



CYSTIC LYMPHANGIOMA IN ADULT-A RARE CASE REPORT

Pathology

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ABSTRACT

cystic lymphangioma (hygroma) is a benign congenital malformation of the lymphatic system that occurs due to obstruction or sequestration of developing lymphatic vessels. It is seen in infants and children who are less than two years of age, but rarely seen in adults. Occurrence in adults is rare and few cases are reported in literature. Here we are presenting a case report of 50 years old male reported in our patient department with a swelling in neck. Local examination and Ultrasound revealed a cystic mass in the lesion. Fine needle aspiration cytology was done and then total surgical excision of the mass was done. Histopathologically diagnosis of cystic lymphangioma was made. After an uneventful postoperative period patient was discharged. This case report discusses differential diagnosis, approach to the diagnosis and up-to-date management of cystic lymphangioma in adults. The objective of this case report is to discuss clinical presentation, pathological findings and diagnosis of this malformation.

KEYWORDS

Cystic Lymphangioma, Lymphatic System, Malformation, Cystic Mass

INTRODUCTION

Lymphangioma (cystic hygroma) is a benign congenital malformation of the lymphatic system that occurs as a result of obstruction or sequestration of developing lymphatic vessels^{4,5}. These lesions are rare in adults and only 150 cases are reported in literature so far^{6,7}. They are usually discovered in infants or children younger than two years of age. They are classified into septated (multiloculated) or non-septated (non-loculated) single cavity types. They are most frequently found in the neck (75%) and axilla (15%)⁸, while only 10% are found in the abdominal cavity and mediastinum^{9,10} including mesentery, retroperitoneal areas, and bones¹¹.

Cystic lesions of neck can be benign or malignant, are rare and thus difficult for clinician to interpret. Posterior triangle of neck is the most common location where lymphoproliferative diseases, inflammatory and metastatic adenopathies can occur. Other conditions like brachial cyst, lymphangiomas and hemangiomas can occur in this area¹.

hemangiomas and brachial cysts can easily be diagnosed in children who are below 2 years of age due to their clinical characteristics. These lesions manifest themselves as a mass which has no inflammatory changes and has a slow and painless growth². Treatment for lymphangioma is surgical excision but other conservative interventions like sclerotherapy can be used in adults if it is not an emergency³. As lymphangiomas are rare in adults we are presenting a case report of lymphangioma in adult and discuss about the clinical features, pathological findings and diagnosis of this malformation.

CASE REPORT

A 50 years old male presented in outdoor department with chief complaint of swelling over the upper right side of neck extending into the posterior triangle of neck (Figure 1 and 2) since one year. The swelling was associated with pain which was mild, dull aching, intermittent and non-radiating in nature. Past and family histories of patient were unremarkable. There was no history of dysphagia, dyspnoea, trauma, upper respiratory tract infection, fever, toothache, pus discharge from skin. Local examination revealed a single diffuse swelling over right side of neck near parotid region and angle of mouth, measuring 5.2×5.0, fluctuant in nature, not fixed to underlying structure.

The ultrasound of neck revealed cystic swelling measuring 4.5×4.4×1.5 cm in right side of neck just lateral to parotid region (figure 3). According to clinical and radiological findings differential diagnosis of brachial cyst, cystic teratoma, lipoma, lymphangioma were made.

Then patient was referred to pathology department for Fine needle aspiration cytology. On aspiration straw color fluid was obtained from

the affected area (approximately 15 ml). The swelling decreases in size considerably after aspiration. Both air dried and alcohol fixed smears were made from the aspirated material. Alcohol fixed smear was stained with Hematoxylin and eosin stain and air dried smear was stained with Giemsa stain. Microscopic examination of smears showed predominantly mature lymphocytes along with few inflammatory cells in a proteinaceous background (Figure 4 and 5). According to the location of the swelling, nature of the aspirated material and cytomorphology it was suggestive of cystic lymphangioma. Surgical excision was done and specimen (Figure 6) was sent to pathology department for histopathological examination. Slides were prepared from that specimen and stained with Hematoxylin and eosin stain and studied microscopically. Histopathology confirmed the diagnosis of cystic lymphangioma (Figure 7 and 8).

Figure 1 and 2:- Cystic swelling in posterior triangle of neck (after FNAC)

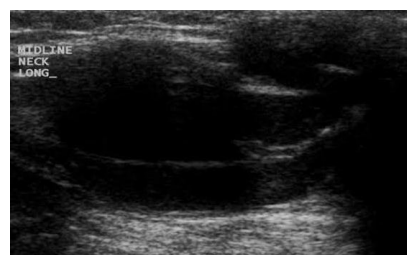


Figure 3:- ultrasound showing hypoechoic neck swelling

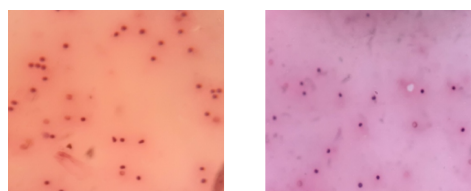


Figure 4 and 5:- aspiration cytology slides showing few mature

lymphocytes



Figure 6:- cystic specimen ,brownish in colour , cut in two parts , collectively measuring 3×1.5×0.6 cm

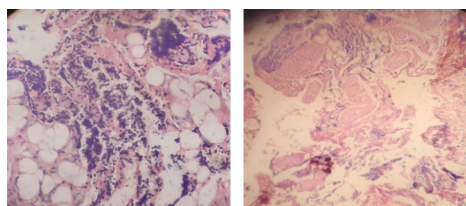


figure 7 (40x) and 8 (10x) H&E staining :- Histological features showing dilated lymphatic spaces lined by endothelial cells which are separated by connective tissues and contains lymphoid aggregates

DISCUSSION

Cystic lymphangioma are believed to be a congenital abnormality of the lymphatic system, there is a failure in the communication between lymphoid vessels and venous system which leads to accumulation of fluid. Most of them develop in-utero or in infancy therefore most cases are reported in pediatric age group. Most common presentation in adult is a painless lump, rapidly growing in size in an asymptomatic patient.

Thompson¹² in 2006 classify them into cystic (present most commonly in the neck), cavernous (located on tongue and mouth) or capillary (generally located on subcutaneous tissue). Colbert et al¹³ classified them into macrocysts, formerly known as cystic hygromas, and in microcysts, which may occur simultaneously in a lesion. Another classification done by Bhayya et al¹⁴ is lymphangioma simplex (composed of small thin-walled lymphatics), cavernous lymphangioma (comprised of dilated lymphatic vessels with surrounding adventitia), cystic lymphangioma (consisting of a large lesion, with macroscopic lymphatic spaces surrounded by fibrovascular tissue and smooth muscle) and benign lymphangioma (lymphatic channels that seem to be dissected through dense collagen bundles).

Microscopically, lymphangioma is characterized by large, dilated lymphatic vessels in a fibrotic or loose stromal background. Lymphoid aggregates when exist in large amount may be confusing and needs to be differentiated from atypical lymphoid proliferation. The main histopathologic differential diagnosis of cystic hygroma is cavernous hemangioma in which blood filled large cystic spaces were described to be similar to lymphangioma. Cavernous hemangioma lining endothelium shows positivity with Factor VIII, and pancytokeratin on immunohistochemistry. The presence of lymphatic spaces with thin walls containing fibrous tissue and lymphoid aggregates suggest the diagnosis of lymphangioma.

Surgical resection is advocated as the best treatment option, but where the lesions extend into deep neck spaces, as the floor of the mouth or parapharyngeal space, complete removal may be difficult and can cause damage to the nearby nervous and vascular structures. Thus alternatives such as drainage and sclerotherapy with different substances such as tetracycline, bleomycin and triamcinolone can be used in such cases. Radiofrequency ablation and interferon alpha are another therapeutic option to treat with^{13,16}. Sclerotherapy with OK-432—a strain of Group A *Streptococcus* can be used. It produces an inflammatory reaction when applied with intracystic injection and then causes the destruction of endothelium, sclerosis, and cicatricial contraction of cyst wall. Complications like infections, tissue necrosis, cranial nerve lesions, vascular thrombosis and endocrine disorders can occur¹⁵. Complication with OK-432 are fever and local inflammation^{16,17,18}, hypopharyngeal edema was reported when it was administered in cysts located near the airway¹⁹. Complication with

laser or ablation therapy is damage to overlying skin. Interferon alpha causes complication like fever, diarrhea and neutropenia²⁰.

Imaging studies such as ultrasound, computed tomography and MRI are important to assess extension of the lesion. Although ultrasound scan is sufficient to establish the diagnosis, computed tomography (CT) or MRI are useful to show extension to adjacent tissue.

This case was unusual as cystic lymphangioma (hygroma) presented in an adult with no history of trauma or respiratory airway infection. This causes a diagnostic challenge due to rarity of this lesion in adult cases. Complete surgical excision was done which was successful and prognosis is good.

CONCLUSION

Cystic lymphangioma in cervical region are rare, benign malformation which can easily be diagnosed. Clinical suspicion of lymphangioma helps in management of patient and radiological imaging with ultrasound, computed tomography and MRI are helpful in finding the extent of the lesion in adjacent structure. Final diagnosis can be made by pathology. Surgical excision is the treatment of choice but cases where surgery is not possible other alternative therapies can be used, most commonly is sclerotherapy.

CONFLICT OF INTEREST

None stated by authors

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