

Rare Case of Adult Cystic Hygroma of Neck

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Abstract: Cystic hygroma is a congenital malformation of the lymphatic system that occurs in children less than 2 years of age. Although it is well recognised in paediatric practice, it seldom presents de novo in adults. These are commonly present in head and neck but can occur anywhere. Cystic hygroma is very rare in adults, but it should be considered in the differential diagnosis of adult neck swellings. Patients presenting with a painless, soft, fluctuant and enlarging neck mass should have a careful history and physical examination along with radiological imaging to help in diagnosis. Here, we are reporting a case of cystic hygroma in a 42-year-old female patient in the neck region.

Keywords: Adult, Cystic hygroma, Lymphangioma, Neck.

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I. Introduction

Cystic Hygromas or lymphangiograms are originally reported by Redenbacker in 1828 and the name “cystic hygroma” was coined by Werner in 1834[1]. It can occur in the head, neck, axilla, cervico-facial regions, groin and below the tongue[2]. The exact aetiology of lymphangioma in adults is still debated. According to literature, only 150 cases of adult lymphangioma have been reported[3]. Cystic Hygromas are classified into septated or non-septated single cavity types. It always poses a diagnostic challenge in both children and adults and most of the time it is based on post-operative histological findings[3].

II. Case Report

A 42-year-old female patient known hypertensive, presented with the complaint of a painless, progressively enlarging mass in the neck for two years. The mass was initially smaller in size, then gradually enlarged to occupy the whole neck on the left side. She had some discomfort on moving her neck on left side. She did not have any obstructive features or signs of malignancy. There was no history of trauma in the neck region or recent upper respiratory tract infections. On examination, a soft, fluctuant, non-tender, trans-illuminant, well-defined mass measuring approximately 14x10cm could be palpated on the left side of the neck extending from beneath the ear lobe to the supra-clavicular fossa in the vertical plane, and from the para-tracheal region to the posterior triangle of the neck in the horizontal plane(**Figure 1**). There was no thrill, bruit or cervical lymphadenopathy and the remaining neck and systemic examination was normal.



Figure 1 - showing large, soft, fluctuant swelling occupying the left side of neck

Ultrasound of neck revealed an multilocular, cystic anechoic lesion in the postero-lateral region of the neck, superficial to major neck vessels. FNAC revealed straw-coloured fluid showing lymphocytes suggesting of chronic inflammatory exudate. CECT scan of the neck and thorax showed a large loculated benign cystic mass lesion with few enhancing septa within. It occupied the posterior cervical space on the left side of the neck lateral to the carotid artery and displacing the left internal jugular vein antero-medially. There was no intra thoracic extension. A diagnosis of cystic hygroma was made and the patient was prepared for surgical excision.

Under general anaesthesia, through a transverse incision was made over the swelling, subplatysmal flaps were raised and the sternocleidomastoid muscle was divided to expose the multiloculated cystic lesion throughout its extent. The spinal accessory nerve lying on the anterior aspect of the lesion and the internal jugular vein were carefully preserved, and the cystic lesion was excised intoto (**Figure 2**).



Figure 2 - intra operative image showing the large multiloculated, cystic swelling being

Histopathology revealed thin connective-tissue stroma separating the cystically dilated spaces lined by a single layer of benign endothelial cells which was consistent with cystic hygroma.

III. Discussion

Acquired cystic hygroma is one of the unusual differential diagnosis of cystic neck lesions in adults. It is an aberrant proliferation of lymphatic vessels resulting from abnormal development of the lymphatic system[4]. It is often misdiagnosed with other cystic neck swellings like branchial cleft cyst, dermoid cyst, lipomas, hemangiomas or thyroid mass[5]. Most of the cases present with painless enlargement of neck mass. However, Kalsotra et al[6], reported a massive enlargement of lymphangioma in an adult causing compression of major structures of neck like larynx, trachea, oesophagus and brachial plexus. Certain patients may have intra thoracic extension of lymphangioma, where prior investigations like computed tomography/ magnetic resonance imaging (MRI) is needed for proper pre-operative evaluation and decision making. There are three histological subtypes of cystic hygroma- capillary lymphangioma (composed of small lymphatics), cavernous lymphangioma (composed of large lymphatics), cystic lymphangioma (composed of large lymphatics spaces with collagen and smooth muscle)[7].

Surgical excision has been the standard treatment of choice. However, certain studies show that sclerotherapy (with alcohol, bleomycin and OK-432) may be a more appropriate as first-line therapy of lymphangioma. In recurrent lesions, intravenous chemotherapy have also been tried with some success[8].

IV. Conclusion

Cystic hygromas are a rare differential diagnosis in adult neck masses and should be kept in mind for the adult patients who present with neck, axillary, mediastinal or retroperitoneal masses. Preoperative imaging for diagnosis and to look for intrathoracic extension is essential. Surgical excision is the treatment of choice. The paucity of literature and atypical presentation pose a diagnostic challenge due to rarity of this lesion.

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