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Various anatomic localizations of cystic hygroma: a retrospective analysis of 16 cases

Kistik higromanın farklı anatomik lokalizasyonları: 16 olgunun retrospektif analizi

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ABSTRACT

Background: This study aims to report the results of cystic hygroma operations performed in our clinic.

Methods: Medical records of 16 patients (7 males, 9 females; mean age 39.9±17.5 years; range 11 to 69 years) operated for cystic hygroma in our clinic between January 2011 and December 2015 were retrospectively analyzed. Data including age and gender of the patients, anatomic localization of the lesion, length of hospital stay, postoperative complications, and mortality and morbidity rates were recorded.

Results: The most common symptoms were soft tissue swelling, pain, and cough. The most common anatomical localization was cervical region (n=8, 50%), followed by axillary region (n=3, 18.8%), mediastinal region (n=3, 18.8%), and thoracic wall (n=2, 12.5%). Five patients experienced minor complications.

Conclusion: Although common in childhood, cystic hygroma may also affect adults and occur in various anatomic localizations. Cystic hygroma should be considered in case of soft fluctuating masses or asymptomatic cervicomediastinal lesions. Surgery is the primary management approach, and when a cyst is not totally excised, there is a high risk of recurrence.

Keywords: Adult; cystic hygroma; surgery.

ÖZ

Amaç: Bu çalışmada kliniğimizde yapılan kistik higroma ameliyatlarının sonuçları bildirildi.

Çalışma planı: Ocak 2011 - Aralık 2015 tarihleri arasında kliniğimizde kistik higroma nedeni ile ameliyat edilen 16 hastanın (7 erkek, 9 kadın; ort. yaş 39.9±17.5 yıl; dağılım 11-69 yıl) tıbbi kayıtları retrospektif olarak incelendi. Hastaların yaşı ve cinsiyeti, lezyonun anatomik lokalizasyonu, hastanede kalış süresi, ameliyat sonrası komplikasyonlar ve mortalite ve morbidite oranları dahil olmak üzere veriler kaydedildi.

Bulgular: En sık görülen semptomlar yumuşak doku şişliği, ağrı ve öksürük idi. En sık görülen anatomik bölge servikal bölgeyi (n=8, %50) takiben, aksiller bölge (n=3, %18.8), mediastinal bölge (n=3, %18.8) ve toraks duvarı (n=2, %12.5) idi. Beş hastada minör komplikasyonlar gelişti.

Sonuç: Genellikle çocukluk çağında görülmesine rağmen, kistik higroma erişkinleri de etkileyebilir ve çeşitli anatomik lokalizasyonlarda görülebilir. Yumuşak fluktuasyon veren kitlelerde veya asemptomatik servikomediastinal lezyonlarda akılda tutulmalıdır. Başlıca tedavi yaklaşımı cerrahi olup, kist total olarak çıkarılmaz ise, nüks ihtimali yüksektir.

Anahtar sözcükler: Erişkin; kistik higroma; cerrahi.

Cystic hygroma (CH) is a congenital malformation of the lymphatic system, which develops when a connection is formed between the lymphatic vessels and venous system for an unknown reason.[1,2] It is the second most common benign vascular tumor of childhood.[1] Its incidence is 1/6000 live births.[1] Studies on aborted fetuses have estimated an incidence of 1/750.[2] It accounts for 0.7 to 4.5% of all mediastinal tumors.[3] Although it typically involves the neck (75 to 90%), it may also involve the axillary region in

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20%, mediastinum in 5% of cases, and retroperitoneal region and, rarely, thoracic wall.[3]

Cystic hygroma grows slowly and may, even rarely, regress spontaneously. Hemorrhage or infection in a cyst may lead to a rapid increase in its diameter. Aspiration of the cyst content may provide a partial and temporary benefit; however, cyst infection or recurrence may ensue thereafter. Complete resection of a cyst is the most optimal treatment approach.[3]

In this article, we report the results of CH operations in our clinic and discuss the results in the light of the relevant literature.

PATIENTS AND METHODS

The medical records of 16 patients (7 males, 9 females; mean age 39.9±17.5 years; range 11 to 69 years) operated for CH in our clinic between January 2011 and December 2015 were retrospectively analyzed. Data including age and gender of the patients, anatomic localization of the lesion, length of hospital stay, postoperative complications, and mortality and morbidity rates were recorded. All patients underwent posteroanterior (PA) chest X-ray, thoracic computed tomography (CT), complete blood count, and routine biochemistry analysis. A neck CT was obtained for patients with a cervical lesion. Magnetic resonance imaging (MRI), which plays an important role for the radiological diagnosis of CH, was only performed in certain cases in whom a clear distinction between cystic structures and surrounding tissues was unable to be done, to prevent unnecessary costs.

Operative technique

All patients were operated on under general anesthesia at the operation theatre. A double-lumen endotracheal intubation was performed in three patients with amediastinal cyst localization, while a single-lumen intubation was performed in the remainder patients. The patients were positioned on the operating table depending on the lesion localization (supine, right or left lateral decubitus position). All lesions were totally excised including their walls. Following achieving hemostasis, the layers were closed anatomically. A hemovac drain (B-Vak 400 mL Wound Drainage System 14 CH, Bıçakcılar Tıbbi Cihazlar San. ve Tic. A.Ş., İstanbul, Turkey) was placed in cases without an entry into the thoracic cavity while a single 28-F chest tube was used for three patients operated through thoracotomy. All patients were extubated on the operating table and, then, transferred to the ward.

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 21.0 software (IBM Corp., Armonk, NY, USA). Quantitative variables were expressed in mean ± standard deviation (SD), while qualitative variables were presented in number and percentage. The chi-square test was used to compare the frequencies of the expected and observed qualitative variables.

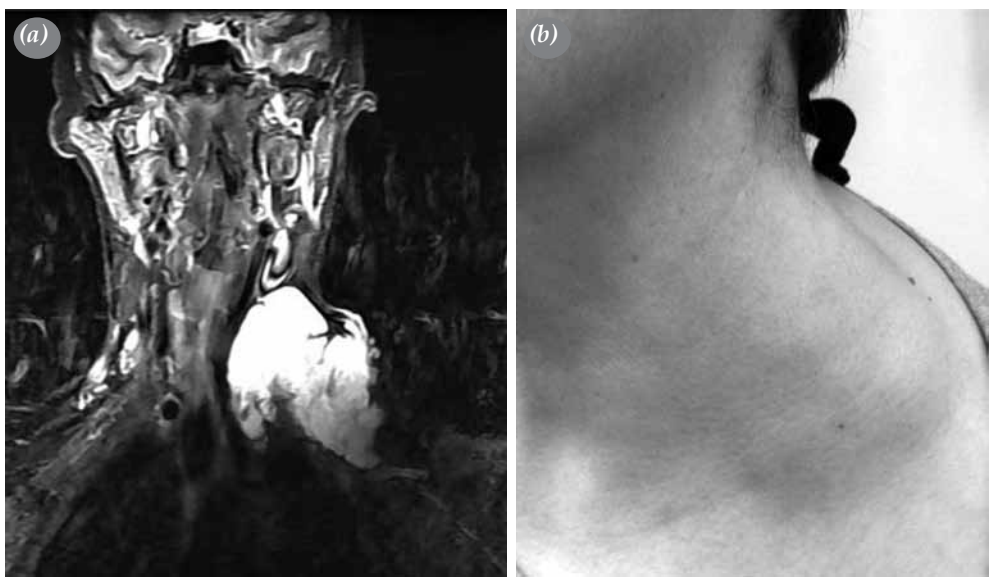

Figure 1. A 59-year-old woman. **(a)** a hyperintense lesion on T_2-weighted coronal section, **(b)** a mass lesion in the left cervicofacial region.

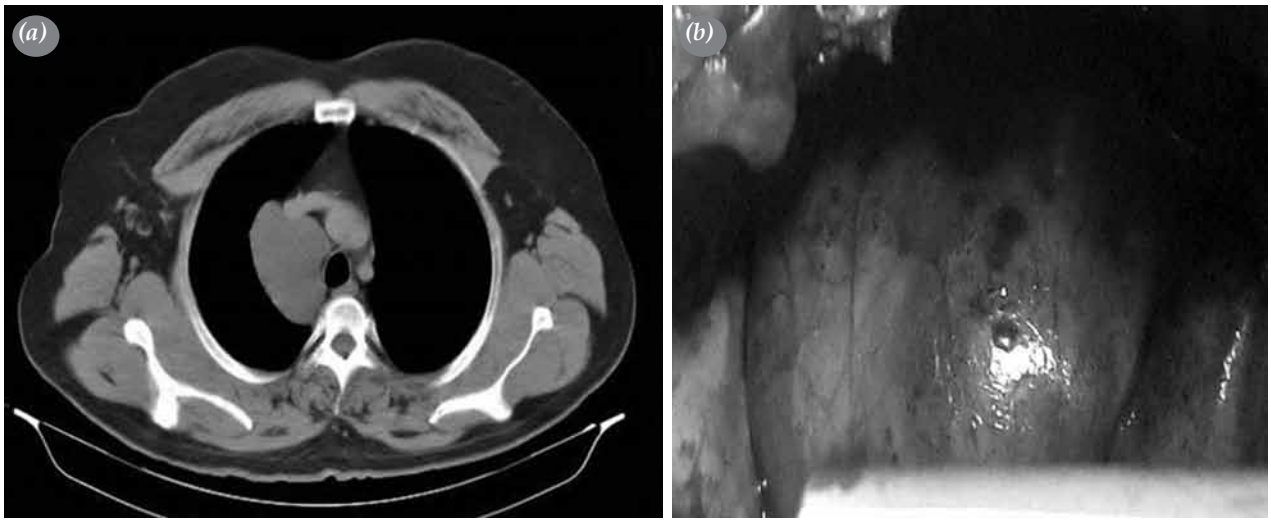

Figure 2. A 43-year-old man presenting with cough and mediastinal cystic lesion. **(a)** thoracic computed tomography showing the extent of the mass, **(b)** the intraoperative view of the mass lesion filled with fluid.

A *p* value of less than 0.05 was considered statistically significant.

RESULTS

Of all patients, two were of childhood age group (11 to 14 years), while 14 patients were adult cases (19 to 69 years). The most common symptoms were soft tissue swelling, pain, and cough. The lesions were most commonly localized to the cervical region (Figure 1) (n=8, 50%), followed by mediastinal region (n=3, 18.8%) (Figure 2), thoracic wall (n=2, 12.5%) (Figure 3), and axillary region (n=3, 18.8%) (Figure 4). Of the patients having CH in the cervical region, five (62.5%) were females (55.6% of female subjects) and three (37.5%) were males (42.9% of male subjects). Postoperative pain was rated using the visual analog scale (VAS). Three patients had a VAS score equal to or greater than 6 within the first 24 hours. A total of five patients (31.3%) developed minor complications, of which one was minor hemorrhage and one was wound infection. No major vessel injury or death occurred. The mean length of hospital stay was 3.2 (range 2 to 5) days

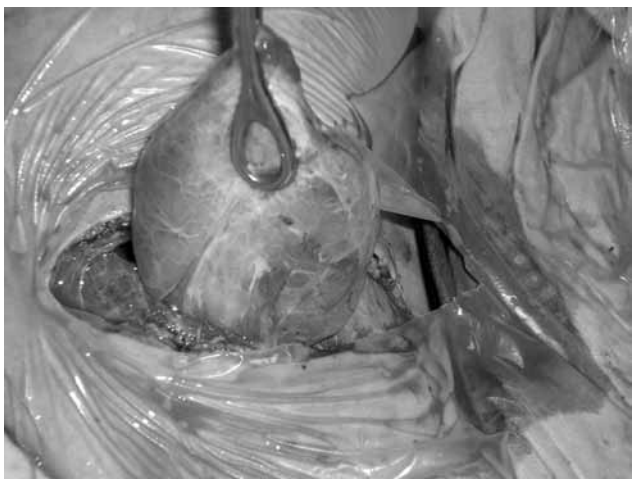

Figure 3. An 11-year-old male patient. The intraoperative view of the septated structure of the lesion.

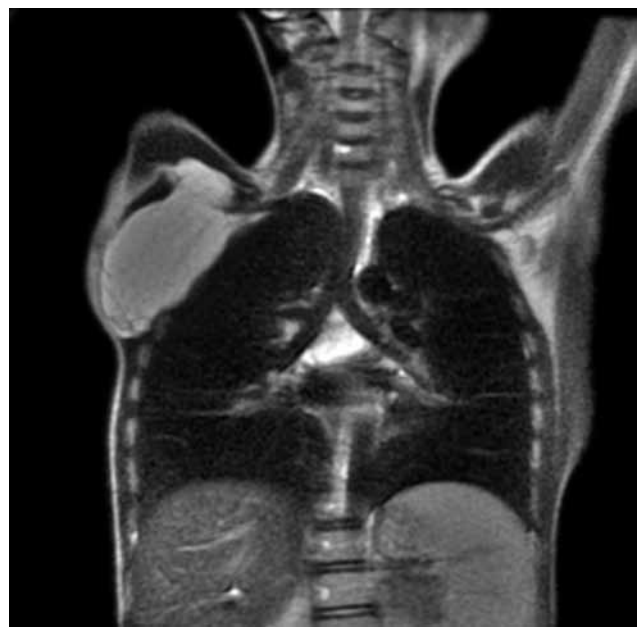

Figure 4. A 14-year-old male patient. Hyperintensity of the lesion with fluid content on T_1-weighted coronal section was due to its possible protein content.

Table 1. Demographic characteristics of patients

Variables	Patients (n=16)			
	n	%	Mean±SD	Min-Max
Age (year)			39.9±17.5	11-69
Duration of stay (day)			3.2±1.0	2-5
Gender				
Male	7	43.8		
Female	9	56.2		
Symptoms				
Swelling	13	81.3		
Pain	5	31.3		
Cough	3	18.8		
Complications				
Pain	3	18.7		
Minor hemorrhage	1	6.3		
Wound infection	1	6.3		
Anatomic localization				
Cervical	8	50.0		
Axillary	3	18.8		
Mediastinal	3	18.8		
Thoracic wall	2	12.5		

SD: Standard deviation; Min: Minimum; Max: Maximum.

(Table 1). There was no statistically significant correlation between the anatomical localizations of CH and age and gender (Table 2).

The histopathological examination of tissue samples macroscopically revealed a cross-section composed of multiple small and large cysts of a colorless, gelatinous fluid oozed. Microscopically, multiple lymphoid aggregates and lymphoid follicles were seen inside a fibroadipose cyst wall. Cross-sectional examination revealed that a single-layer of squamoid benign endothelial cells lined the inner surface of the large, dilated, irregular cystic structures (Figure 5). A small amount of an eosinophilic proteinaceous granular material and lymphocytes were present in a few cysts. In immunohistochemical examination, the endothelial cells lining the inner cyst surface were stained with D2-40 and CD 31 (Figures 6 and 7). Based on all histopathological and immunohistochemical findings, all specimens were reported as CH.

DISCUSSION

Lymphangiomas, benign congenital malformations of the lymphatic system, are most commonly located in the head and neck region; however, they may involve any body site where the lymphatic system originates.[4] Although these lesions may encircle and sometimes invade surrounding anatomic structures, they do not carry malignancy potential.[5] They are more prevalent in women, compared to men.[6]

The basic pathology of CH is the absence of connections supposed to form between the jugular vein

Table 2. Distribution of lesion localization according to gender

	Male (n=7)		Female (n=9)		
Localization	n	%	n	%	*p*
Cervical	3	42.9	5	55.6	0.818*
Axillary	1	14.3	2	22.2	0.981
Mediastinal	2	28.6	1	11.1	0.981
Thoracic wall	1	14.3	1	11.1	0.955

* Chi-square test.

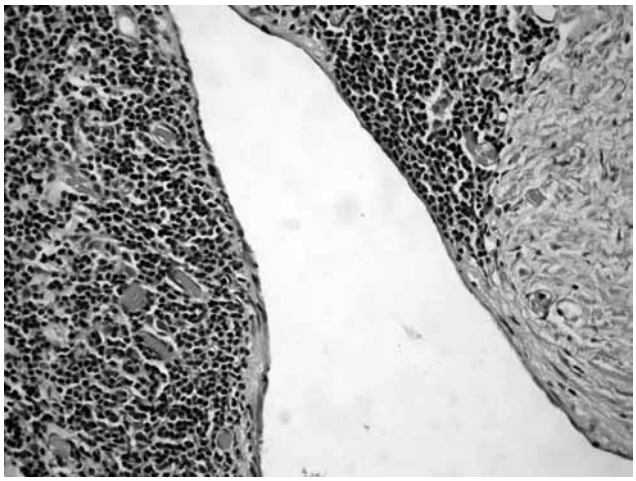

Figure 5. Sections showing lymphoid aggregates and a cyst lined by a single-layer of endothelial cells within a fibroadipose wall. (H-E x 40).

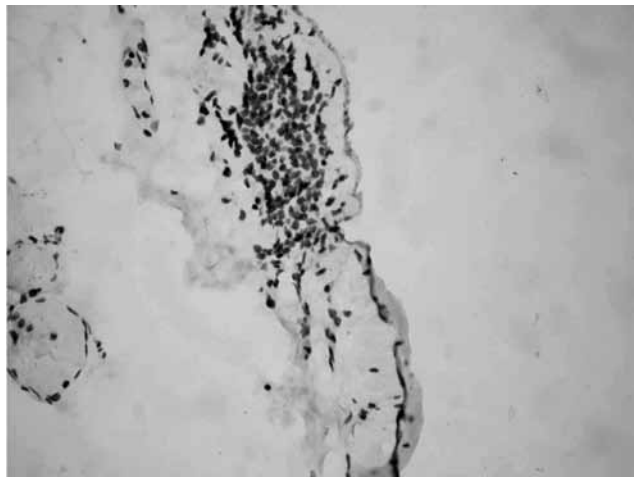

Figure 6. Immunohistochemical examination with D2-40 showing staining in endothelial cells. (D2-40, x40)

and lymphatics.[7] In fetus, the lymphatic system starts to develop at around fifth gestational week and expected to unite with the jugular vein in neck at seventh gestational week, and if the connection fails to develop by the 11th to 12th gestational week, CH process may begin.[8]

The etiology of this condition frequently involves an abnormal development of the lymphatic system.[9] A previous study demonstrated that lymphangiomas develop as congenital sequestrations of primitive lymphatic tissue with abnormal excretory channels.[10] In adults, in addition to congenital causes, other possible causes of CH include trauma, infection, previous surgery, and radiation.[11] However, no clear etiological factor was found in our cases.

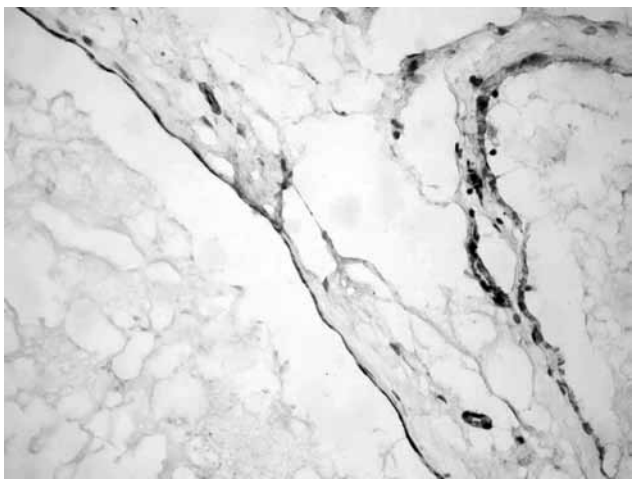

Figure 7. Immunohistochemical examination with CD31 showing staining of the cells lining the inner surface of the cyst. (CD31, x40).

Furthermore, there is no globally standardized pathological classification of lymphangiomas. In general, morphological and clinical classifications are made. Smith et al.[12] morphologically classified CH into macrocystic, microcystic, and mixed CH, whereas Mulliken et al.[13] proposed a more widely accepted histopathological classification including capillary, cavernous, and cystic lymphangiomas. However, the most practical classification is the one that is made according to lesion localization.[14] All histopathological types contain lymphatic channels lined by endothelial cells which are separated by a stroma composed of the connective tissue. Cystic hygroma consists of dilated lymphatic sacs, while cavernous and capillary lymphangiomas consist of smaller lymphatic channels.[5] Immunohistochemical studies in our study showed that endothelial cells lining the inner surface of a cyst were specifically stained with D2-40 and CD31.

The localization of CH is the cervicofacial region in 75%, axillary region in 20%, and mediastinal and retroperitoneal region in 5% of cases.[3,15] In our study, CH were localized in the cervical region in eight, in the axillary region in three, in the mediastinal region in three, and in the thoracic wall in two patients. In accordance with the literature, our patients were largely female and the CHs were mostly located in the cervical region. Another study reported that the lesions developed in less than six months prior to presentation and progressively grew in size in a couple of weeks.[16] The reason why CH reaches a greater size than other lymphangioma types is that their anatomic localization surrounded by loose

connective tissue allows lesion growth.[5] In our study, two patients had soft tissue masses which were progressively growing for the past six months; one of these lesions was located in the thoracic wall and the other in the axillary region.

In addition, a connection between CH and certain chromosomal anomalies has been reported.[17] It is most commonly (40 to 80%) associated with Turner syndrome. Other associated karyotype anomalies include Down syndrome, trisomy 21, Klinefelter syndrome, partial trisomies, partial monosomies, translocations, and mosaicisms.[18] However, a review of the literature did not provide any information about the prevalence of chromosomal anomalies in adult cases.[19] It was observed that two-thirds of mothers having children with CH had oligohydramnios during their pregnancies. In these cases, using transvaginal ultrasonography (USG), in addition to high-resolution USG, increases the rate of diagnosis, particularly within the first trimester.[18]

In addition to clinical findings, the diagnosis of lymphangioma frequently requires aspiration of the lesion content for cytological, histopathological, and radiological examinations. The commonly used radiological diagnostic methods include USG, thoracic CT, and MRI. On USG, these lesions appear as septae composed of multilocular masses, in which cystic appearance predominates.[19] In addition, MRI plays an important role for making the diagnosis of CH and assessing its extension to the adjacent anatomical structures. Also, CH is characterized by a high-signal intensity on T_2-weighted MRI images. On T_1-weighted images, on the other hand, it may have a low- or high-signal intensity, depending on the protein content of the lymphatic fluid.[5,9] In three of our patients diagnosed with mediastinal CH, MRI images were obtained to identify the relationship of the lesion with both surrounding tissues and great vessels. The MRI appearances were consistent with those reported in the literature, and no vascular invasion was detected.

Surgical excision is the recommended approach for lymphangioma treatment. It is technically important to remove the cyst wall totally. Complete resection provides cure in 81% of cases.[20] As important structures surround these lesions, however, excision is typically limited to 50 to 70% of the mass to avoid possible complications and anesthetic problems.[8,21] In addition, CH recurs at a rate of 88% after partial excision.[20] Riechelmann et al.[22] reported no recurrence after total excision, although 56% of the patients and 100% of the patients had recurrences after subtotal excision and after partial excision or aspiration, respectively. In our study, we performed total excision in all cases and found no recurrence during a six-month follow-up period.

Radiotherapy, embolization, cyclotherapy, electrocautery, cryotherapy, and laser therapy have been recently introduced as alternatives to surgical treatment for the management of CH.[15] In particular, successful results have been reported with the use of intra-lesional sclerosing agents.[23] The two most commonly used agents are bleomycin and OK-432; the former is commonly used and has a considerably favorable side effect profile.[24]

Although some surgeons recommend a conservative management for asymptomatic cases, spontaneous regression of the lesions has been also reported, albeit rare.[17] In these cases, it is believed that a connection between lymphatics and the jugular vein is formed later, leading to the regression of the cystic formation.[8]

In conclusion, although cystic hygroma is usually seen in childhood, it may also affect adults and can be located in different regions of the body. It should be considered in case of a soft fluctuating mass or an asymptomatic cervicomediastinal lesion. Surgery is the primary treatment approach, and the risk for recurrence increases, when a cyst is not totally excised.

Declaration of conflicting interests

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REFERENCES

1. Stringel G. Hemangiomas and lymphangiomas. In: Aschraft KWH, editor. Pediatric Surgery. Phidelphia: Elsevier; 1990:802-22.
2. Chen CP, Liu FF, Jan SW, Lee CC, Town DD, Lan CC. Cytogenetic evaluation of cystic hygroma associated with hydrops fetalis, oligohydramnios or intrauterine fetal death: the roles of amniocentesis, postmortem chorionic villus sampling and cystic hygroma paracentesis. Acta Obstet Gynecol Scand 1996;75:454-8.
3. Akar E. A rare location of cystic hygroma: a case report. Turk J Thorac Cardiovasc Surg 2013;21:1145-7.
4. Beech AN, Farrier JN. An Interesting Association of Cystic Hygroma of the Neck and Lymphangioma Causing a Paediatric Swollen Tongue. Case Rep Pediatr 2016;2016:7930945.
5. Onur MR, Özel K, Demir F, Özdemir H. Dev Kistik. Higroma: US ve MRG özellikleri F.Ü. Sağ Bil Derg 2007;21:141-4.
6. Yüksel M, Öztek İ, Çelik M, Halezeroğlu S, Sili M. Mediastinal kistik higromalar (iki olgu nedeni ile). Turk Patol Derg 1989;5:54-7.

7. Bahl S, Shah V, Anchlia S, Vyas S. Adult-onset cystic hygroma: A case report of rare entity. Indian J Dent 2016;7:51-4.
8. Ünüvar Ş, Tanrıverdi Hİ, Şenel U, İnce DA, Takçı Ş, Çakmak B. Intralesional Bleomycin Injection at Newborn with Huge Cystic Hygroma: Case Report. Causa Pedia 2014;3:777.
9. Togo S, Ouattara MA, Maiga IB, Lu Y, Jin D, Sangaré I, et al. A rare case of a giant cavernous lymphangioma of the chest wall in a child. Open Journal of Respiratory Diseases 2016;6:1-6.
10. Dowd CN. XI. Hygroma Cysticum Colli: Its Structure and Etiology. Ann Surg 1913;58:112-32.
11. Çipe G, Genç V, Karaca AS, Çakmak A, Baskan S. Üst Ekstremite yerleşimli kistik higroma: Olgu sunumu. Ankara Üniversitesi Tıp Fakültesi Mecmuası 2009;62:181-2.
12. Smith RC, Sherk HH, Kollmer C, Javitt MC. Cystic lymphangioma in the adult: an unusual axillary mass. Magn Reson Imaging 1989;7:561-3.
13. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg 1982;69:412-22.
14. de Serres LM, Sie KC, Richardson MA. Lymphatic malformations of the head and neck. A proposal for staging. Arch Otolaryngol Head Neck Surg 1995;121:577-82.
15. Fidan V, Sütbeyaz Y. Lingual cystic lymphangioma in an elderly patient. Kulak Burun Bogaz Ihtis Derg 2008;18:260-2.
16. Schefter RP, Olsen KD, Gaffey TA. Cervical lymphangioma in the adult. Otolaryngol Head Neck Surg 1985;93:65-9.
17. Gedikbasi A, Gul A, Sargin A, Ceylan Y. Cystic hygroma and lymphangioma: associated findings, perinatal outcome and prognostic factors in live-born infants. Arch Gynecol Obstet 2007;276:491-8.
18. Öztürk A, Sırmatel Ö, Gültekin E, Bitiren M. Dev kistik higroma: prenatal tanı ve bulgular. Diagn Interv Radio 2002;8:407-9.
19. McCaffrey F, Taddeo J. Surgical management of adult-onset cystic hygroma in the axilla. Int J Surg Case Rep 2015;7:29-31.
20. Rolekar NG, Shah DK. Recurrent cystic lymphangioma of the neck in an adult: A case report of rare entity. Int J Med Sci Public Health 2014;3:243-5.
21. Ameh EA, Nmadu PT. Cervical cystic hygroma: pre-, intra-, and post-operative morbidity and mortality in Zaria, Nigeria. Pediatr Surg Int 2001;17:342-3.
22. Riechelmann H, Muehlfay G, Keck T, Mattfeldt T, Rettinger G. Total, subtotal, and partial surgical removal of cervicofacial lymphangiomas. Arch Otolaryngol Head Neck Surg 1999;125:643-8.
23. Akyüz C, Ataş E, Varan A. Treatment of a tongue lymphangioma with sirolimus after failure of surgical resection and propranolol. Pediatr Blood Cancer 2014;61:931-2.
24. Sainsbury DC, Kessell G, Fall AJ, Hampton FJ, Guhan A, Muir T. Intralesional bleomycin injection treatment for vascular birthmarks: a 5-year experience at a single United Kingdom unit. Plast Reconstr Surg 2011;127:2031-44.