ACCSCIENCE PUBLISHING

Tumor Discovery

CASE REPORT

Cystic hygroma in a young adult: A case report and recent management

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Abstract

We are reporting a case of a 27-year-old young female who presented with right side neck swelling without any associated obstructive symptoms and any other grave signs and symptoms. She noticed a gradual increase in the size of the swelling within a period of 2 years. After investigation and surgical excision, the swelling was diagnosed as cystic hygroma. The root cause of the development of cervical lymphangioma is the congenital malformation of the developing lymphatic system. Cystic hygroma is benign in nature and the cause in adults is still unclear. The most common site of origin is in head and neck region, and cystic hygroma accounts for 75% of lymphatic malformations. The most common presentation of cystic hygroma is painless swelling with ill-defined lesion, most commonly located at the posterior triangle of the neck. The common age group is between birth and 2 years of age, with very rare presentation in adults. Hence, it is necessary to rule out all differential diagnosis of cervical lymphangioma, which is presented with cystic neck swelling. Complete surgical excision is the recommended standard treatment.

Keywords: Cystic hygroma; Young adult; Cervical lymphangioma; Congenital lymphatic malformations

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1. Introduction

The incidence of cervical lymphangioma (cystic hygroma) in adults is very rare, and very few cases have been reported in the literature[1,2]. Lymphangiomas are congenital malformations of the developing lymphatic system, and these conditions are benign in nature. The most common cause of lymphangioma development is an obstruction or sequestration of the developing lymphatic vessels[3,4]. The cause of cystic hygroma in adults is not known; however, upper respiratory tract infection and trauma have been reported in the literature[5,6]. Diagnosis of cystic hygroma in adults is difficult, and the definitive diagnosis is purely based on the final histopathological examination. The most common location of cystic hygroma is at the posterior triangle of the neck, while the most common pathologies are inflammatory, metastatic adenopathies or lymphoproliferative diseases. Under 2 years of age, the common pathologies of cystic hygroma are branchial cysts, hemangiomas, and lymphangiomas[7]. They are characterized as slow-growing tumors, and chances of spontaneous regression are

very rare[8]. Surgical excision is the choice of treatment as described in most literature[9].

2. Case presentation

A young female of age 27 years with no co-morbidity with Eastern Cooperative Oncology Group Performance Status 1 approached our clinic and complained of right side neck swelling. There was no supportive family, medical and surgical history. The patient has a history of swelling on the right side of the neck around 2 years ago, and the swelling gradually grew (in size) over the years. There was mild pain with the movements of the neck. There was no associated history of trauma, difficulty in swallowing, and previous procedure. During clinical examination, there was mobile and fluctuating large swelling of approximately 10 cm × 9 cm in size, which was present in the right posterior triangle of the neck without neck lymphadenopathy. She had been evaluated outside of our clinic with contrast-enhanced computed tomography (CECT) of the neck and fine needle aspiration cytology (FNAC), and we also had advised her to undergo computed tomography (CT) of the chest. The finding of CECT was suggestive of a multiseptated cystic lesion of 10.5 cm × 9.5 cm × 8 cm in the posterior triangle of right side of the neck with preservation of all fat planes, as shown in Figures 1 and 2. Meanwhile, the finding of FNAC was suggestive of lymphangioma, which was probably cystic hygroma. The patient was advised to undergo surgery, and she was treated with wide local excision with intact capsule (Figures 3 and 4). The post-operative course was uneventful, and she was discharged on the 5th post-operative day. The final histopathology report confirmed that the cystic lesion was a cystic hygroma. Recurrence was not reported by the patient even after 1 year of completion of treatment.

3. Discussion

The incidence of lymphangiomas is in the range of 1.2 – 1.8/1000 of new births[10] or 1 in 2000 – 4000 live births[11], as reported in different studies. It has been found that in 90% of reported cases, the lesion occurs commonly in the age range between birth and 2 years[12]. The most common site or origin of lymphangiomas is the head and neck region[13]. The other reported sites of lymphangiomas with the lower incidence rate are retroperitoneum, axilla, pelvis, and mediastinum[14,15]. Out of all head and neck lymphatic malformations, cystic hygroma accounts for the majority with an accountability of 75% in the head and neck region[16]. The causes of lymphatic malformations are still unclear; however, some of the etiologies have been reported like misplacement of lymphatic channels during embryogenesis, arrest of lymphatic growth, and failure of lymphatic system to reach the venous drainage[17]. In addition, an association has been found between cystic hygroma and other conditions such as chromosome aneuploidies, hydrops fetalis, and intrauterine death[18].

The most common presentation of cystic hygroma is a painless and ill-defined swelling or mass. It never involves skin and it shows positive transillumination test. The common location of cystic hygroma is in the anterior and posterior triangles of the neck, with the posterior triangle of the neck being the most common site of occurrence[19]. Some of the cystic hygromas present with large neck masses with obstructive symptoms such as dysphagia or adult respiratory distress if they are located in the suprahyoid region, and these lesions are associated with a higher rate of recurrence, complications, and morbidity[20]. Incomplete surgical resection, midline location, and multiple lesions are in favor of higher recurrence rate. Hence, complete

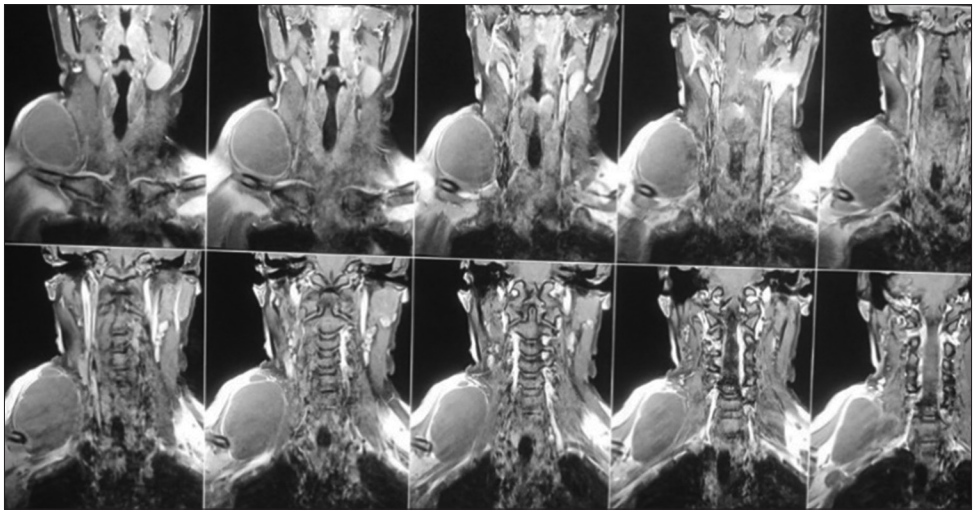

Figure 1. Coronal view of contrast-enhanced computed tomography.

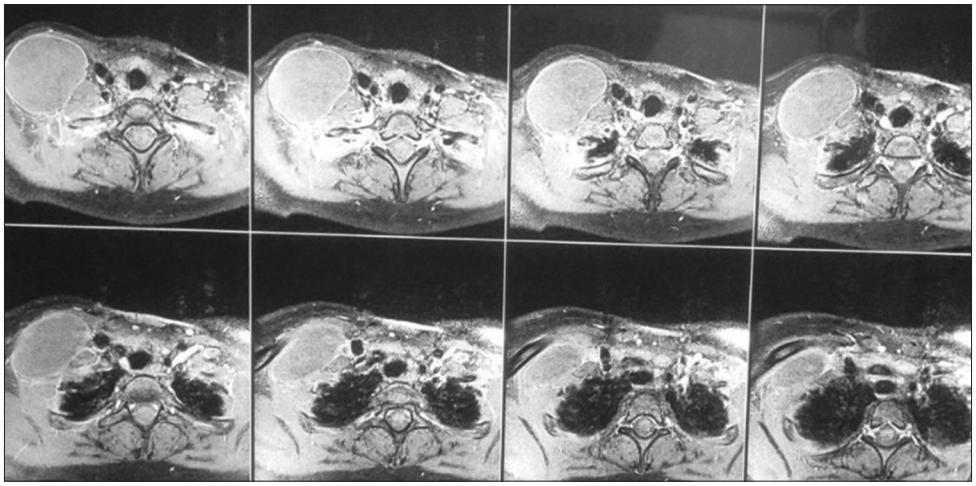

Figure 2. Axial view of contrast-enhanced computed tomography.

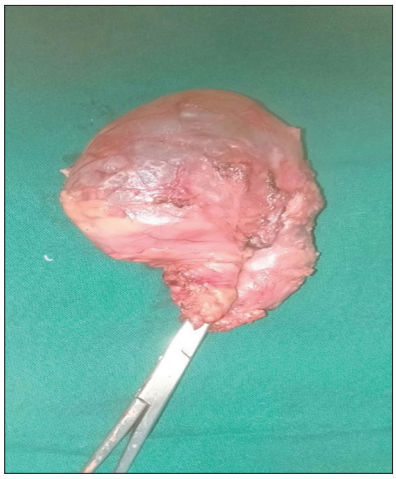

Figure 3. Specimen resected *en bloc*.

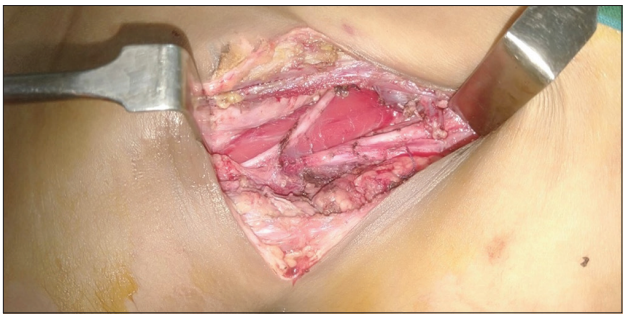

Figure 4. Post-resection.

surgical excision with intact capsule is mandatory to avoid future complications.

The choice of radiographic investigations is ultrasound, CT, and magnetic resonance imaging (MRI). CT defines the extent of the lesion with characteristics of the inner content of the cyst, while MRI helps in defining the relationship of the cystic lesion with surrounding soft tissues[21]. In most of the cases, radiographic investigations are enough for diagnostic purposes. Tissue diagnosis before surgery is only indicated if there is a dilemma in diagnosis to differentiate lymphangiomas from neck sarcoma, lymph node mass, lymphoma or other benign neck tumors. If obstructive symptoms are present like respiratory distress, prior tracheostomy is essential to maintain airway track. Classification based on anatomical location has been published in 1995 by de Serres *et al.*[22] (Table 1).

Different treatment modalities have been proposed for the treatment of cystic hygromas. In individuals with age <3 years and lesion size <4 cm, observation is a treatment option as there are chances of spontaneous regression[22,23]. The next proposed options are sclerotherapy with doxycycline or radiotherapy, which were recommended by Miceli and Stewart[24]. The other non-surgical options are percutaneous drainage, carbon dioxide laser, Nd-YAG laser, and diathermy which were proposed by Fageeh *et al.*[23] Previously sclerosing agents were used for the treatment, including boiling water, quinine, sodium morrhuate, urethane, iodine, doxycycline, and nitromin; however, sclerosing agents have been found to cause more complications with a low success rate in treatment[25-27]. Several case reports have been published establishing the role of bleomycin as primary intra-lesional sclerosing agent for the treatment of cystic hygroma[28,29]. Aspiration of cystic hygroma is one of the temporary treatment options, which helps in reducing the size of the hygroma and thereby reduces the pressure effects on the respiratory and feeding tract[30,31]. The surgical resection of the cystic hygroma is a traditionally accepted, standard treatment. However, when the lesion extends into the floor of mouth, parapharyngeal spaces or deep neck spaces, complete removal of the lesion will be a difficult task. In these

Table 1. Classification of lymphatic malformations based on anatomical location

Class	Description
Stage I	Unilateral Infrahyoid lesion
Stage II	Unilateral Suprahyoid lesion
Stage III	Unilateral Suprahyoid and Infrahyoid lesion
Stage IV	Bilateral Suprahyoid lesion
Stage V	Bilateral Suprahyoid and Infrahyoid lesion

cases, alternative procedures, such as sclerotherapy with tetracycline, bleomycin, and triamcinolone or drainage, have been recommended. The next therapeutic treatment option is radiofrequency ablation[21]. OK-432 (Picibanil), a sclerosing agent recommended by Ogita *et al.*[32], was prepared by incubating streptococcal pyogenes with penicillin[33,34]. It is used to perform sclerosis in cystic lesions of the neck as it has a property of inducing fibrosis secondary to inflammatory and cicatricle changes with the consequent contraction of the lymphangioma.

4. Conclusion

Although cystic hygroma is rare in adults, differential diagnosis among all cervical lymphangiomas is necessary. Surgical excision is the gold standard for the treatment of cystic hygroma, except in complex cases, while histopathology is the definitive diagnostic modality.

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Conflict of interest

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Ethics approval and consent to participate

Informed consent was obtained from the patient for being included in this study.

Consent for publication

Informed consent to publish this case was obtained from the patient.

Availability of data

Not applicable.

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