JoCASS
Çukurova Anestezi ve Cerrahi Bilimler Dergisi
2018 Adana

HEAD AND NECK LYMPHANGIOMAS IN ADULTS: A SINGLE CENTER'S EXPERIENCE

ERİŞKİNLERDE BAŞ VE BOYUN LENFANJİOMLARI: TEK MERKEZ DENEYİMİ

Meryem İlkay Eren Karanis

Konya City Hospital, Department of Pathology, Konya, Turkey

Sorumlu Yazar/Corresponding Author: Meryem İlkay Eren Karanis E-mail: dr-ilkay@hotmail.com

Geliş Tarihi/Received: 29.05.2021 Kabul Tarihi-Accepted: 18.07.2021 Available Online Date/Çevrimiçi Yayın Tarihi: 31.08.2021

Cite this article as: Eren Karanis Mİ. Head and Neck Lymphangiomas in Adults: A Single Center's Experience. J Cukurova Anesth Surg. 2021;4(2):122-30.

Doi: 10.36516/jocass.2021.80

Abstract

Introduction: To reveal the clinicopathologic features of adult head and neck lymphangiomas that underwent surgical resection in our hospital.

Materials and Methods: All surgical resection materials of patients aged older than 18 years from the head and neck region that were examined at our clinic between 2008 and 2020 were retrospectively reviewed. Cases diagnosed as lymphangioma were included in the study.

Results: Of the 34 lymphangiomas included in the study, 17 were from females and 17 from males. The ages of the patients ranged between 19 and 76 years. The median lymphangioma size was 0.8 cm. Lesions were located on the face in 15 (44.1%) patients, in the oral cavity in nine (26.5%) patients, on the neck in six (17.6%), on the orbital and periocular region in three (8.8%), and the ear in one (2.9%) patient. All lesions larger than 1.5 cm were located in the neck or oral cavity. All lesions in the oral cavity were diagnosed as cavernous lymphangioma. All four cystic lymphangiomas were identified in the neck. Lymphangioma recurred in two cases; no malignant transformation was detected.

Conclusion: Head and neck lymphangiomas are not uncommon in adults, they can be seen at any age, even in elderly patients. Cystic lymphangioma is often seen in the neck and cavernous lymphangioma in the oral cavity. Total surgical resection provides both treatment and definitive diagnosis. Recurrence can be seen, but malignant transformation is not expected.

Keywords: Lymphangioma; head and neck; cavernous lymphangioma; cystic lymphangioma; lymphangioma circumscriptum

Öz

Giriş: Hastanemizde cerrahi rezeksiyon uygulanan erişkin baş boyun lenfanjiomlarının klinikopatolojik özelliklerini ortaya koymayı amaçladık.

Gereç ve Yöntem: 2008-2020 yılları arasında laboratuvarımızda incelenen 18 yaş üstü hastaların baş boyun bölgesinden yapılan tüm cerrahi rezeksiyon materyalleri retrospektif olarak incelendi. Lenfanjiyom tanısı konulan olgular çalışmaya dahil edildi.

Bulgular: Çalışmaya dahil edilen 34 lenfanjiomdan 17'si kadın, 17'si erkekti. Hastaların yaşları 19 ile 76 arasında değişmekteydi. Medyan lenfanjiyom boyutu 0,8 cm idi. Lezyonlar 15 (%44,1) hastada yüzde, dokuz (% 26,5) hastada ağız boşluğunda, altı (% 17,6) hastada boyunda, üç hastada (% 8,8) orbital ve perioküler bölgede, bir hastada (%2,9) kulakta lokalize idi. Lenfanjiyomların 16'sı (%47,0) lenfanjiyoma sirkumskriptum, 14'ü (%41,2) kavernöz lenfanjiyom ve dördü (%11,8) kistik lenfanjiyom olarak tanımlandı. 1.5 cm'den büyük tüm lezyonlar boyun veya ağız boşluğunda yerleşmişti. Ağız boşluğundaki tüm lezyonlara kavernöz lenfanjiom tanısı konuldu. Dört kistik lenfanjiomun tamamı boyunda tespit edildi. Lenfanjiom iki olguda nüksetti; malign transformasyon saptanmadı.

Sonuç: Baş boyun lenfanjiyomları erişkinlerde nadir değildir, her yaşta görülebilirler. Kistik lenfanjiomlar sıklıkla boyunda ve kavernöz lenfanjiomlar ağız boğluğunda görülür. Total cerrahi rezeksiyon hem tedavi hem de kesin tanı sağlar. Nüks görülebilir ancak malign transformasyon beklenmez.

Anahtar Kelimeler: Lenfanjiyom; baş ve boyun; kavernöz lenfanjiyom; kistik lenfanjiyom; lenfanjiyoma sirkumskriptum; vasküler malformasyon

Introduction

Lymphangiomas are not considered as true neoplasms, they are thought to be malformations that occur as a result of a communication failure of lymphatic channels with the venous system. Congenital and early childhood lymphangiomas favor developmental malformations and genetic abnormalities also play a role[1]. Although the etiology is not well known in adults, it may occur secondary to infection, trauma and iatrogenic injuries[2]. Lymphangiomas are frequently observed in the pediatric age group and most frequently in the head and neck. Although most patients are aged younger than 2 years, lymphangiomas are also seen in adults[3]. It is slightly more common in males[1].

Lymphangiomas may be superficially or deeply located. Superficial lymphangiomas are known as lymphangioma circumscriptum, and deep lymphangiomas as cavernous or cystic[4]. Radiologic investigations are useful in diagnosis, but the definitive diagnosis is affirmed by histopathologic examination of surgical resection materials. Sclerotherapy or surgical excision can be used in treatment[5,6]. While evaluating the treatment options, it is taken into consideration whether the lesion is suitable for complete resection due to its location, size, and its relationship between vessels and vital organs. It is important not to have residual lesions during surgical resection because of the risk of recurrence of incomplete resections[7].

In this study, we aimed to reveal the clinicopathologic features of adult head and neck lymphangiomas that underwent surgical resection in our hospital.

Materials and Methods

Ethical approval was obtained for the study from the Ethics Committee of KTO Karatay University, Faculty of Medicine (2020/007). The study was conducted in accordance with the principles of the Helsinki Declaration.

All excisional biopsy materials of patients aged over 18 years from head and neck region that were examined at the Pathology

Clinic of Konya Education and Research Hospital between August 1st, 2008, and August 1st, 2020, were retrospectively reviewed. Cases diagnosed as lymphangioma were included in the study. Clinical and pathologic information such as the age and sex of the patients, location and size of the lesions, and follow-up information were obtained from patient files.

Statistical analyses were performed using the SPSS 22.0 for Windows software package (SPSS, Chicago, IL, USA). The Shapiro-Wilk test was used for examining continuous variables with normal and abnormal distribution, and one-way analysis of variance (ANOVA) was used for normally distributed continuous variables. The Kruskal-Wallis test was used for abnormally distributed continuous variables. When the Kruskal-Wallis test indicated statistically significant differences, the causes of the differences were determined by using a Bonferroni-adjusted Mann-Whitney U test. Continuous variables are presented as mean ± standard deviation (SD) or median (min-max), and categorical variables are presented as the number of cases and percentage.

Results

Samples from a total of 34 patients were included in the study; 17 were female and 17 were male. The median age of the patients was 52 (range, 19-76) years. The median lymphangioma size was 0.8 (range, 0.3-5) cm. Lesions were located on the face in 15 (44.1%) patients, in the oral cavity in nine (26.5%), on the neck in six (17.6%), the orbital and periocular region in three (8.8%), and the ear in one (2.9%) patient. The age, sex, and tumor size distributions of the patients according to lesion locations are given in Table 1. Of the lymphangiomas, 16 (47.0%) were defined as lymphangioma circumscriptum, 14 (41.2%) as cavernous lymphangioma, and four (11.8%) as cystic lymphangioma.

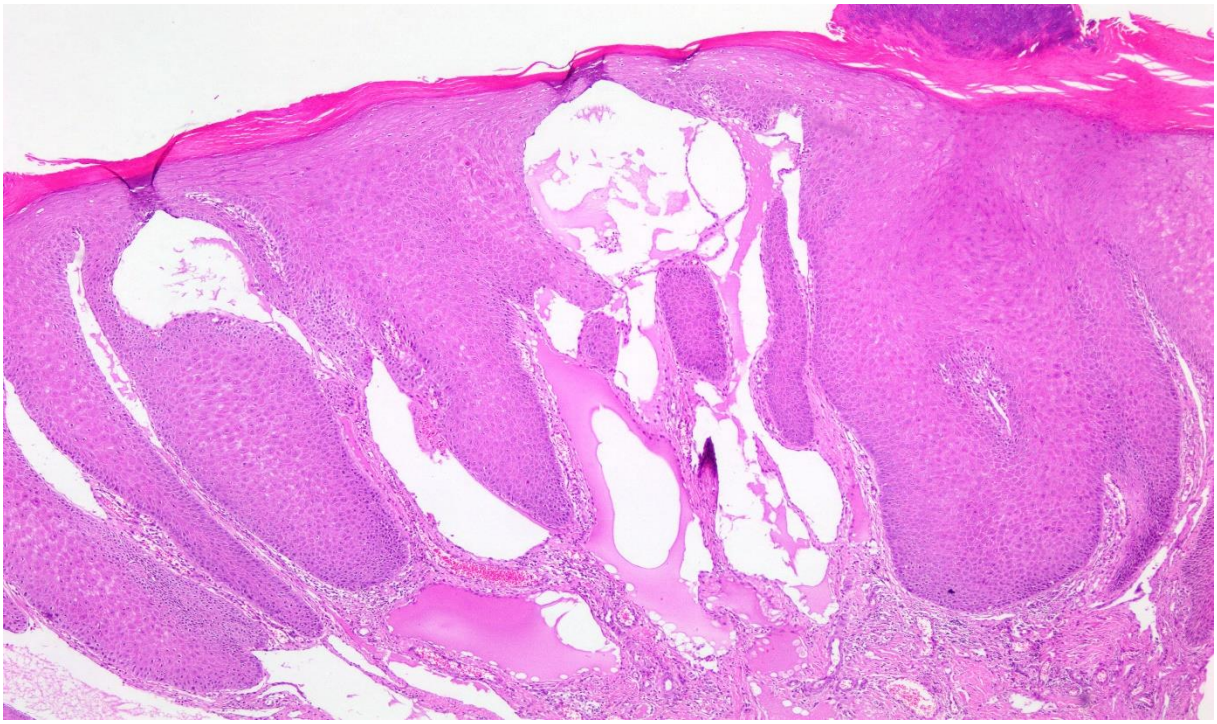

Figure 1. Lymphangioma circumscriptum: Hyperkeratosis, acanthosis and papillomatosis of the epidermis and markedly dilated lymphatic channels containing homogenous eosinophilic proteinaceous material in the papillary dermis. H&E x100

Table 1. The age, sex and tumor size distributions of the patients according to the lesion locations of head neck lymphangiomas.

Location (n)	Age (year) Median (min-max)	Sex Female/Male	Size(cm) Median (min-max)
Head			
• Face (n=15)	53 (19-68)	10/5	0,6 (0.3-1,5)
• Oral cavity (n=9)	43 (28-62)	5/4	1.7 (0.3-3)
• Orbital and periocular (n=3)	54 (48-62)	1/2	0,3 (0,3-0,6)
• Ear (n=1)	62	0/1	0.7
Neck (n=6)	35 (24-76)	1/5	3 (0,9-5)

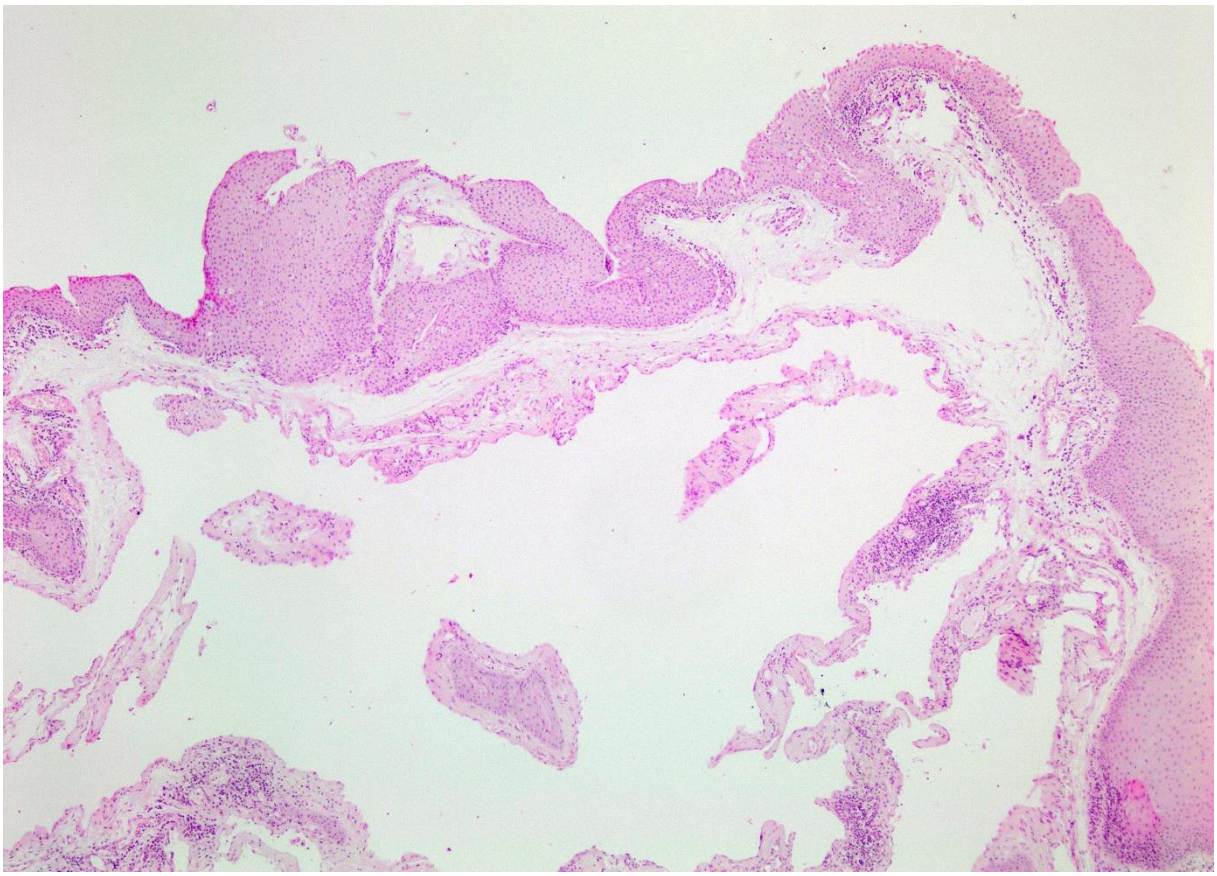

Figure 2. Cavernous lymphangioma in the conjunctiva: Lesion composed of dilated lymphatic vessels. H&E x40

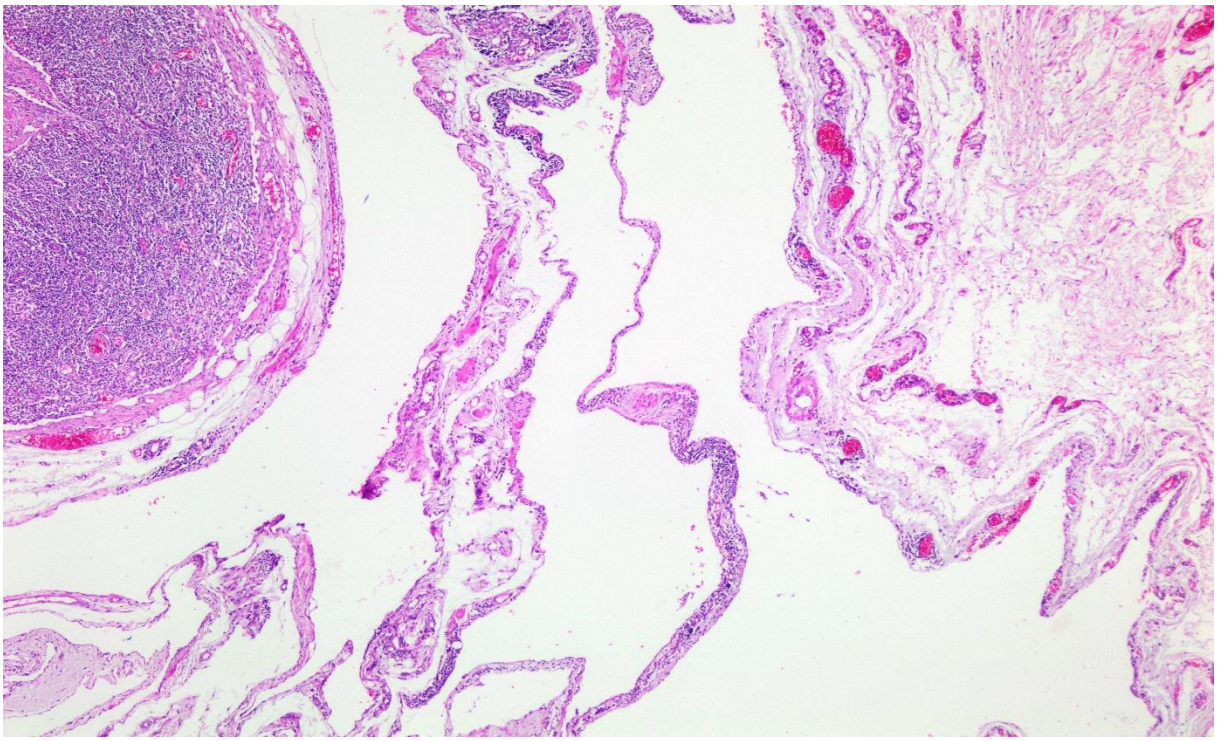

Figure 3. Cystic lymphangioma: Lesion composed of ectatic lymphatic vessels of different sizes lined with flattened endothelium. Lymphoid aggregates of varying densities are observed in septae. H&E x100.

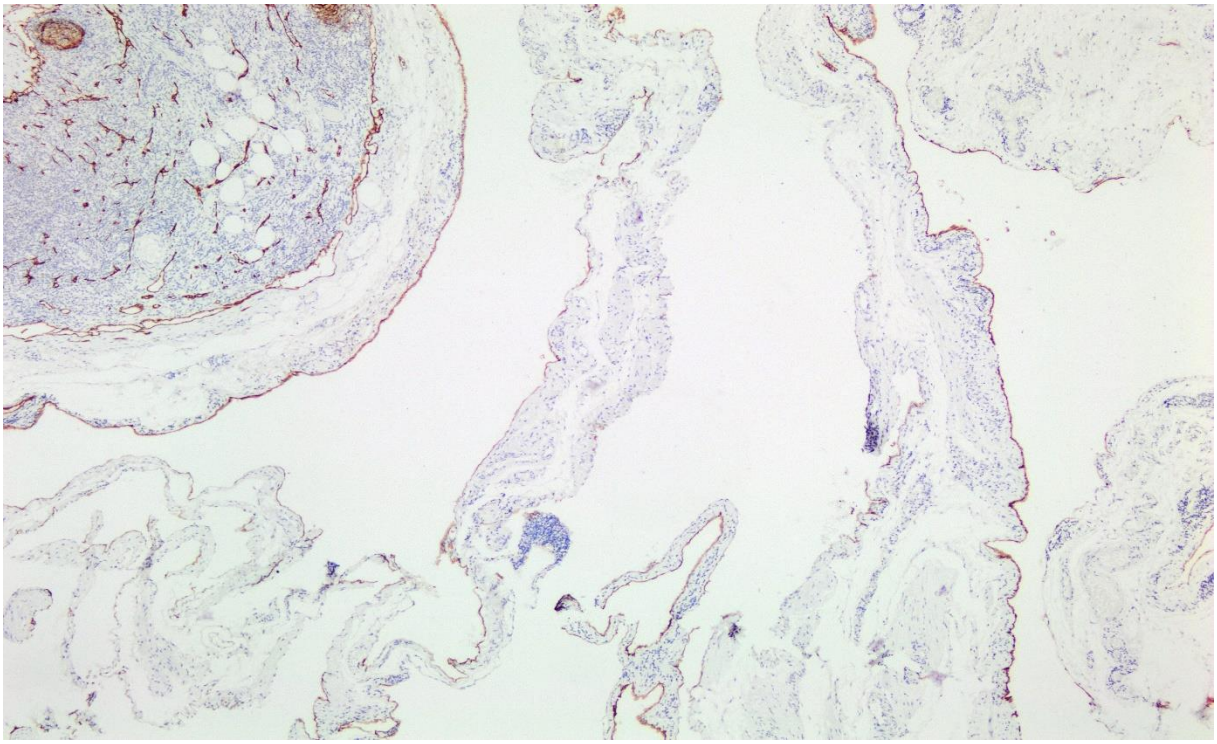

Figure 4. Cystic lymphangioma: D2-40 positivity in the endothelium lining the dilated vessels forming the lesion. D2-40 x100

Three of the lymphangiomas located on the face were defined as cavernous lymphangiomas and 12 as lymphangioma circumscriptum (Figure 1). All lesions in the oral cavity were diagnosed as cavernous lymphangioma. One of the oral lesions was located on the tongue. One of the orbital and periocular lesions was located on the conjunctiva, which was defined as cavernous lymphangioma (Figure 2). The other two lesions were located in the eyelids and were determined as lymphangioma circumscriptum, histopathologically. The lesion identified in the ear was lymphangioma circumscriptum. Four of the lesions located in the neck were cystic lymphangioma, one was cavernous lymphangioma, and the other was lymphangioma circumscriptum (Figure 3).

All lesions larger than 1.5 cm were located in the neck or oral cavity. All of the lymphangiomas in the oral cavity were cavernous. All of the cystic lymphangiomas were located in the neck. Lymphangiomas were multiple in two cases, one on the face and the other in the oral cavity. There were no patients with a family history of lymphatic malformation. Genetic examinations were not performed in any patients. Patients with superficial lymphangioma presented with pale or pink vesicular lesions on the skin. The other patients presented with a painless, growing mass. All patients had cosmetic symptoms but none had symptoms related to compression of surrounding tissues such as dysphagia, dyspnea, and voice disorder.

Immunohistochemical D2-40 and CD31 were positive in all patients (Figure 4). The follow-up period of the patients ranged from 5 months to 65 months; no malignant transformation was detected in any patients. Lymphangioma recurred in two patients. One case was 4.5 cm in size and a cervical cystic lymphangioma. The other patient with recurrence had an oral cavernous lymphangioma 3 cm in size. Sclerotherapy with bleomycin was administered to both patients.

Discussion

Lymphangiomas are benign malformations frequently seen in children. Sixty-five percent of lymphangiomas are congenital and 80% occur in the first two years of life[2]. Although lymphangiomas primarily occur in children and young adults, they can be encountered at any age and have been reported in adults[8]. The oldest patient in our series was a 76-year-old male.

Lymphangiomas can affect superficial or deep tissues and they are in generally grouped as superficial and deep subject to the depth and size of lymphatic channels. Superficial lymphangiomas are defined as lymphangioma circumscriptum, deep lymphangiomas include cavernous and cystic lymphangiomas. Lymphangioma circumscriptum is mostly seen on the skin of the extremities. They usually present as multiple clusters of pale, pink or red vesicles. Histopathologically, hyper-keratosis and acanthosis of the epidermis are observed and dilated lymphatic channels containing eosinophilic proteinaceous material in the papillary dermis is distinctive. The presence of lymphatic fluid in these dilated channels is critical in distinguishing lymphangioma circumscriptum from angiokeratoma, which has a similar appearance but contains blood in the vessel lumen[9]. In our series, lymphangioma circumscriptum was located on the face, periocular, and ear skin, and all patients presented with a pink or pale vesicle on the skin. The largest lymphangioma circumscriptum in our series was 1 cm in diameter and in one patient it was multiple.

Cavernous lymphangiomas are most commonly observed in the head and neck, followed by the gastrointestinal tract, mesentery, retroperitoneum, extremities, groin and axilla[10-15]. Cavernous lymphangiomas, which consist of enlarged lymphatic vessels, usually cause painless, growing swellings. All of the lesions we detected in the oral cavity were cavernous

lymphangiomas. We also described a cavernous hemangioma on the face and in one patient on the conjunctiva.

Cystic lymphangiomas are lesions seen in the neck, axilla, groin, and abdomen, presenting as large cystic masses, also called cystic hygromas[16-18]. Microscopy examinations of cystic lymphangiomas reveal that these lesions are composed of ectatic lymphatic vessels of different sizes, which are lined with flattened endothelium. In addition, lymphoid aggregates of varying densities are observed in septae. In some cases, lymphoid cells can even form lymphoid follicles[3]. Cystic lymphangiomas, like cavernous lymphangiomas, also present as a mass of painless, growing, swellings. Cervical cystic lymphangiomas can cause dyspnea, dysphagia or voice changes due to compression of the trachea, larynx or esophagus.

Although various treatment modalities are defined, surgical resection still seems to be the best treatment option for lymphangiomas. However, complete surgical resection of head neck lymphangiomas may be difficult or impossible. When cervical lymphangiomas are close to the upper airways and great vessels, and when they are too large, total surgical resection may not be performed or surgical complications may develop. Surgical resection may not be possible because deformity and loss of function may occur in lesions located in the retropharynx, tongue-base or oral-floor[19]. Especially in lymphangiomas for which complete surgical resection cannot be performed, the recurrence rate is around 39%. Recurrence may occur even after many years, but malignant transformation has not been reported[20]. In cases where complete resection is difficult or impossible due to the location of the lesion, other treatment methods such as sclerotherapy, radiotherapy, cryotherapy, electro-coagulation or laser applications can be performed. Bleomycin or doxycycline can be used as a sclerosing agent[5,6]. These treatment methods can be prefered either alone or together with surgery.

Lymphangiomas in adults can be clinically confused with dermoid cysts, branchial cleft cysts, lymph node neoplasms, and especially cystic metastatic lymph nodes[3]. In addition, teratoma, dermoid cyst, thyroglossal duct cyst, and neurofibroma should be kept in mind in the differential diagnosis of lymphangiomas[21]. Histopathologically, lymphangiomas are similar to hemangiomas and both consist of dilated vascular structures. However, it is typical to have homogeneous eosinophilic material in lymphangiomas, whereas there is blood in the dilated vessels in hemangiomas. Panendothelial immune-histochemical markers such as CD31 and CD34 are positive in both lymphangiomas and hemangiomas, and markers such as D2-40, vascular endothelial growth factor receptor 3 and podoplanin are specific for lymphatic vessels, which is useful in distinguishing lymphangiomas from hemangiomas[22]. D2-40 was used in all of our cases and an immunoreaction was detected in all of them in the endothelium lining the dilated vessels forming the lesion.

In conclusion, head and neck lymphangiomas are not uncommon in adults. They can be seen at any age, even in elderly patients. The lesions are generally asymptomatic and cause cosmetic problems. Cystic lymphangioma is often seen in the neck and cavernous lymphangioma in the oral cavity. Total surgical resection provides both treatment and definitive diagnosis. Recurrence can be seen, but malignant transformation is not expected.

Conflict of Interest

The authors declared they do not have anything else to disclose regarding conflict of interest with respect to this manuscript.

Funding

None

Ethical approval

Ethical approval was obtained for the study from the Ethics Committee of Karatay University, Faculty of Medicine (2020/007). All procedures performed in this study were in accordance with the ethical standards of the national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1- Thway K, Doyle LA. Lymphangioma and lymphangiomatosis. In: WHO Classification of Tumours Editorial Board. eds. Soft Tissue and Bone Tumours. 5th ed. Lyon: IARC Press, 2019:154-5.
2- Student PG, Rao CA. Lymphangioma of Neck in a 35-year Old Female: A Case Report of Rare Entity. Journal of Chalmeda Anand Rao Institute of Medical Sciences. Vol 2018;16(2): 207.
3- 3- Kaira V, Kaira P, Agarawal T. Cervical Cystic Lymphangiomas in Adults: A Case Series of a Rare Entity with Literature Review. Head and Neck Pathology. 2020;1-6. https://doi.org/10.1007/s12105-020-01227-y
4- Patel GA, Schwartz RA. Cutaneous lymphangioma circumscriptum: frog spawn on the skin. International journal of dermatology. 2009;48(12):1290-5. https://doi.org/10.1111/j.1365-4632.2009.04226.x
5- Jha SK, Jha S, Ranjan R, et al Management of Head And Neck Lymphangioma. Orissa Journal of Otolaryngolgy and HNS. 2019;13(1):39-43.
6- Porwal PK, Dubey KP, Morey A, et al. Bleomycin sclerotherapy in lymphangiomas of head and neck: prospective study of 8 cases. Indian Journal of Otolaryngology and Head & Neck Surgery. 2018;70(1):145-8. https://doi.org/10.1007/s12070-017-1243-x
7- Valletti PA, Brucoli M, Boffano P, et al. A single-center experience in the management of head and neck lymphangiomas. Oral and Maxillofacial Surgery. 2020;24(1):109-115. https://doi.org/10.1007/s10006-020-00832-z
8- Lerat J, Mounayer C, Scomparin A, et al. Head and neck lymphatic malformation and treatment: clinical study of 23 cases. European annals of otorhinolaryngology, head and neck diseases. 2016;133(6):393-6. https://doi.org/10.1016/j.anorl.2016.07.004
9- Fatima S, Uddin N, Idrees R, et al. Lymphangioma circumscriptum: clinico-pathological spectrum of 29 cases. Journal of the College of Physicians and Surgeons Pakistan. 2015;25(9):658.
10- Eren S, Cebi AT, Isler SC, et al. Cavernous lymphangioma of the tongue in an adult: a case report. Journal of Istanbul University Faculty of Dentistry. 2017:51(2):49. https://doi.org/10.17096/jiufd.64259
11- Lee DH, Yoon TM, Lee JK, et al. Cavernous Lymphangioma in the Maxillary Sinus. Journal of Craniofacial Surgery. 2019;30(8):2520-21. https://doi.org/10.1097/SCS.0000000000006003
12- Tan B, Zhang SY, Wang YN, et al. Jejunal cavernous lymphangioma manifested as gastrointestinal bleeding with hypogammaglobulinemia in adult: A case report and literature review. World Journal of Clinical Cases. 2020;8(1):140. https://doi.org/10.12998/wjcc.v8.i1.140
13- Wang X, Meng S, Duan K, et al. Treatment of Retroperitoneal Cavernous Lymphangioma: A Case Report. Chinese Medical Sciences Journal. 2020;35(3):283-5. https://doi.org/10.24920/003760
14- Leong JF, Levin KB, Rajkumar V. Cavernous Lymphangioma of the Digits: A Rare Cause of Macrodactyly. Medicine & Health. 2019;14(2). https://doi.org/10.17576/MH.2019.1402.24
15- Loetscher KCQ, Jandali AR, Garzoli E. Axillary cavernous lymphangioma in pregnancy and puerperium. Gynecologic andobstetric investigation. 2005;60(2):108-11. https://doi.org/10.1159/000085584
16- Cadena-Piñeros E, Rojas Gutiérrez A. Cervical lymphangioma in adults: case report and current treatment. Case reports. 2018;4(1):61-8. https://doi.org/10.15446/cr.v4n1.67067
17- Azim MT, Hussain SM, Mughal MA. Primary Supraclavicular Cystic Lymphangioma in an Adult: A Rare Presentation. Journal of the College of Physicians and Surgeons--Pakistan: JCPSP. 2019;29(6):S11-S12.
18- Aprea G, Guida F, Canfora A. Mesenteric cystic lymphangioma in adult: a case series and review of the literature. In BMC Surgery. 2013;13(S1):A4 https://doi.org/10.1186/1471-2482-13-S1-A4
19- Lerat J, Mounayer C, Scomparin A. Head and neck lymphatic malformation and treatment: clinical study of 23 cases. European annals of

otorhinolaryngology, head and neck diseases. 2016;133(6):393-6. https://doi.org/10.1016/j.anorl.2016.07.004
20- Kotsis T, Exarchos G, Metaxa L. Recurrent Neck Lymphangioma in a Young Adult: Twenty-Three Years After Successful Treatment. Vascular and endovascular surgery. 2019;53(2):170-6. https://doi.org/10.1177/1538574418814057
21- Usha V, Sivasankari T, Jeelani S. Lymphangioma of the tongue-a case report and review of literature. Journal of clinical and diagnostic research: JCDR. 2014;8(9):ZD12. https://dx.doi.org/10.7860%2FJCDR%2F2014%2F9890.4792
22- Kolay SK, Parwani R, Wanjari S. Oral lymphangiomas–clinical and histopathological relations: An immunohistochemically analyzed case series of varied clinical presentations. Journal of oral and maxillofacial pathology: JOMFP. 2018;22(Suppl 1):S108. https://dx.doi.org/10.4103%2Fjomfp.JOMFP_157_17