REVIEW ARTICLE

Adult Onset Unilocular Cystic Hygroma in posterior triangle neck: An unusual patient with detailed Review of Literature

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ABSTRACT

Cystic hygroma (CH) also known as macrocystic lymphatic malformation is a well- recognized lesion in a pediatric patient, mostly under 2 years of age. This lesion is rarely seen in adults with only about 150 cases on record. Most of them are multilocular with infiltrative nature, unilocular CH being a rare entity. This lesion can occur in any part of body, having lymphatics but the most common site is neck region. In adults, CH is a diagnostic challenge because of its rarity and difficulty in differentiating it from other cystic lesions in the region. Multiple diagnostic modalities are taken into consideration before giving the final diagnosis. The gold standard is the histopathology. Surgical removal of lesion remains the preferred treatment for adult CH, but because of its multilocular, invasive nature, and proximity to the vital structures in the head and neck makes its surgery quite a challenging task. We present a case of adult-onset unilocular cervical CH with a detailed review of the literature highlighting different aspects of this disease process.
Keywords: Cervical Cystic hygroma, Head & Neck, Lymphatic malformation, lymphangioma

INTRODUCTION

CH also known as lymphangioma which is basically a congenital benign malformation of the lymphatic system when the lymphatic system fails to communicate with the venous system.The lesion was first reported by Reden Bacher in 1828 and the name “Cystic Hygroma “assigned by Werner in 1834[1,2]. It occurs in pediatric age group under 2 years[3]. Although CHis well known in pediatric practice it seldom occurs in adults. Adult cystic hygroma of the cervicofacial region recorded in English literature is less than 150 and optimum management of these lesions is still a matter of debate.[1]Theseswellings are in cervicofacial region but can present anywhere in the body where lymphatics are present[3]. CH may be multilocular or unilocular, these are locally aggressive benign lesions with a recurrence rate of 21%[4]. In the pediatric age group, the lesion is congenital, although the cause ofthis lesion in adults is not knownbut trauma and upper respiratory tract infections are considered as possible contributory factors for its onset[1]. The management of CH in adult age group is more challenging than in children, different diagnostic modalities are in practice but the final diagnosis is usually based on postoperative histopathology report[1]. Different treatment modalities are available, but the ideal treatment is still under debate, because of the rarity this disease process in adults and lack of experience internationally. Surgical excision is the preferred treatment and remains the treatment of choice in adult age group[5]. When surgical excision is not possible because of inaccessible site, proximity to vital structures, and recurrence, other treatment modalities like sclerotherapy by chemical drugs[6,7]is a better treatment option and withtimeis getting more popular. We present a case of unilocular cystic hygromain posterior triangle of neck in a 37 year healthy male along with a detailed review of theliterature.

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PATIENT PRESENTATION

A 37 years old man noticed a swelling in his left lower neck while taking bath about 04 months ago. It was a lemon size to start with, then gradually increased to the cricket ball size. Initially, he felt a slight discomfort in swelling which vanished in about a month. There were no complaints of difficulty in swallowing or respiration, but there was a history of repeated attacks of upper respiratory tract infections for the last 3 years. Clinical examination revealed that there was a smooth globular non- pulsatile swelling 6x7 cm in the left lower part of the posterior triangle of the neck just above the clavicle (fig 1-a & b). The overlying skin was normal with no scar mark, non-adherent tounderlying swelling. It was soft, non-tender, and fluctuant.

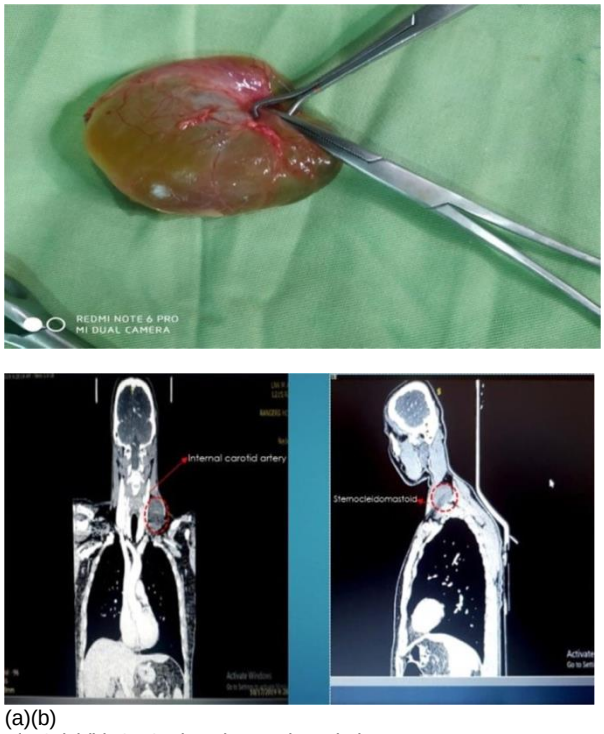
(a) (b)
Fig 1 (a)(b) Pre-operative pictures of patient revealing swelling in left posterior triangle of the neck

This swelling had ill-defined and indistinct margins. The fluid thrill was positive and the lesion was brilliantly trans illuminant. The swelling was dull to percuss with no bruit on auscultation. The loco regional lymph nodes were not palpable. Examination of chest, axilla, left upper limb, Pharynx, head, and neck were inconclusive. (Figure:1 a & b)

Based upon clinical presentation provisional diagnosis of adult onset cervical cystic hygroma was suspected with a differential diagnosis of lipoma, cold abscess, and branchial cyst. For confirmation or rejection of provisional diagnosis investigations were carried out, the results of which were confusing and notdefinitive.

Ultrasonography of the neck showed a subcutaneous cystic lesion measuring 42 x 47 x 29 mm with a volume of 29 ml, noted in the left lower posterior triangle of theneck. There were internal echoes suggestive of an abscess.

CECT scan of the chest and cervicofacial region was performed for clarity of diagnosis and to see the extent of the lesion (fig 2- a & b). It revealed a well- defined rounded fluid attenuation lesion measuring 5 x 5.4 x 5 cm in the posterolateral aspect of the left lower neck. Extending posterior to the sternocleidomastoid muscle and lateral to the left common carotid artery. There were no internal septations, solid components, or wall enhancement. The chest and axilla were clear. Findings suggested of 3rd branchial cleftcyst.

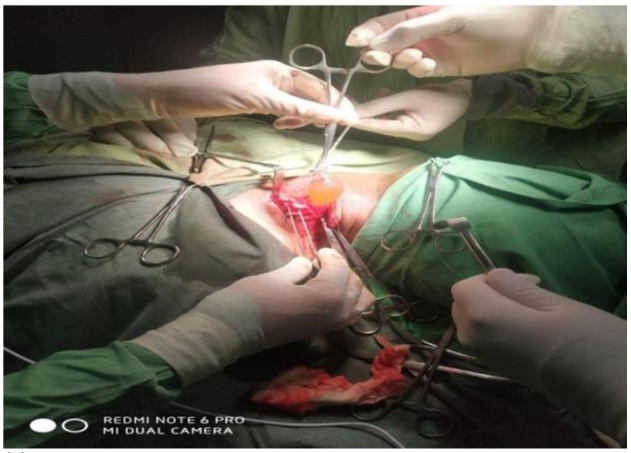


(a)(b)

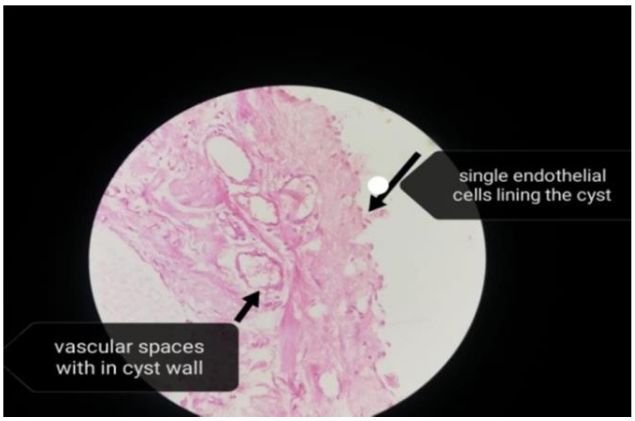
Fig 2 (a)(b) CECT head & neck and chest

Because of ambiguous clinical diagnosis, USG, and CECT diagnosis, FNAC was conducted to get a possible answer for diagnosis. 10 ml of yellowish fluid was aspirated which was subjected to cytology and chemical analysis. The report was again inconclusive. There were no WBCs or RBCs. PH 9, Specific Gravity 1.010, Glucose 1.9 mm0l/l, Proteins 35 gm./l, LDH 38U/L. Small lymphocytes were not seen in the aspirated fluid. These findings were not typical of a CH.

The excision of the lesion was planned for treatment and definitive diagnosis. Surgery was carried out under general anesthesia after informed consent. A transverse skin crease incision was made over the most prominent part of the underlying cyst, flaps raised; the cyst was inclose vicinity of left common carotid artery and left spinal accessory nerve. The lesion was dissected out and removed in toto preserving the nerve and vessel (fig 3- a & b). Skin and soft tissue closed in layers with a radivacsuction drain. Postoperatively there was uneventful recovery, drain was removed after 48 hours while the stitches were removed on the 7th post-operative day.



(a)



(b)

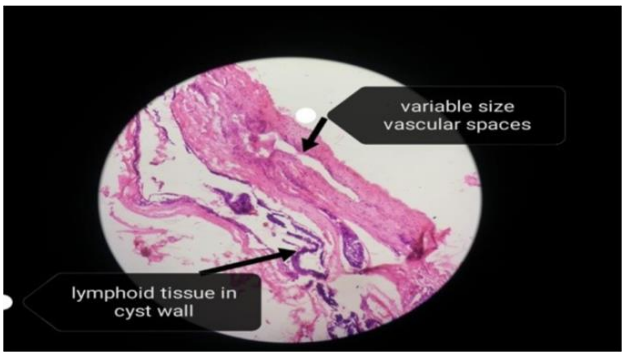
Fig 3 (a) Per operative findings with spinal accessary nerve visible. (b) CH specimen removedintact.

The cyst specimen was subjected to histopathology. The histopathology report revealed on gross appearance an intact cyst measuring 6 x 5 x 3 cm. Cut surface revealed unilocular cyst containing serous material with no solid areas and paper-thin wall.

Microscopic examination of the sections (fig 4-a & b) revealed cystic lesion comprised of large irregular vascular space lined by a single layer of cuboidal epithelium. The wall of the cyst had many variable-sized vessels and reactive lymphoid tissue consistent with cavernous

lymphangioma tissues that confirmed the diagnosis of unilocular cystichygroma.

Postoperatively patient had an uneventful recovery. There was no swelling or recurrence on follow up after 3, 6, and 9 months. Still the patient is followed up.



(a)(b)
Fig 4 (a)(b) Histopathology slides of the specimen

Table1: DE Serres Staging System of CH[16]

Stage	Location
I	Unilateral Infrahyoid
II	Unitateral Suprahyoid
III	Unilateral supra And infrahyoid
IV	Bilateral Infrahyoid
V	Bilateral supra and infrahyoid

DISCUSSION

CH is a benign hamartomatous lymphatic malformation typically presenting in infants and children of age 2 years[8]. The reported incidence in the pediatric age group is 2.8: 1000 to 1: 16000 live births[8,9]. Synonyms for CH are lymphangioma and macrocystic lymphatic malformations[10]. CH is a rare presentation in the adult age group posing diagnostic and treatment challenges. The exact etiology oftheselesionsisunknown.Inthepediatric age group, it is considered to be an embryonic malformation where the lymphatic channels fail to communicate with the venous system[1]. There is controversy regarding the etiology of CH in adults, most of the cases areconsidered idiopathic while some may be acquired secondary to trauma, infection, surgery, or repeated upper respiratory tract infections[12]. Our patient gave a history of repeated upper respiratory tract infections for the last 3 years which could have been the etiological cause in this case. Some authors attribute adult-onset CH to the delayed proliferation of sequestered lymphatic tissue due to some triggering etiological factor[12]. CH can present in any place but the most common site being the head and neck region (70 - 80 %) with a preference for a posterior triangle as was observed in our case, other anatomical locations are axilla (20%), mediastinum (5%), groin and retroperitoneal space[13].

CH is classified grossly into multiloculatedor unilocular types[14]. Wooleyet all classified into three subtypes of lymphangiomas on histological basis, 1.Capillary lymphangioma (composed of small lymphatics) 2. Cavernous lymphangioma the most common type (composed of larger lymphatics) 3. Cystic lymphangioma (CH – composed of large macroscopic lymphatic spaces with collagen and smooth muscles)[15]. De Serres suggested a staging system for CH depending upon the location and extent of the lesions {Tab-1}[16].

As per de Serres staging system, the lesion in our patient was placed in stage I (Table1):CH commonly presents as a slowly growing painless swelling in adults[11]. Occasionally it may enlarge in a short span of time as mentioned in literature[17]. Vital structures in neck like brachial plexus, larynx, trachea esophagus and great vessels may be compressed or incorporated within the lesion[17]. Other presentation may be pain, hoarseness, dysphagia, and breathlessness[12,14,18]. Translucency may not be elicited in cases of adult-onset CH[11]. The patient who presented in our department gave the history of a painless swelling noticed four months back which gradually increased to the present size of 6x7 cm, translucency test was positive and it had proximity to the common carotid artery and spinal accessorynerve.

Diagnosis of CH in adults is a challenging task because of a rarity in this age group and should always be kept in mind while dealing with swellings in the head and neck region. The differential diagnosis includes thyroglossal cysts, lipoma, dermoidcyst,laryngocele, hemangiomas, branchial cleft cyst, abscess, sarcomas, lymphomas, and ranulas[10]. Diagnosis of CH in adults is based on history, clinical examination, and a number of diagnostic modalities including FNAC, USG, CECT head neck and chest, and MRI. Despite having most of these investigations still there could be confusion in diagnosis of CH. The final diagnosis is normally confirmed on histopathology of the excised specimen. FNAC of CH typically reveals a clear or straw-colored fluid containing mature lymphocytes and histiocytes[11,14]. USG is the initial radiological investigation advised which may reveal septated or non-septated fluid filled cystic lesions. CECT head and neck, as well as chest, is important not only for diagnosis but also to see the extent of the lesion and its relationship with the vital structures ofthe head and neck region and hence plays an important role in planning its management. On CECT the lesion appears as non-enhancing homogenous cystic lesions[19]. On MRI T1-weighted image reveals lesionshaving low to intermediate intensity, and on T2-weighted image shows no enhancement with contrast[19,20]. In our patient FNAC revealed straw-colored fluid with no lymphocyte. There was confusion about the diagnosis on both USG and CECT scans. USG was suggestive of an abscess while the CECT indicated the 3rd branchial cleft cystinthe left posterior triangle extending behind the left sternocleidomastoid muscle with close proximity to the left common carotid artery. The final diagnosis in our case was confirmed on the histopathology report of the excised specimen.

There are different treatment modalities available, but because of the rarity of this lesion in adults, there is no consensus on a single treatment. The treatment alternatives available for adult CH, include surgical excision, laser therapy, cryotherapy, electro cautery, steroid injections, sclerotherapy, embolization, and radiation therapy, but still, the preferred treatment is the surgical excision[21]. One has to take into consideration many factors like the lesion is

complex or simple, its location, extent, proximity to vital structures in the region or a recurrent lesion while deciding treatment modality. Because of the lack of facilities, expertise, and complications as well as recurrence, cryotherapy, electro cautery, embolization, radiation, simple aspiration, and incision drainage are not being commonly practiced in adult CH. In our case, it was a simple unilocular lesion that was removed intact without any injury to the surrounding vital structures. The cure rate after complete excision of CH in adults is 81%. When only part of CH is excised, the recurrence rate is 88%[22]. The recurrence depends upon different factors like multilocularity, site, and relationship with the vital structures[11,15]. Our patient was reviewed at 3, 6, and 9 months postoperatively, he was asymptomatic without any recurrence.

The complication rate of surgery is 15–30%, the complications commonly experienced are hemorrhage, lymphoria, infection, vascular injury, neural injury and fistula formation[11,17].Sometime complete surgical excision is not possible, because of its large size and structural complexity , then other treatment modalities can be used in these cases. Simple aspiration is not recommended except for emergency relief of pressure because of recurrence, infection, and fistula formation. Sclerotherapy with OK-432 is an effective alternative mode of treatment in adults[8]. Sclerotherapy studies are mainly available in the pediatric age group. Sclerotherapy has a low success in adults thus limiting its use as primary treatment modality in this age group[8,15]. Other sclerosing agents used are steroids, alcohol, bleomycin sulfate, doxycycline, cyclophosphamide, interferon, and hypertonic saline[15]. Bleomycin sulfate and OK-432 are the mainstay agents used for sclerotherapy. Limitations in this treatment modality includes, requirement of multiple injection sessions, anaphylaxis like reactions have been reported, pain, fever, localized edema, infection, sepsis, myalgia, and sudden enlargement of the lesion are other possible complications[8,15,23]. Sclerotherapy is successful in simple CH which is 1-5cm in size and below mylohyoid[8]. Laser therapy can be used effectively for superficial microcytic lesions. Additional different options availableinclude ND: YAG laser,CO^2 laser, pulsed dyelaser, and diode laser[24]. Radiofrequency ablation was initially used for microcytic lesions but has been abandoned later on due to risk of malignant transformation[25,26]. Long termfollowup is required as recurrence has been reported even after six years of surgical excision[11,27].

CONCLUSION

The adult-onset cervical cystic hygroma is a rare presentation which not only causes diagnostic confusion but also treatment is technically challenging because of its complexity and close proximity to the vital structures in the cervico-facial region. The CECT scan of the head and neck region is important preoperative aid in the diagnosis and planning of treatment modality and especially surgery. The definitive diagnosis of cystic hygroma is confirmed on the histopathological report. Although different treatment modalities are available, but surgical excision remains the mainstay of treatment in adult-onset cervical cystic hygroma. We hope that our experience in dealing with this rare lesion will add up to a better understanding of thisdisease.

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Conflict of Interest: The authors have no conflict of interest regarding the publication of this paper.

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