

Case Report

Adult-onset cystic hygroma: A case report of rare entity

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Abstract

Cystic hygroma is a benign congenital malformation of the lymphatic system that occurs in infant or children younger than 2 years of age. Although cystic hygroma is well recognized in pediatric practice, it seldom presents *de novo* in adulthood. These are commonly present in head and neck but can be present 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 assist with diagnosis. Surgical intervention is the treatment of choice for this rare condition. Here, we are reporting a case of cystic hygroma in a 32-year-old male patient in the neck region. The objectives of this case report are to discuss the clinical presentation, diagnosis, histopathological findings and management of this malformation.

Key words: Cystic hygroma, lymphatic system, neck masses, surgical excision

INTRODUCTION

Cystic hygroma also known as water-tumor or lymphangioma is a benign malformation of lymphatic vessels which usually occurs when the lymphatic system fails to communicate with the normal jugular vein. It can occur in the head, neck, axilla, cervico-facial regions, groin, and below the tongue.^[1] Lymphangioma, soft tissue tumor of disputed pathogenesis was originally reported by Reden Backer in 1828 and "cystic hygroma" name was first given by Wernker in 1834.^[2] Lymphangioma is generally known as a disease of childhood when there is active lymphatic growth. Cystic Hygromas are single or multiple cysts found mostly in the neck region. A cystic hygroma can be present as a birth defect (congenital) or develop at any time during a person's life^[3]

They are locally aggressive, benign lesions that are difficult to manage due to recurrence of the tumor following surgery with a recurrence rate of 21%.^[1] Cystic hygroma is usually histologically characterized by the proliferation of small lymphatic vessels with

intervening fibrous tissue.^[1] 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 the onset.^[4] Most commonly these malformations occur in the head and neck, although they have been described in a variety of other anatomical locations. Until date, there have been fewer than 150 reports of adult cervicofacial cystic hygroma in the English language literature and the optimum management of these lesions is still a matter of debate.^[4] Diagnosis in adults is considered to present a greater challenge than in children, and final diagnosis is usually based on postoperative histology.^[4]

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

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CASE REPORT

A 32-year-old male patient reported with a chief complaint of pain and swelling over the right lower third region of face extending into the neck until [Figure 1a and b] the midline since 8 months. The swelling was associated with pain that was mild, dull aching, continuous and nonradiating in nature. There was no history of fever, trauma, toothache, pus discharge, but there was a history of restricted mouth opening since 1-month. On extraoral examination and palpation [Figure 1b], a single diffuse swelling was present over right submandibular and a submental region extending into neck till midline that was soft, tender, and fluctuant in nature. On intraoral examination [Figure 1c], no abnormal finding was seen, occlusion was normal. On aspiration light brown colored and stingy fluid was obtained from the affected area. The computed tomography axial contrast [Figure 2a] showed a well-defined lobulated, hypodense cystic lesion of size (27.2 mm × 45.9 mm) present on right side in the neck of the midline just below the submandibular gland and compressing the same. Postcontrast axial computed tomography [Figure 2b] shows no enhancement. Radiologically features were suggestive of a cystic lesion in the right side of the neck. Based on clinical and radiographic findings, provisional diagnosis of plunging ranula of right side of neck was made with differential diagnosis of enlarged submandibular and submental lymph nodes, branchial cyst, dermoid cyst, lipoma, thyroglossal cyst, subhyoid bursitis, and lymphangioma.

The patient was taken under general anesthesia and Risdon's incision placed in the right submandibular region. The lesion was excised in toto [Figure 3a]. Macroscopically biopsy specimen [Figure 3b] was multiloculated measuring 4.8 cm × 1.8 cm × 1.5 cm. The biopsy specimen was soft in consistency with irregular borders, color was creamish pink. Microscopically specimen showed [Figure 4a] cystic lumen formed by cystically dilated vascular lymphatic channels containing pinkish eosinophilic proteinaceous lymphatic fluid along with lymphoplasmacytic cell infiltration. Cystic lumen was lined by single flattened endothelium layer. The connective tissue wall [Figure 4b] was loosely fibroblastic with engorged capillaries, adipocytes and extravasated red blood cells. The final diagnosis correlating with clinical and radiological findings was made as adult-onset cystic hygroma. Postoperatively patient recovered well [Figure 5] without neurological symptoms. The patient is on regular follow-up of 1, 3, and 6 months.

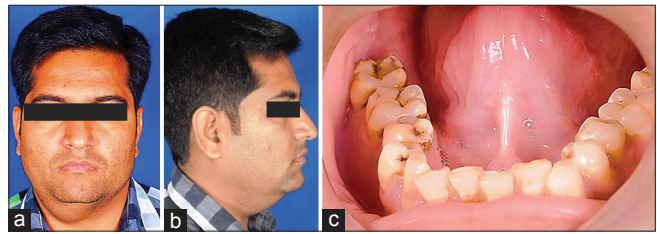


Figure 1: (a) Diffuse swelling present on right submandibular and submental region extending into the neck till the midline. (b) Diffuse swelling present on right submandibular and submental region. (c) Intraorally no abnormal findings were seen

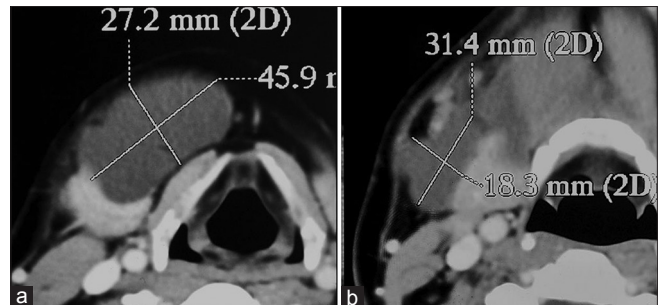


Figure 2: (a) Computed tomography axial contrast is showing well defined, lobulated, hypodense cystic lesion present on right side in neck off the midline below the submandibular gland and compressing the same. (b) Postcontrast axial computed tomography shows no enhancement

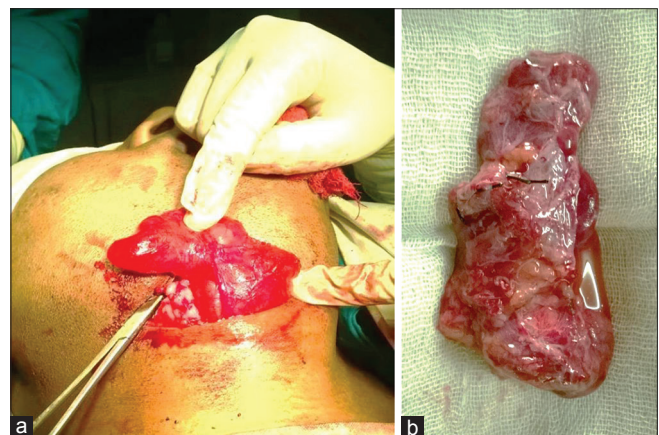


Figure 3: (a) Cystic lobulated swelling present in the neck region. (b) Surgically excised specimen

DISCUSSION

Lymphangioma is a benign hamartomatous tumor of lymphatic channels, with the marked predilection of head and neck region.^[5] Within the literature, the term cystic hygroma is used interchangeably with lymphangioma and lymphatic malformation.^[4] Most commonly these malformations occur in the head and neck, although they have been described in a variety of other anatomical locations such as axilla (20%), mediastinum (5%), groin, retroperitoneal space and groin.^[3] Hygromas

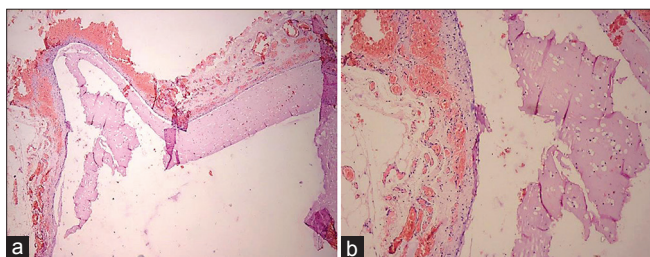


Figure 4: (a) Photomicrograph, showing cystic lumen formed by cystically dilated vascular lymphatic channels containing pinkish eosinophilic proteinaceous lymphatic fluid along with lymphoplasmacytic cell infiltration (H and E, $\times 10$). (b) Photomicrograph of cystic hygroma showing cystic lumen lined by single flattened endothelium layer (H and E, $\times 20$). The connective tissue wall is loosely fibroblastic with engorged capillaries, adipocytes and extravasated red blood cells

are probably the result of sequestration of lymphatic tissue that has retained its potential for growth in any of these areas.^[3] However, the etiology in the adult population is controversial. Some authors attribute adult lymphangioma to delayed proliferation of the congenital or acquired lymphoid rests following trauma or preceding respiratory infections.^[6] Cystic hygroma could be classified into septated (multiloculated) or nonseptated single cavity (nonloculated) types.^[7] There are three histological subtype of lymphangioma's. Capillary lymphangioma (composed of small lymphatics), cavernous lymphangioma (composed of larger lymphatics), cystic lymphangioma (cystic hygroma-composed of large macroscopic lymphatic spaces with collagen and smooth muscle). Cavernous lymphangioma is the most common subtype.^[8] Most lymphangiomas are asymptomatic. They have no gender predilection and present as a painless mass that progressively enlarges. Typically, the mass is soft, nontender and ill-defined. Symptoms may develop when the lymphatic malformation enlarges to where it compresses surrounding tissue. There may be obstructive symptoms such as dysphagia, dysphonia, and airway obstruction, however, this is rare in adults.^[9] Most cystic hygromas present in-utero or in infancy and, therefore, most of the literature on management considers pediatric cases.^[8] There is no consensus in the literature concerning the use of fine-needle aspiration cytology to diagnose these lesions. Differential diagnosis of the thymic cyst, pericardial cyst, bronchogenic cyst, cystic teratoma should be kept in mind.^[6] Complete excision of a cystic hygroma has been shown to have an 81% cure rate. When only part of a lymphatic malformation is excised, there is an 88% recurrence rate.^[10]

The present case was unusual, as a large cervical cystic hygroma presented *de novo* in an adult with no history of trauma or upper respiratory tract infection.^[7] Many treatment alternatives exist for lymphangiomas,



Figure 5: Postoperatively patient recovered well with no signs of neurological dysfunctions

including surgical excision, laser surgery, cryotherapy, electrocautery, steroid administration, sclerotherapy, embolization and radiation therapy, but surgical excision is the most preferred option.^[3] Total surgical excision was performed in this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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