Kulak Burun Bogaz Ihtis Derg 2008;18(5):326-329

CASE REPORT

Cystic hygroma colli in adults: a report of two cases, one with atypical location

Yetişkinlerde kistik higroma kolli: biri atipik yerleşimli olan iki olgu

Fikret KASAPOĞLU, M.D., Nadir YILDIRIM, M.D.[1]

We presented two adult cases of cystic hygroma colli (CHC), one with atypical location. The first patient was a 23-year-old male with CHC located in the posterior cervical triangle, its common location in the neck. The latter was a 22-year-old male whose lesion occupied the submandibular region and extended to the skull base. The diagnosis was made by computed tomography, magnetic resonance imaging, and fine-needle aspiration biopsy, and confirmed by postoperative histopathologic examination. The lesions were surgically removed in both patients, and no recurrence was encountered during postoperative 12 and 14 months, respectively.

Key Words: Adult; head and neck neoplasms; lymphangioma, cystic/surgery; skull base neoplasms.

İki yetişkin hastada, birinde atipik yerleşimli olan kistik higroma kolli (KHK) sunuldu. Yirmi üç yaşında bir erkek olan ilk olguda lezyon, boyundaki tipik yerleşim yeri olan posterior servikal üçgende idi. Diğer olgu 23 yaşında bir erkekti ve KHK submandibüler alanda yerleşimliydi ve oldukça ender görülen atipik kafa tabanı uzanımı gösteriyordu. Tanı aşamasında bilgisayarlı tomografi, manyetik rezonans görüntüleme ve ince iğne aspirasyon biyopsisinden yararlanıldı. Kesin tanı ameliyat sonrası histopatolojik inceleme ile kondu. Olguların tedavisinde cerrahi eksizyon uygulandı. İki olgunun sırasıyla 12 ve 14 aylık takibi sırasında nüks bulgusuna rastlanmadı.

Anahtar Sözcükler: Erişkin; baş-boyun neoplazileri; lenfanjiyom, kistik/cerrahi; kafa tabanı neoplazileri.

Cystic hygroma or cystic lymphangioma is a congenital malformation of the lymphatic system that manifests itself as a soft, benign, and painless mass. The lesion occurring as a neck mass is called as cystic hygroma colli (CHC). It is widely accepted that they arise from the remnants of embryonic lymphatic tissue which retains the potential for proliferation. They grow in the fashion of sprouting and are capable of transgressing anatomical boundaries.[1,2] They can occur almost at any anatomical site. However, 75-80% of CHCs are located in the head and neck region.[3-5] In the neck, they are typically located within the posterior cervical triangle.[1,2,6-9]. The majority of cases (80-90%) are diagnosed under the age of two. In adults, it usually presents as a rapidly growing mass. Infections, tumors, and trauma are thought to trigger its growth and, during the periods of expansion, it may cause dyspnea, dysphagia, and symptoms of toxemia.[2,10-12] We presented two young adults with CHC.

◆ *Department of Otolaryngology, Medicine Faculty of Uludağ University (Uludağ Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Hastalıkları Anabilim Dalı), Bursa; [1]Department of Otolaryngology, Medicine Faculty of Yüzüncü Yıl University ([1]Yüzüncü Yıl Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Hastalıkları Anabilim Dalı), Van; all in Turkey.*

◆ *Received - October 11, 2006 (Dergiye geliş tarihi - 11 Ekim 2006). Request for revision - May 1, 2007 (Düzeltme isteği - 1 Mayıs 2007). Accepted for publication - June 6, 2007 (Yayın için kabul tarihi - 6 Haziran 2007).*

◆ *Correspondence (İletişim adresi): Dr. Fikret Kasapoğlu. Uludağ Üniversitesi Tıp Fakültesi, Kulak Burun Boğaz Hastalıkları Anabilim Dalı, 16069 Görükle, Bursa, Turkey. Tel: +90 224 - 442 83 03 Fax (Faks): +90 224 - 442 80 91 e-mail (e-posta): fkasapoglu@uludag.edu.tr*

CASE REPORT

Case 1– A 23-year-old male was referred to our clinic with a painless, slowly-growing swelling of three-year history in the left lateral side of the neck. The mass was palpated beneath the sternocleidomastoid muscle (SCM), extending to the supraclavicular region. It was soft, fluctuating, nontender and nondemarcated. Contrast-enhanced cervical computed tomography (CT) revealed a finely septated, hypodense cystic lesion, 5x4x2.5 cm in size, adjacent to the SCM medially and the great vessels posteriorly (Fig. 1). At surgery, the cystic mass was found lying over the deep cervical fascia, encapsulated by a fine membrane, and adjacent to the great vessels. Total excision of the mass was performed.

Case 2– A 22-year-old male was admitted to our department with a mass in the right submandibular region. He first noticed the lump two years before. It had undergone a slow but steady growth until the past two months, after which its growth became rapid. On examination, a soft, fluctuating, and painless mass was felt over the submandibular salivary gland. Histopathology of fine-needle aspiration biopsy (FNAB) material was consistent with CHC. Computed tomography of the head and neck showed a septated, hypodense, and lobulated cystic mass, 5x3.5x1 cm in size, surrounded by the submandibular gland medially and SCM posteriorly. T_2-weighted magnetic resonance imaging (MRI) of the same region revealed the extension of the lesion towards skull base (Fig. 2a, b). Excision of the mass lesion was carried. The mass was removed from the submandibular space together with the left submandibular salivary gland to which it was attached. Its

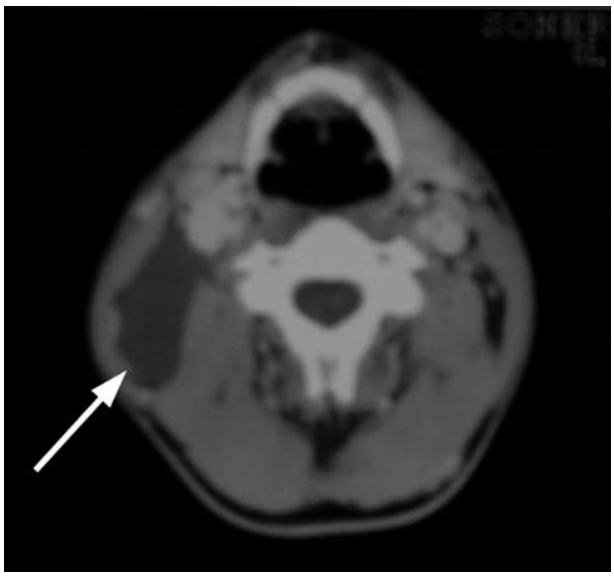

Fig. 1. *Computed tomography image of cystic hygroma located in the posterior cervical triangle (Case 1).*

superior extension up to the level of the jugular foramen was confirmed. With dissection over the capsula of the lesion, it was followed through the jugular foramen and excised totally. It had a soft, grayish, lobular, invasive, and noncapsulated structure.

Histopathologic diagnosis of surgical specimens in both patients was cystic hygroma (Fig. 3a, b). There was no evidence for recurrence in both patients during 12 and 14 months of follow-up, respectively.

DISCUSSION

Cystic hygroma, or cystic lymphangioma, is described as an abnormal proliferation of the lymphatic tissue that is congenital and benign in nature.

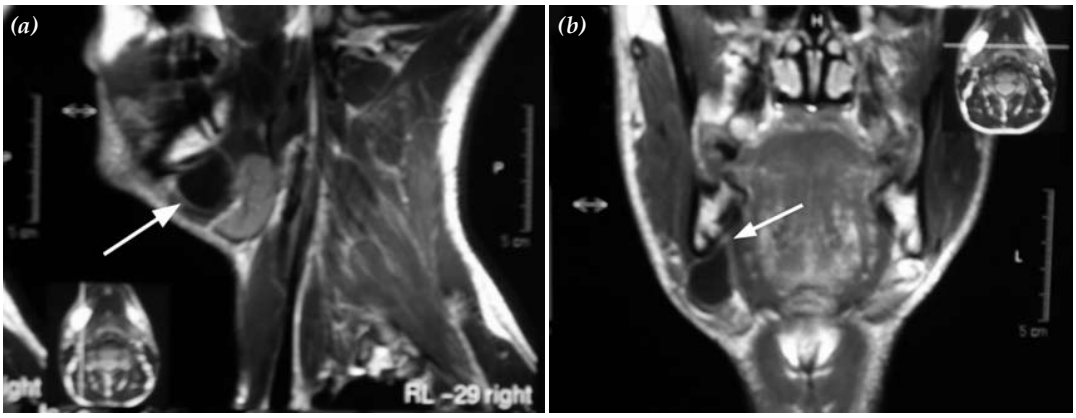

Fig. 2. ***(a)*** *Sagittal T_2-weighted magnetic resonance scan illustrates displacement of the submandibular gland by the cystic mass.* ***(b)*** *Extension of the mass through the level of the foramen jugulare on the coronal section (Case 2).*

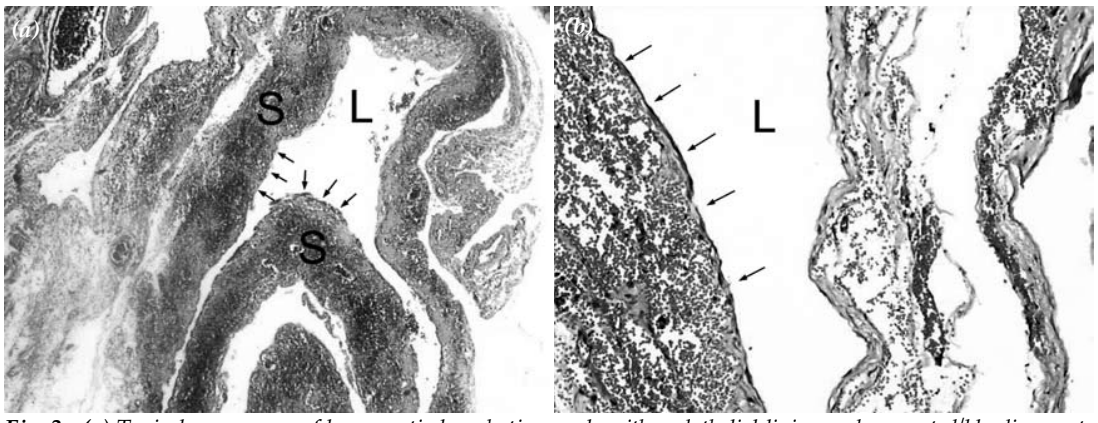

Fig. 3. ***(a)*** *Typical appearance of large cystic lymphatic vessels with endothelial lining and congested/bleeding septa separating each of these vessels (Case 2) (H-E x 25).* ***(b)*** *No blood elements are seen within the lumen and cavities that are lined by fine endothelium (Case 2) (H-E x 100). S: Septa; L: Lumen; Arrows: Endothelial lining.*

Many authors classify lymphangiomas into three categories: lymphangioma simplex, made up by thin-walled capillary lymphatic vessels, cavernous lymphangiomas by dilated lymphatic spaces, and cystic hygroma by cysts of various sizes.[1,2]

It can occur at any age, but 30-40% of them are detected in the newborn period, and in 80-90% of the patients within the first two years of life. Rarely, it is seen in adults in their 4th to 6th decades and its occurrence in younger adults, as in our patients, is even less common.[1,2,5,13] Schefter et al.[14] reported the largest series of adult CHC, with 32 patients. So far, nearly 100 adult cases have been reported.[3,5-9,11-15] Apart from our cases, the only series of CHC with skull base extension was reported by Tibesar et al.[16] with three cases.

In the differential diagnosis of CHC, branchial cysts, plunging ranula, thyroglossal cysts, lymphoma, lipoma, metastatic diseases, and hematoma should be considered.[1,6,10] The location of the masses and their consistency helped us in the diagnosis of our patients. In hygromas of atypical locations, FNAB is also a very useful tool for the diagnosis and differential diagnosis. Usually, a yellow liquid with mature lymphocytes and histiocytes are seen in the cystic lesion.[2,5,7,8,14] Contrast-enhanced CT and MRI are valuable in the differential diagnosis, and are also helpful in planning surgery by displaying the relation of the mass with neighboring structures. In this respect, T_2-weighted MRI is superior to CT especially for visualizing skull base extension, as it is not hindered by bony artifacts.[2-4,11]

Nonsurgical management of CHC has been advocated by some authors in order to avoid tedious and risky task of surgical resection.[5,17-23] Kennedy et al.[24] recommended awaiting spontaneous resolution in order to avoid potential hazards of surgery. Radiotherapy (RT) which was used in the past has now been almost abandoned due to the risk of malignant transformation of the lesion.[25] Sclerosing agents and RT are not suggested in adult CHC.[14] Sclerotherapy with bleomycin and OK-432 have been used in the treatment of CHC with some success, especially in children.[5,17-21] Sclerosing agents such as boiling water, sodium morrhuate, alcohol, and 50% dextrose are also used, but the disadvantages of these agents are their unpredictable results and extensive sclerosis, which may make future surgery extremely difficult when it is required.[2,7,8,25] Especially in children, nonsurgical treatment options are suggested for lesions located over the parotid region in order to avoid the complications of surgery.[18] Therefore, surgical excision still remains as the treatment of choice. In adult patients with more circumscribed hygromas, like the ones we presented, complete or near-complete resection can be achieved, considerably reducing the risk for recurrence.

In conclusion, CHC is a very rare entity in adults and must be considered in the differential diagnosis of neck masses. In diagnosis and treatment planning of CHC, FNAB, MRI and CT are useful tools. Total excision is the first choice of treatment.

REFERENCES

1. Baer S, Davis J. Cystic hygroma presenting in adulthood. J Laryngol Otol 1989;103:976-7.
2. Kennedy TL. Cystic hygroma-lymphangioma: a rare and still unclear entity. Laryngoscope 1989;99(10 Pt 2 Suppl 49):1-10.

3. Godin DA, Guarisco JL. Cystic hygromas of the head and neck. J La State Med Soc 1997;149:224-8.
4. Mahboubi S, Potsic WP. Computed tomography of cervical cystic hygroma in the neck. Int J Pediatr Otorhinolaryngol 1989;18:47-51.
5. Sichel JY, Udassin R, Gozal D, Koplewitz BZ, Dano I, Eliashar R. OK-432 therapy for cervical lymphangioma. Laryngoscope 2004;114:1805-9.
6. Guarisco JL. Congenital head and neck masses in infants and children. Part II. Ear Nose Throat J 1991;70: 75-82.
7. Çelenk F, Ceylan A, Köybaşıoğlu A, Gönül İI. Erişkin hastada servikal kistik lenfanjiom: olgu sunumu. KBB-Forum 2006;5:62-5.
8. Güner A, Aydın A, Çelik F. Cystic hygromas in adults: reports of two cases. Bakırköy Tıp Dergisi 2006;2:101-3.
9. Gidvani VK, Bhowmick SK. Midline posterior cervical cystic hygroma. South Med J 1999;92:340-3.
10. Cohen SR, Thompson JW. Lymphangiomas of the larynx in infants and children. A survey of pediatric lymphangioma. Ann Otol Rhinol Laryngol Suppl 1986; 127:1-20.
11. Gleason TJ, Yuh WT, Tali ET, Harris KG, Mueller DP. Traumatic cervical cystic lymphangioma in an adult. Ann Otol Rhinol Laryngol 1993;102:564-6.
12. Antoniades K, Kiziridou A, Psimopoulou M. Traumatic cervical cystic hygroma. Int J Oral Maxillofac Surg 2000;29:47-8.
13. Ricciardelli EJ, Richardson MA. Cervicofacial cystic hygroma. Patterns of recurrence and management of the difficult case. Arch Otolaryngol Head Neck Surg 1991;117:546-53.
14. Schefter RP, Olsen KD, Gaffey TA. Cervical lymphangioma in the adult. Otolaryngol Head Neck Surg 1985;93:65-9.
15. Aslan İ, Başerer N, Yenice H, Yazicioğlu E, Başaran B. Postpubertal parotis lenfanjiomları. KBB ve Baş Boyun Cerrahisi Dergisi 2000;8:226-9.
16. Tibesar RJ, Rimell FL, Michel E. Cystic hygroma of the skull base. Arch Otolaryngol Head Neck Surg 1999;125:1390-3.
17. Mathur NN, Rana I, Bothra R, Dhawan R, Kathuria G, Pradhan T. Bleomycin sclerotherapy in congenital lymphatic and vascular malformations of head and neck. Int J Pediatr Otorhinolaryngol 2005;69:75-80.
18. Ozen IO, Moralioglu S, Karabulut R, Demirogullari B, Sonmez K, Turkyilmaz Z, et al. Surgical treatment of cervicofacial cystic hygromas in children. ORL J Otorhinolaryngol Relat Spec 2005;67:331-4.
19. Ikarashi T, Inamura K, Kimura Y. Cystic lymphangioma and plunging ranula treated by OK-432 therapy: a report of two cases. Acta Otolaryngol Suppl 1994;511:196-9.
20. Ogita S, Tsuto T, Nakamura K, Deguchi E, Iwai N. OK-432 therapy in 64 patients with lymphangioma. J Pediatr Surg 1994;29:784-5.
21. Sanlialp I, Karnak I, Tanyel FC, Senocak ME, Buyukpamukcu N. Sclerotherapy for lymphangioma in children. Int J Pediatr Otorhinolaryngol 2003;67:795-800.
22. Katsuno S, Ezawa S, Minemura T. Excision of cervical cystic lymphangioma using injection of hydrocolloid dental impression material. A technical case report. Int J Oral Maxillofac Surg 1999;28:295-6.
23. Molitch HI, Unger EC, Witte CL, vanSonnenberg E. Percutaneous sclerotherapy of lymphangiomas. Radiology 1995;194:343-7.
24. Kennedy TL, Whitaker M, Pellitteri P, Wood WE. Cystic hygroma/lymphangioma: a rational approach to management. Laryngoscope 2001;111:1929-37.
25. Di Carlo I, Gayet B. Lymphangioma of the diaphragm (first case report). Surg Today 1996;26:199-202.