Airway obstruction caused by rapid enlargement of cervical lymphangioma in a five-month-old boy

Junji Shimizu¹, Takashi Taga², Takuma Kishimoto¹, Motoki Ohta², Kouji Tagawa², Tomoaki Kunitsu², Tetsunobu Yamane¹, Yasuyuki Tsujita¹, Yoshihiro Kubota² & Yutaka Eguchi¹

¹Department of Emergency and Intensive Care Unit, Shiga University of Medical Science, Otsu, Shiga, Japan
²Department of Pediatrics, Shiga University of Medical Science, Otsu, Shiga, Japan
³Department of Pediatric Surgery, Shiga University of Medical Science, Otsu, Shiga, Japan

Correspondence
Junji Shimizu, Department of Emergency and Intensive Care Unit, Shiga University of Medical Science, Seta Tsukinowa-cho, Otsu, Shiga 520-2192, Japan. Tel: +81-77-548-2929; Fax: +81-77-548-2929; E-mail: jushimi77@gmail.com

Funding Information
No sources of funding were declared for this study.

Received: 23 March 2016; Revised: 13 May 2016; Accepted: 24 July 2016

Clinical Case Reports 2016; 4(9): 896–898
doi: 10.1002/ccr3.659

Introduction

Lymphangiomas are rare congenital malformations of the lymphatic system that tend to affect the head and neck [1]. These lesions are histologically benign and usually asymptomatic; therefore, they are usually followed up in the outpatient department. However, life-threatening complications including airway obstruction can arise due to sudden enlargement of a lymphangioma by hemorrhage or infection [1]. Percutaneous sclerotherapy for lymphangioma, such as that by intralesional injection of OK-432, has recently been proposed as an alternative to surgical resection [2]. However, sclerotherapy may take a long time to exhibit an effect, and airway obstruction may occur due to temporary enlargement of the lesion by inflammation. In this report, we describe a five-month-old boy with cervical lymphangioma who needed emergency tracheal intubation because of rapid enlargement of the tumor by infection and underwent prophylactic elective tracheostomy for tracheal compression.

Case Report

A five-month-old boy was brought to the emergency department with fever and stridor associated with rapid growth of a neck mass. One month before presentation, a soft mass had been found on the left side of his neck, and cervical lymphangioma was diagnosed by computed tomography and magnetic resonance imaging. He had no respiratory symptoms and was followed as an outpatient. One day before presentation, he developed a fever of 39°C and stridor when crying. On arrival, his oxygen saturation was 99% and stridor was noted with crying, but no effort was observed while breathing. He was admitted to the hospital, and antibiotics were administered. However, the stridor worsened and his oxygen saturation declined to 80% despite oxygen administration; therefore, he was intubated and ventilated. Fentanyl, midazolam, and rocuronium were administered for induction of anesthesia. A 3.5-mm tracheal tube was inserted. Chest and neck computed tomography revealed a large cystic mass that compressed the trachea (Fig. 1). Fine needle
aspiration of the cyst was performed, and 20 mL of ser-
ous liquid was removed; however, the cervical swelling
did not improve. Intralesional injection of OK-432 was
planned, but there was concern that transient swelling
due to the local inflammatory response after OK-432
injection would cause airway obstruction and prolonged
intubation. Moreover, after OK-432 injection and tran-
sient swelling, reintubation seemed likely to be more dif-
cult if accidental extubation occurred. Therefore,
tracheostomy was performed by an otolaryngologist, and
OK-432 was then injected. After transient swelling and
erthema, the mass markedly decreased in size. Three
weeks after OK-432 injection, the tracheostomy tube was
removed.

**Discussion**

Lymphangiomas are rare congenital malformations of the
lymphatic system that affect 1 in 6000–16,000 live-born
neonates. Approximately 75% of lymphangiomas occur in
the head and neck region [3]. They are histologically
benign and commonly asymptomatic, so affected patients
are often followed up as outpatients without any treat-
ment. However, lymphangiomas of the neck can cause
life-threatening complications, including airway obstruc-
tion [1]. Patients can present to the emergency depart-
ment with airway obstruction resulting from sudden
enlargement of a lymphangioma by hemorrhage or infec-
tion. Respiratory distress secondary to a lymphangioma
can progress rapidly, and intervention to secure the air-
way should be considered immediately. In this case, the
respiratory distress progressed rapidly, and the patient
required emergency tracheal intubation. In such cases of
upper airway obstruction, the clinician should consider
having the anesthesiologist intubate the patient and avoid
using muscle relaxants.

Indications for treatment of lymphangioma are compres-
sion of surrounding structures such as the airway,
cosmetic problems, and recurrent infection [4]. Various
methods are available for treatment of lymphangioma,
including surgical excision and sclerotherapy with bleo-
mycin or OK-432 [2]. Complete surgical excision is the
definitive approach to lymphangioma, but it is often diffi-
cult to achieve because of diffuse lesions and the risk of
damage to important organs [4]. Percutaneous sclerother-
apy has been proposed as an alternative to surgical resec-
tion. Bleomycin has been used, but it must be used with
extreme caution because of the risk of pulmonary fibrosis
[5]. Several previous reports have suggested that intrale-
sional injection of OK-432 should be a first-line treatment
for lymphangioma [2].

Lymphangioma is classified as cystic lymphangioma,
cavernous lymphangioma, and lymphangioma simplex
[6]. It can appear at any site, but cystic lymphangioma is
seen in the cervical region more commonly than is
cavernous lymphangioma. OK-432 is reportedly more
effective for cystic lymphangioma than cavernous lym-
phangioma. Cavernous lymphangioma is often intractable
to medical treatment [7]. Additionally, sildenafil is report-
dedly effective for severe lymphangioma. In cases that are
intractable to medical treatment, the use of sildenafil
might be considered [8]. In the present case, computed
tomography showed a multicellular cystic mass progress-
ing from the left parotid gland to the pharyngeal space.
Serous liquid was removed by fine needle aspiration; the
mass was thus regarded as a cystic lymphangioma, and
OK-432 injection was effective.

OK-432 injection produces a local inflammatory reac-
tion that leads to shrinkage of the lesion [2]. OK-432
injection is safe in the majority of children, but complica-
tions of OK-432 injection have been reported, including
transient pyrexia, swelling, tenderness, severe upper

![Figure 1. Computed tomography showing the multilocular cystic mass progressing from the left parotid gland to the pharyngeal space and compressing the trachea.](image)
Airway obstruction caused by lymphangioma

J. Shimizu et al.

airway obstruction, and enlargement of the mass [4]. When a lymphangioma seen around the trachea or cervical region is treated with intralesional OK-432, the physician should be aware that such airway complications may occur and prepare for possible airway obstruction.

Airway protection is a major indication for tracheostomy [9, 10]. Complications of tracheostomy include granuloma, infection, and bleeding. Granuloma is induced by contact of the airway wall with the tracheostomy tube and increases in size as the duration of tube placement becomes longer. The timing of and indications for tracheostomy in children are still controversial. The reported average mechanical ventilation period before tracheostomy is 26–32 days [9, 11, 12], which is longer than that in our case. On the other hand, two patients with lymphangioma reportedly died of hypoxic damage related to difficulty with reintubation after endotracheal tube displacement [13]. In our case, reintubation seemed difficult when accidental extubation occurred. Securing the airway was given the highest priority in our patient; therefore, prophylactic elective tracheostomy was performed with a shorter period of mechanical ventilation than in previous reports. In cases of cervical lymphangioma with tracheal compression, when reintubation seems difficult or the duration of intubation may be prolonged, securing the airway is most important and prophylactic and elective tracheostomy should be considered.

Conclusion

Physicians should be aware of the risks of airway obstruction caused by sudden enlargement of cervical lymphangioma. Furthermore, because of transient swelling related to injection of OK-432, an intervention to secure the airway may be needed. In patients with tracheal compression, when reintubation seems difficult or the duration of intubation may be prolonged, prophylactic and elective tracheostomy should be considered.

Informed Consent

Informed consent to report this case was obtained.

Conflict of Interests

None declared.

References

1. Kennedy, T. L., M. Whitaker, P. Pellitteri, and W. E. Wood. 2001. Cystic hygroma/lymphangioma: a rational approach to management. Laryngoscope 111:1929–1937.
2. Laranne, J., L. Keski-Nisula, R. Rautio, M. Rautiainen, and M. Airakasinen. 2002. OK-432 (Picibanil) therapy for lymphangiomas in children. Eur. Arch. Otorhinolaryngol. 259:274–278.
3. McGill, T., and J. B. Mulliken. 1993. Vascular anomalies of the head and neck. Otolaryngol. Head Neck Surg. 1:333–346.
4. Hall, N., N. Ade-Ajayi, C. Brewis, D. J. Roebuck, E. M. Kiely, D. P. Drake, et al. 2003. Is intralesional injection of OK-432 effective in the treatment of lymphangioma in children? Surgery 133:238–242.
5. Acevedo, J. L., R. K. Shah, and S. E. Brietzke. 2008. Nonsurgical therapies for lymphangiomas: a systematic review. Otolaryngol. Head Neck Surg. 138:418–424.
6. Landing, B. H., and S. Farber. 1956. Tumor of the Cardiovascular system, Atlas of Tumor Pathology, Section III, Fascicle 7. 124–138.
7. Okazaki, T., S. Iwata, T. Yanai, H. Kobayashi, Y. Kato, T. Marusasa, et al. 2007. Treatment of lymphangioma in children: our experience of 128 cases. J. Pediatr. Surg. 42:386–389.
8. Swetman, G. L., D. R. Berk, S. S. Vasanawala, J. A. Feinstein, A. T. Lane, and A. L. Bruckner. 2012. Sildenafil for severe lymphatic malformations. N. Engl. J. Med. 364:385–386.
9. Dursun, O., and D. Ozel. 2011. Early and long term outcome after tracheostomy in children. Pediatr. Int. 53:202–206.
10. Da Silva, P. S. L., J. Waisberg, C. S. T. Paulo, F. Colugnati, and L. C. Martins. 2005. Outcome of patients requiring tracheostomy in a pediatric intensive care unit. Pediatr. Int. 47:554–559.
11. Karapınar, B., M. T. Arslan, and C. Özcan. 2008. Pediatric bedside tracheostomy in the pediatric intensive care unit: six-year experience. Turk. J. Pediatr. 50:366–372.
12. Graf, J. M., B. A. Montagnino, R. Hueckel, and M. L. McPherson. 2008. Pediatric tracheostomies: a recent experience from one academic center. Pediatr. Crit. Care Med. 9:96–100.
13. Kitagawa, H., H. Kawase, M. Wakisaka, Y. Satou, H. Satou, S. Furuta, et al. 2004. Six cases of children with a benign cervical tumor who required tracheostomy. Pediatr. Surg. Int. 20:51–54.