Thoracic duct cysts: A rare differential diagnosis

FRANK GOTTWALD, MD, HEINRICH IRO, MD, CARSTEN FINKE, MD, and JOHANNES ZENK, MD, Erlangen, Germany

OBJECTIVE: **Cysts of the thoracic duct located in the supraclavicular region are uncommon. To date only 12 cases in this topographic area have been described in the literature. Between 1998 and 2002, 5 patients presented to our department with the primary symptom of a palpable soft left-supracavicular swelling that could be displaced relative to adjacent structures.**

SETTING: **In each case, sonography showed a hypoechogenic, almost echo-free, distinctly outlined polycyclic structure with distal echo enhancement at the junction of the left internal jugular vein and the subclavian vein. All 5 patients underwent surgery, the cysts were extirpated, and the numerous communicating lymph vessels localized and meticulously ligated. Pathohistologic analysis of the milky, yellowish fluid obtained by intraoperative puncture confirmed the initial suspicion of a thoracic duct cyst in all patients.**

CONCLUSION: **In the case of left supraclavicular masses, the rare differential diagnosis of a thoracic duct cyst must be considered as a possibility. Sonography as the imaging method of choice is sufficient for primary diagnosis. In addition, a thorax x-ray should be performed in order to exclude an intrathoracic involvement. Surgical extirpation marks the therapy of choice in treating such cysts.**
(Otolaryngol Head Neck Surg 2005;132:330-3.)

Cervical swelling is known to have a variety of possible causes. Apart from malignancies, lymphadenitis, lymphoma and lateral, as well as medial cysts of the neck, more rarely occurring disorders such as lipoma or cystic hygroma must also be taken into consideration. Thoracic duct cysts represent a rare differential diagnosis of the head and neck space occupying lesions.

In literature, one finds reports on only 11 cases of left-lateral thoracic duct cysts and merely a single report on a right-lateral case.[1] We report here on the patient history and problems encountered in the diagnosis and therapy of 5 patients presenting at our department between 1998 and 2002.

CASE DESCRIPTIONS

The 5 patients had a mean age of 45.2 years (24, 38, 49, 47, and 68 years.) and included 3 males and 2 females. Age, gender, symptoms, and cyst localization are listed separately in Table 1.

No previous underlying disorders had been determined at initial presentation or in the patient histories. All patients initially presented with painless left-supraclavicular swelling that was perceived as cosmetically disturbing. Symptoms had persisted for 3 to 6 months, on average for 4.2 months. In 2 patients, increased swelling had been observed during physical examination; another patient complained of an unspecific pressure sensation in the left region of the head and shoulder. All patients had been referred to the clinic for further diagnosis and therapy after initial suspicion of a lateral cyst of the neck.

At initial presentation, all patients showed a soft, indolent, partly fluctuating swelling that was mobile and compressible relative to the subcutaneous tissue and adjacent structures. Sonography showed hypoechogenic to echo-free masses of different dimensions (4 to 10 cm; mean, 6.5 cm) located in the area of the junction of the left internal jugular and sublavian vein, having no discernible connections to these vessels though (Fig 1). Distinct distal echo enhancement was found in each case. In general, the internal structure was homogeneous. In one case, hyperechogenic areas indicating septuations were noted. All of the spatial masses were sonocompressible, sharply outlined, and polycyclic. No internal perfusion was found in any of the patients by color-coded Doppler or power-mode sonographic investigation.

The established findings were in agreement with externally conducted magnetic resonance (1 case, Fig 2) and computed tomography (3 cases) investigations.

We refrained from performing preoperative fine-needle aspiration because of the unambiguous sonographically evidenced cystic structure. Moreover, we aimed to avoid provoking an iatrogenic infection and restricting the possibility of intraoperative preparation.

All patients underwent surgery because of a differentially diagnosed, untypically located lateral cyst of the neck of unclear origin. All interventions involved general anesthesia. Surgical access was attained by

From the Department of Otolaryngology–Head and Neck Surgery, University Erlangen-Nuremberg, Erlangen, Germany.

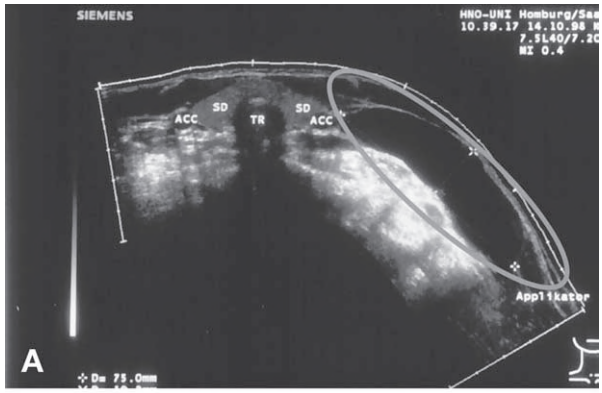
Reprint requests: Frank Gottwald, MD, Waldstr. 1, D-91054 Erlangen, Germany; e-mail, Frank.Gottwald@hno.imed.uni-erlangen.de.

0194-5998/$30.00

doi:10.1016/j.otohns.2004.09.002

Table 1. Seventeen cases of cervical thoracic duct cysts or of right-lateral lymphatic duct cysts

Gender/age	Localization	Symptomatology	Authors	Year
F/18©	Right©	Asymptomatic©	Steinberg[7]©	1964
F/76©	Left©	Asymptomatic©	Barlow@and©Gracey[8]©	1965
F/28©	Left©	Supraclavicular@pain©	Steinberg@and©Watson[11]©	1966
F/55©	Left©	Asymptomatic©	Kolbenstvedt@and©Clanese[12]©	1986
M/17©	Left©	Asymptomatic©	Arnault@t@al[13]©	1990
F/63©	Left@to@mediastinal©	Asymptomatic©	Sakamoto@t@al[14]©	1991
M/56©	Left©	Asymptomatic©	Wax@and©Treolar[15]©	1992
F/64©	Left©	Lump@sensation©	Masuda@t@al[10]©	1992
M/59©	Left@to@mediastinal©	Apnea,@swelling©	Okazaki@t@al[16]©	1996
F/28©	Left©	Asymptomatic©	Maruyama@t@al[17]©	1997
F/49©	Left©	Swelling@of@neck©	Mattila@t@al[1]©	1999
M/52©	Left©	Supraclavicular@swelling©	Gomez@t@al[18]©	2001
F/38	Left	Supraclavicular swelling	presented cases	1998
M/49	Left	Swelling and pressure sensations		2000
M/47©	Left©	Swelling@of@neck©		2001
F/24	Left	Supraclavicular swelling		2002
M/68	Left	Asymptomatic		2002



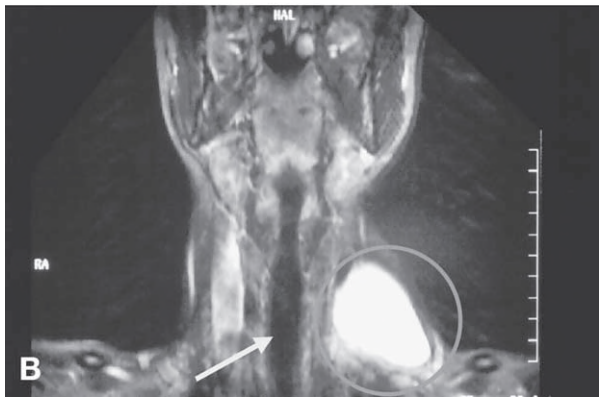


Fig 1. A, Sonographic image of a thoracic duct cyst in a horizontal plane (patient aged 49 years). Distinctly outlined echo-free mass (+. . .+) with distal echo enhancement at the junction of the internal jugular and subclavian vein (dimensions: 75 × 18 mm). (*ACC*, common carotid artery; *SD*, thyroid gland; *TR*, trachea) **B,** T2-weighted noncontrast MRI of a thoracic duct cyst in a female patient (age 38). Signal-dense representation of the cyst, smoothly outlined in the angle formed by the veins (*arrow,* trachea).

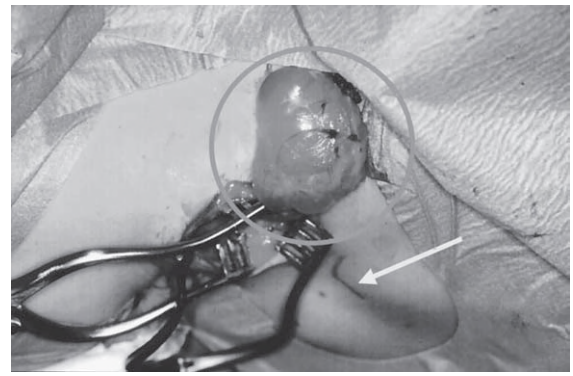


Fig 2. Extirpated cyst at the operation-site in a female patient (age 24) (*arrow*, jugulum).

transversal incision in a skin fold located approximately 2 to 3 cm above the clavicle. The intraoperative state of the cyst was firmly elastic and shimmering yellow. The number of multiple lymphatic ducts (>5-7) connecting the cyst with the surrounding lymph vessel system was highly conspicuous. All the thoracic duct cysts were located in the angle formed by the internal jugular vein and the subclavian vein during exposure as well (Fig 3). In the ventral direction, the divided insertion of the sternocleidomastoid muscle, and in the dorsal direction, the muscles constituted adjacent structures that had to be exposed.

The wall of the cyst was conspicuously thin compared with the walls of the communicating vessels. After careful ligature of all connected lymphatic ducts, the cysts could be removed in toto.

No connection to the venous system was discernible in any of the cases. Three of the cysts were punctured intraoperatively so as to obtain a better surgical field of

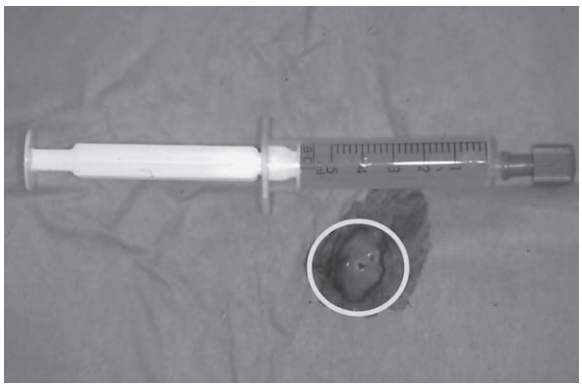


Fig 3. Postoperative cyst after needle aspiration of contents in a 68-year-old patient. Typical yellowish milky secretion in the syringe.

view and to facilitate exposure. The retrieved secretion was slightly milky and amber-colored (Fig 3). We dispensed with further lab analysis of the punctured fluid, since the surgical preparation was routinely submitted for pathologic and histologic work-up.

Pathohistologic analysis confirmed the presence of a thoracic duct cyst in all cases. After meticulous hemostasis a 2-layer wound closure was performed and a redon drain left in place for 2 days.

Wound healing remained without complications in all 5 patients. Follow-up examinations at intervals of 1 to 6 months yielded no clinical or sonographic indication of a recurrence.

DISCUSSION

The thoracic duct originates from the cisterna chyli that drains the small lymphatic ducts of the intestine. It runs upward, paramedially along the right side of the spine, to the level of the fifth or sixth thoracic vertebra. Here it crosses over to the left side and then runs between the aorta and the azygos vein. It enters the left side of the neck dorsal to the innominate artery and arches 3 cm above the clavicle anterior to the phrenic nerve, the thyreocervical trunk and the subclavian artery. In most cases, the duct lies laterodorsally to the common carotid artery, vagus nerve, and internal jugular vein and empties into the junction of the subclavian and internal jugular vein.[1]

The wall of this lymphatic vessel is relatively thick in the thoracic region, consisting of collagenous and elastic fibers with some longitudinally aligned smooth muscle fibers. The cervical portion consists of thin subendothelial connective tissue without elastic lamina and contains only sparsely distributed thin smooth muscle fibers.[1]

The flow of lymph or chylus in the thoracic duct depends on several factors including the variable intrathoracic respiratory pressure conditions, transmitted pulsations from adjacent vessels, and active peristaltic contractions of the duct itself.

The pathogenesis of thoracic duct cysts is unclear. In all 5 patients the cyst was very thin-walled macroscopically, and there were no signs of inflammation or infection. Intraoperatively, it was not possible to expose a direct inlet of the duct into the internal jugular vein. In our opinion, the most probable causes of cyst formation therefore appears to be obstruction of lymphatic drainage in the angle formed by the internal jugular vein and the subclavian vein, either exclusively or in combination with weakness of the wall of the thoracic duct.

Congenital weakness of the ductal wall and degeneration induced by trauma, infection or other inflammation processes have been discussed in literature.[1] Stenoses obstructing drainage in the vein angle may also predispose to cyst formation.

Four of the 12 (23.5%) patients known to have cervical thoracic duct cysts exhibited symptoms other than painless left-supraclavicular swelling (Table 1). These symptoms were caused by compression of adjacent structures and ranged from unspecific cervical pressure sensations via lump sensation in the hypopharynx to apnea.[1-10] Ten of the patients were female. However, no statistical relevance can be established because of the low number of cases.

A conspicuous finding was that only a single case of a right-lateral cyst of the right lymphatic duct has been described in literature.[7] Here one could assume that cyst formation may well be dependent on the amount of transported lymph. But again, the number of cases is too low to provide substantiated statistical proof.

A thorough patient history and clinical examination are essential in establishing a diagnosis. Comparison of different imaging methods has shown that reliable diagnosis and determination of the extension of cervical cysts can be sufficiently achieved by sonography resting in the hands of an experienced investigator. Further imaging modalities furnish no additional information in the case of cervical cysts of the thoracic duct and are thus without prognostic relevance. If an intrathoracic extension or the presence of secondary intrathoracic or abdominal cysts are suspected, however, computed tomography and/or magnetic resonance tomography with contrast medium may well be indicated. For screening purposes, a thorax x-ray is advisable in order to exclude an expanded mediastinum. None of our cases showed a sign of intrathoracic involvement.

In the literature, lymphography has been described as a method for determining extension. Such reports date from before the era of MRI, however. Today, these investigations are no longer indicated.[1-10]

In our view, preoperative puncture of the cysts for lab-chemical analysis of the chylus is not generally indicated. Moreover, there is the risk of fistulation and infection. Differential diagnoses in the context of thoracic duct cysts certainly include, apart from malignancies, untypical lateral and medial cysts of the neck as well as lymphangioma and cystic hygroma.

The therapy of choice is the total excision of the cyst. There is no report of nonoperative management of these neoplasms. In our view, this fact depends on the patient's demand of getting free of the supraclavian mass. However, only histologic examination is able to assure the benign character of these neoplasms. Nevertheless, a monitoring of patients by ultrasound over months to evaluate the progress or stability of the process is a possible alternative option.

Typical complications reported in the literature are wound-healing problems, inflammations, and scars and loss of sensation in the surrounding skin. Neither pneumothorax nor any other severe complications have been described. One important possible risk is fistulation of chylus that sometimes appears for example within neck dissections. Obviously, no case of chylus fistula is reported in the literature up to now. Intraoperatively, especially careful ligation of all communicating lymphatic vessels is essential to avoid a possible chylus fistula. Complete ligation of the entire thoracic duct is possible without any associated co-morbidity being known, because multiple collateral connections to the hemiazygos vein exist intrathoracically, which can provide lymphatic drainage. Other possibilities of complications are injuries of surrounding nerves (eg, vagal and phrenic nerve), brachial plexus paresis or bleeding intra- or postoperatively from the common carotic artery, the thyreocervical trunk, and the subclavian artery or corresponding veins.

In our patients, we encountered only a complication-free development without recurrences over a follow-up period of 6 months.

CONSEQUENCES FOR CLINICAL PRACTICE

Thoracic duct cysts represent a rare differential diagnosis in supraclavicular swelling of the neck. Further symptoms do not necessarily have to occur. The patient history and a thorough physical examination are preeminent in establishing the diagnosis. Diagnostic B scan and Doppler sonography mark the method of choice and definitive strategy in diagnostic imaging.

We would like to thank Mr Stuart Irwin for revising the English manuscript.

REFERENCES

1. Mattila PS, Tarkkanen J, Mattila S. Thoracic duct cyst: a case report and review of 29 cases. Ann Otol Rhinol Laryngol 1999; 108(5):505-8.
2. Hori S, Harada K, Morimoto S, et al. Lymphangiographic demonstration of thoracic duct cyst. Chest 1980;78:652-4.
3. Morettin LB, Allen TE. Thoracic duct cyst: diagnosis with needle aspiration. Radiology 1986;161:437-8.
4. Adachi A, Watanabe J, Kurohiji T, et al. A case report of thoracic duct cyst. Nippon Kyobu Geka Gakkai Zasshi 1989;37:1586-9.
5. Okabe K, Miura K, Konishi H, et al. Thoracic duct cyst of the mediastinum. Scand J Thorac Cardiovasc Surg 1993;27:175-7.
6. Hvid.Jacobsen K, Nielsen SL, Jensen VJ, et al. Demonstration of the thoracic duct by 123-I heptadecanoic acid: report of a case. Acta Radiol 1987;28:783-4.
7. Steinberg I. Roentgen diagnosis of persistent jugular lymph sac. Radiology 1964;82:1022.
8. Barlow D, Gracey L. Cystic dilatation of the thoracic duct. Br J Clin Pract 1965;19:101-2.
9. Fuchs WA. Normal radiologic anatomy of the lymphatics and lymph nodes. In: Abrahams HL, ed. Abraham's angiography: vascular and interventional radiology. Boston: Little, Brown; 1983. pp. 1997-2002.
10. Wax MK, Teolar ME. Thoracic duct cyst: an unusual supraclavicular mass. Head Neck 1992;14:502-5.
11. Steinberg I, Watson RC. Lymphangiographic and angiographic diagnosis of persistent jugular lymph sac: report of a case. N Engl J Med 1966;275:1471-4.
12. Kolbenstvedt A, Aanese J. Cystic dilatation of the thoracic duct presenting as a supraclavicular mass. Br J Radiol 1986;59:1228-9.
13. Arnault G, Winants D, Charles M, et al. Cystes du canal thoracique. A prospes d′une localisation sus-claviculaire. J Radiol 1990;71:61-4.
14. Sakamoto H, Uda H, Sato A, et al. Thoracic duct cyst of the neck: a case report. Lymphology 1991;24:130-4.
15. Masuda R, Furuhata Y, Kasahara D, et al. Preoperative diagnosis of a thoracic duct cyst arising in the supraclavicular fossa: surgical case report. Nippon Kyobu Geka Gakkai Zasshi 1992; 40:320-5.
16. Okazaki T, Shinkai M, Fukiwara H, et al. Thoracic duct cyst—a case report. Nippon Kyobu Geka Gakkai Zasshi 1996;44:830-4.
17. Maruyama M, Kobayashi S, Kasuga Y, et al. Thoracic duct cysts in supraclavicular region. Ulster Med J 1997;66(2):140-3.
18. Gomez E, Tejerina E, Martorell V, et al. Thoracic duct cyst presenting as an hour-glass shaped mass in the left supraclavicular area: case report. Acta Otorhinolaryngol Belg 2001;55(3): 223-5.