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CYSTIC HYGROMA IN ADULTS - A RARE CASE REPORT

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ABSTRACT: Cystic hygroma (lymphangioma) is a benign congenital malformation of the lymphatic system that occurs in infants and children younger than two years of age. It is rarely seen in adults. Occurrence in adults is uncommon, and only few cases of adult lymphangioma have been reported in the literature. Here we present a rare case of cystic hygroma in a 52 year old patient who presented with a swelling in the neck. Computed tomography revealed presence of cystic masses in the lesion. After total excision of the mass, histopathological diagnosis of cystic lymphangioma was made. After an uneventful postoperative period patient was discharged. No recurrence has occurred since then. This case report discusses the differential diagnoses and up-to-date management of cystic hygroma in adults. The objectives of this report are to discuss about the clinical presentation, diagnosis, pathologic findings and management of this malformation.

KEYWORDS: Cystic hygroma, lymphangioma, lymphatic system, congenital malformation.

INTRODUCTION: Cystic hygroma (lymphangioma) is a benign congenital malformation of the lymphatic system that occurs as a result of sequestration or obstruction of developing lymphatic vessels.^{1,2} These lesions are usually discovered in infant or children younger than two years of age. Occurrence in adults is uncommon, and fewer than 150 cases of adult lymphangioma have been reported in the literature.³⁻⁵

Cystic hygroma can be classified into septated (multiloculated) or non-septated single cavity (non-loculated) types. Presentation in adulthood is rare and the cause is uncertain, although trauma and upper respiratory tract infection have both been suggested as possible triggers for onset.^{6,7}

Diagnosis in adults is considered to present a greater challenge than in children and initial misdiagnosis, frequently as branchial cleft cysts as in the case reported here, is common.^{6,8} Definitive diagnosis is usually based on post-operative histological findings.

Here we present a case of cystic hygroma in an adult patient and discuss the management options for such a presentation.

CASE REPORT: A 52-year-old woman was referred to our clinic with history of a slowly enlarging mass on the left side of her neck (Figure 1) since last ten months. She noted discomfort and mild pain while turning her head to the left side. She was further asymptomatic. Physical examination revealed a soft, smooth and mobile mass measuring about 10 cm, in the left supraclavicular fossa and left lateral side of neck. In fine needle aspiration cytology (FNAC), yellowish fluid was aspirated and showed the presence of mature lymphocytes and histiocytes. Computed tomography of the neck revealed presence of a lobulated cystic mass extending from the root of neck to the upper part of lateral neck, deep to the left sternomastoid and lateral to the carotid and jugular vessels (Figure 2). Excision of the mass was performed under general anesthesia. Histopathological diagnosis was cystic lymphangioma which was supported with massively dilated lymphatic spaces lined by endothelial

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cells and separated by intervening connective tissue containing lymphoid aggregates (Figure 3). Follow-up at 12, 24 and 36 months showed no recurrence.

DISCUSSION: Cystic hygromas believed to arise from a congenital malformation of the lymphatic system. Failure of communication between the lymphatic and the venous pathways leads to accumulation of lymph. Most cystic hygromas present in-utero or in infancy. Therefore most of the literatures on management consider pediatric cases. The effect of these lesions depends on their position and relationship to surrounding structures. Although the most common adult presentation is of a painless lump in an otherwise asymptomatic patient⁶, rapid enlargement over a short period of time has frequently been reported⁸ and major structures such as the larynx, trachea, esophagus, brachial plexus and great vessels have known to be compressed or incorporated within the lesion⁹. In this case the lesion had doubled in size over a period of 4 months and had caused a restriction of neck movement; others have reported presentation with pain, hoarseness, dysphagia and breathlessness.^{1, 9, 10}

Lymphangiomas are best visualized by magnetic resonance imaging (MRI). The high water content allows lymphangiomas to appear hyperintense on T2-weighted Images.¹⁰ The other imaging methods are Doppler ultrasonography and computed tomography (CT). Complete surgical excision has traditionally been considered the treatment of choice for cystic hygroma.¹¹⁻¹⁴ However, several authors have suggested that sclerotherapy (with alcohol, bleomycin and OK-432) may be a more appropriate first-line therapy.^{15, 16} Intravenous cyclophosphamide has also been used, with some success, in recurrent lesions following surgery.¹⁷

This case was unusual in that a large cervical cystic hygroma presented de-novo in an adult with no history of trauma or upper respiratory infection. This presented a diagnostic challenge due to the rarity of the lesion in adults. Complete excision was successful and the prognosis is good. The authors would like to emphasize the importance of pre-operative imaging and surgical excision in the management of these lesions.

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Fig. 1: Neck swelling in left supraclavicular space

Fig. 2: CT SCAN: 8x9x10 cm lobulated cystic mass deep to the right sternomastoid and lateral to the carotid and jugular vessels

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Fig. 3: Histological findings: Dilated lymphatic spaces lined by endothelial cells and separated by intervening connective tissue containing lymphoid aggregates

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