision of the mass and histopathological examination confirmed the diagnosis of cavernous lymphangioma. Although benign, lymphangiomas can cause complications through invasion of surrounding structures, leading to mass effects or secondary infections. Definitive management is surgical excision, but thorough imaging and differential diagnosis are critical to ruling out other potential conditions, especially given the rarity of lymphangiomas in adulthood.

Keywords: Cystic lymphangioma, Lymphangioma, Cystic hygroma, Cavernous lymphangioma, Lymphatics, Lymphatic malformation

INTRODUCTION

Lymphangiomas are benign, localized malformations of the lymphatic system, arising from the sequestered lymphatic vessels.[1] They occur on the skin, the mucous membranes or even deeper tissues. Cervical cystic lymphangiomas arise due to a failed connection between the jugular lymphatic sac and the jugular vein.[2,3] They present as slow-growing masses in the lateral aspect of neck, usually not associated with pain, and may be easily mistaken for branchial cysts or abscesses or cervical lymph nodes.[4,5]

CASE REPORT

A 44-year-old female with no known comorbidities presented with a complaint of painless, progressively growing swelling on the left side of her neck for two months. She had no prior history of any infections, trauma or any surgical in the head and neck region. The swelling was in the left supraclavicular region, measuring 5×3 cm in size, soft in consistency, fluctuant, non-tender. The patient did not report any difficulty in breathing, change in voice, or dysphagia. The carotid pulsations were feebly palpable, masked under the swelling. The patient first underwent a fine needle aspiration cytology (FNAC) from the swelling at a private clinic, which was reported as lymphocele. Ultrasonography of the swelling reported a cystic, oval-shaped swelling having smooth, thin walls, lying just anterior to the left common carotid artery (CCA) and the left internal jugular vein (IJV). The differential diagnoses were likely to be a haemangioma, a lymphangioma, a venous aneurysm or a vascular malformation. The patient underwent a computed tomography angiography scan of the head and neck region, which showed a cystic lesion measuring 5×2.5×3.2 cm in size, abutting the left lobe of thyroid gland, and indenting

the left strap muscles, causing mass effect on the left IJV, lying 3 mm anterior to the left CCA.

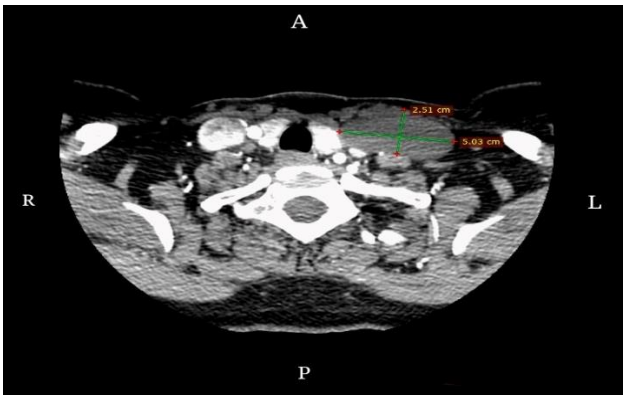

Figure 1: Axial section of CTA head and neck showing cystic lesion over the left supraclavicular region.

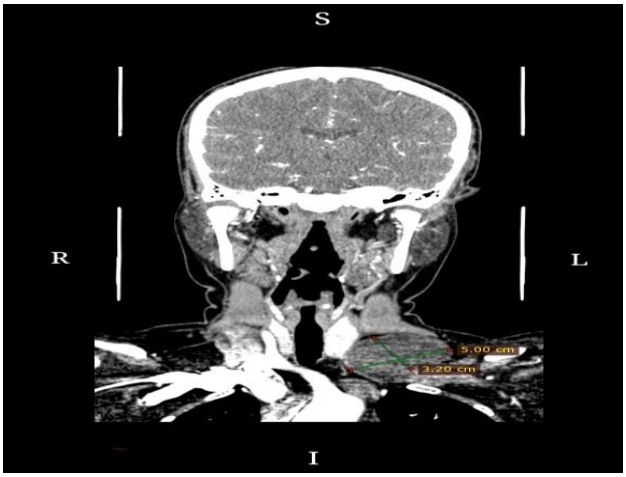

Figure 2: Coronal section of CTA head and neck showing cystic lesion over the left supraclavicular region.

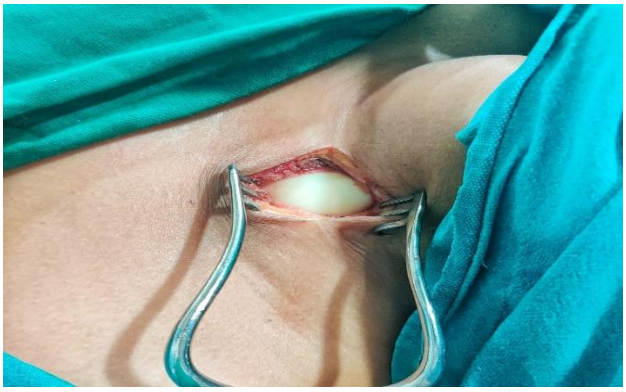

Figure 3: Intra-operative image showing a well-defined cystic lesion in the left supra-clavicular region.

The cyst was excised and sent for histopathological evaluation (HPE), which showed cyst wall lined by lymphocytes and lymphoid follicles and polymorphs, with foamy macrophages and few congest blood vessels – suggestive of a cystic lymphatic malformation, or a cavernous lymphangioma.

DISCUSSION

Lymphangiomas make up about 4% of all benign vascular tumours in infancy and early childhood, and their prevalence in adulthood is extremely rare.[5] They arise from aberrations in normal development of the lymphatic channels. Lymphangiomas are essentially benign tumours but are invasive and cause complications following infiltration into surrounding structures, like carotid or jugular vessels, larynx, and oesophagus. Patients usually present with complaints of a neck mass, which is slow growing in nature, associated with features of compression, like dyspnoea, stridor, dysphagia, change in voice, episodes of unconsciousness, and dizziness. They may also lead to secondary infection, haemorrhage, seroma formation or abscess formation.[6,7] Diagnosis is usually established by routine imaging techniques like ultrasonography or magnetic resonance imaging (MRI) or computed tomography (CT) scans. Radiologically, they appear as cystic swellings, having thin walls, arising from an underlying lymphatic channel, with features of invasion or abutting surrounding structures. One must always rule out other differential diagnoses, especially due to their rare occurrence in adults; before taking the patients up for surgery, which is the definitive management as described in literature in multiple case reports and series. Sharma et al described the occurrence of a massive cervical cystic lymphangioma in a 45-year-old female, managed surgically. In another case report, Qooz et al reported a case of a 24-year-old female with a cavernous type of a lymphangioma in the left supraclavicular region.[4,5] In a series of two patients from 2016, the authors talk of two young adults, a 23-year-old male and a 28-year-old female, both in the head and neck region. The first patient had a posterior triangle lymphangioma, while the second patient was diagnosed with a recurrent submandibular cystic lymphangioma. Wang et al published a case series of 7 adult patients over a period of 13 years (from 2008 to 2021), which only highlights the rarity of this malformation in adults.[7,8] In our case, we describe a 44-year-old female with a cavernous type of cervical cystic lymphangioma.

CONCLUSION

Cervical cystic lymphangiomas are a rare entity in adults. Surgeons must exhibit high degree of clinical suspicion with supportive radiological evaluation for diagnosis of lymphangioma in adults, as patients may present in later stages with features of invasion and compression. Management is via surgical excision of the cyst, and extreme care must be taken to not injure surrounding vascular structures. More comprehensive studies are required to study the phenomenon and effective management of the same.

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