Case Report

A Case of Head and Neck Lymphatic Malformation in which Sclerotherapy with Polidocanol was Useful for Hemostasis

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ABSTRACT

Lymphatic malformations that occur in the head and neck are often difficult to treat. Sclerotherapy using polidocanol, which has little tissue damage, can be treated with minimal dysfunction and complications. In particular, it is considered an effective treatment for superficial lesions that bleed from the mucosal surface. In this study, we performed sclerotherapy using polidocanol for a 17-year-old woman with lymphatic malformations with blood containing lymphorrhea in the pharynx and larynx. A total of 3 sclerotherapy treatments were performed, and lesion reduction and bloody lymphorrhea control were observed. At 1 year after treatment, the course of treatment was good with no re-enlargement of the lesions.

Key words: bleeding, head and neck, lymphatic malformations, polidocanol, sclerotherapy

Introduction

Treatment of head and neck vascular malformations is often difficult due to anatomical features. Lymphatic malformations that develop in the head and neck often cause upper airway stenosis due to increased lesions. Superficial lymphatic malformations with lesions on the skin and mucous membranes can cause hemorrhage and lymphorrhea with minor irritation. We experienced a case of sclerotherapy using polidocanol for lymphatic malformations that occurred in the respiratory tract, in which the superficial bleeding was successfully controlled.

Case report

A 17-year-old female had lymphatic malformations of the pharynx and larynx since birth. Sclerotherapy with OK-432 was performed at the age of 0. However, the lesion was not reduced, and laryngeal stenosis and leakage of bloody lymph into the trachea were confirmed. A tracheotomy was performed at 3 months after birth. Subsequently, 4 sessions of sclerotherapy with OK-432, CO₂ laser ablation, and partial resection were performed, but the lesion did not improve. Therefore, she required airway management with tracheostomy. In addition, the presence of bloody lymphorrhea rendered her school life difficult. She was introduced to our department when she was 17 years old.

At the first consultation, swelling of the left neck and outflow of bloody lymph from the tracheal incision were observed. On pharyngolaryngoscopy, punctate blood vessels were found in the mucous membranes and lesion from the pharynx to the oral cavity, and it was not possible to pass the fiberscope through the larynx (Fig. 1). Contrast-enhanced CT showed soft shadows in the larynx, bilateral parapharyngeal space, and left submandibular space to the sublingual space and also depicted a pharyngolaryngeal lesion with slight enhancement but no apparent vascular space within it (Fig. 2). Based on these findings, we made the diagnosis of lymphatic malformations of the pharynx and larynx, including slight
venous malformations.

A total of 3 sessions of sclerotherapy with polidocanol were performed by collaboration with otolaryngologists, radiologists, and plastic surgeons in our hospital. After spreading the larynx with a laryngoscope, the lesion was punctured with an endoscopic puncture needle (23G), and the extent of the lesion was confirmed with a small amount of contrast medium. A solution of 3% polidocanol (Polidocasclerol®, Kaigen Pharma) was mixed with air at a ratio of 1:4 to form foam and used as a sclerosing agent. The first and second sclerotherapy reduced the lesion. At the second treatment, a biopsy was performed. Vascular cells in the lesion were positive for CD31 and D2-40 but negative for CD34 and were diagnosed as lymphatic malformations. Two months after the second sclerotherapy, the lesions showed intermittent superficial bleeding and a tendency to increase again. The source of bleeding was the superficial layer of the pharyngeal and laryngeal lesions, which flowed out of the tracheostomy to the body surface. A third sclerotherapy was performed to control bleeding. The injection was shallower than that in the previous treatment (Fig. 3). The superficial bleeding stopped, and there was no outflow from the tracheostomy to the body surface. There were no complications after treatment. One year after the third treatment, the lesion was stable without bleeding or enlargement (Fig. 4).

Discussion

Lymphatic malformation is a vascular anomaly that was previously called lymphangioma or hygroma, though it is not a neoplastic lesion. Currently, it is often referred to as a lymphatic malformation, according to the Society for the Study of Vascular Anomalies (ISSVA) classification\(^1\).

Vascular malformations tend to increase slowly and asymptptomatically. When they occur in the head and neck, they can give rise to psychological and social challenges\(^2\). They can also cause life-threatening problems due to the potential for massive hemorrhage. About 70% of lymphatic malformations occur in the head and neck\(^3\). When the lesion is present in the floor of the mouth, pharynx, or larynx, the airway is
compressed and upper airway stenosis occurs. Among the lymphatic malformations that occur in the head and neck, 65% require tracheostomy. If the lesion is present on the surface of the skin or mucous membrane, bleeding with slight stimulus and repetitive bouts of acute swelling and inflammation can occur. Lymphatic malformations with chronic inflammation show increased blood and vascular hyperplasia, resulting in increased internal bleeding. Bleeding occurs in 35% of lymphatic malformations in the head and neck. Therefore, airway management and bleeding control are important for lymphatic malformations that occur in the head and neck. In the case presented herein, lymphatic malformations were observed in the pharynx and larynx from the neonatal period and were difficult to treat. Tracheotomy and airway management were performed, and it is thought that the bleeding was caused by blood vessel growth due to chronic and long-term inflammation, not venous malformation. Blood containing lymphorrhea from the lesion flowed out through the tracheostomy to the body surface, causing a cosmetic problem.

The treatment of lymphatic malformations is broadly divided into surgical excision, sclerotherapy, and medical...
Treatment. In particular, sclerotherapy is considered for widespread and unclear lesions. OK-432, bleomycin, absolute ethanol, STS, doxycycline, and polidocanol are used as sclerosing agents\(^5\-\!)\(^9\), and they are selected with consideration of their specific characteristics. Among them, polidocanol is one of the hardeners that can be used safely because it is associated with less tissue damage and few complications\(^10\). Therefore, it has a relatively wide range of uses and is used to stop bleeding from the mucosa caused by vascular lesions in the digestive tract and trachea.

In this case, surgical resection, CO\(_2\) laser ablation, and sclerotherapy with OK-432 were performed but were all ineffective and the patient had to undergo tracheostomy. Superficial bleeding from the lesion was also observed. In this study, sclerotherapy with polidocanol was performed for the purpose of lesion reduction and hemostasis. Owing to the superficial bleeding, a sclerosing agent had to be injected into the small space in the surface layer of the lesion. As a result, the lesions were reduced and superficial bleeding was controlled without any tissue damage or complications.

The head and neck have complex anatomical structures, with important organs concentrated in a narrow area, making it difficult to select the appropriate treatment for vascular malformations that occur there. Radical resection may cause dysfunction due to tissue defects\(^3\), and even when sclerotherapy is selected, mucosal necrosis may occur due to damage caused by the sclerosing agent\(^5\). Sclerotherapy with polidocanol may be one of the most effective options. In particular, in cases of superficial mucosal hemorrhage, as in the present case, we can expect that bleeding can be controlled without damaging the surrounding tissues by injecting a small amount of fluid into the superficial cavity.

**Conclusion**

Sclerotherapy using polidocanol demonstrated effectiveness in the treatment of lymphatic malformations in the pharynx and larynx presenting with superficial bleeding.

**Statement of ethics**

Written consent was obtained from the patient. The ethics committee of the institution approved the use of polidocanol (#20120476) and the retrospective data (#20160049).

**COI statement**

There is no conflict of interest to declare.

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