Case report

A case of postoperative nasopharyngeal reflux associated with retropharyngeal lymphangioma in newborn infant

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Abstract

Lymphangioma is a rare benign congenital tumor of the lymphatic system, which is commonly diagnosed before 2 years of age. In the natural report, cystic lymphangioma was usually reported as a huge translucent mass located in the head and neck area. It’s occurrence in retropharyngeal space with respiratory obstruction and swallowing difficulty in neonate is extremely rare and postoperative nasopharyngeal reflux has rarely been reported. Complete resection is the standard therapy. However, involvement of the upper airway may be determining prognosis in the extensive lymphangiomas because of the difficulty of complete excision. We present a case of cystic lymphangioma in neonate which was initially asymptomatic but gradually progressed to cause respiratory obstruction due to enlargement. After resection, nasopharyngeal reflux developed with dysfunction of the soft palate and gradually improved with conservative care over 5 months. (Korean J Pediatr 2010;53:258-261)

Key Words: Cystic Lymphangioma, Deglutination disorder, Excision, Pharyngeal Diseases

Introduction

Lymphangioma is a benign congenital tumor of the lymphatic system which involves chiefly the head and neck. But retropharyngeal lymphangioma with respiratory obstruction and swallowing difficulty is extremely rare in a neonate. Retropharyngeal lymphangioma are mostly asymptomatic. But occasionally obstructive symptoms, failure to thrive and respiratory distress may be present when the tumors extend into the upper airway and oral cavity and these symptoms sometimes rarely sustain after operation. Complete excision is the best choice for treatment of lymphangioma. However, it is difficult to execute complete excision of the retropharyngeal lymphangioma around the upper airway. In this paper, we present a case of retropharyngeal lymphangioma with postoperative nasopharyngeal reflux in a 14 day-old neonate.

Case report

A male baby was born at 40+1 weeks of gestation by normal vaginal delivery. The birth weight was 3,280 g. His mother had received prenatal screening check-ups routinely without any specific findings. Immediately after birth, he had transient tachypnea and intermittent vomiting but improved in several hours. Initial physical examination revealed no specific abnormal findings. However, on the eleventh day after birth, the patient had an episode of sudden desaturation and continued to display chest retractions, coarse breath sounds and stridor. We intubated the patient for respiratory care and enhanced computed tomography (CT) scan of the neck was performed for the evaluation of airway on the fourteenth day. CT scan of the neck showed a large retropharyngeal cystic mass which highly suggested the possibility of cystic lymphangioma. The cystic lesion was measured 3.1×2.9×1.1 cm. The mass was mainly located in anteroinferior aspect of the clivus reaching the...
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The anterior portion of C4 body with compression of retropharyngeal and pharyngeal airway and extended along the left parapharyngeal space and anterior aspect of left carotid space to the left lower neck, 2.2 cm in length (Fig. 1A). On the same day, surgical excision was performed under general anesthesia via transoral approach. Bulging mass was exposed on the retropharyngeal area from nasopharynx level to oropharynx level. Its capsule was removed easily with an incision over the cyst. Yellowish clear discharge was sucked out, and the tissue pathology confirmed the lesion as a cystic lymphangioma. A follow-up CT scan of the neck was done on the next day. The previously noted retropharyngeal cystic lesion had disappeared at the central neck portion (Fig. 1B).

Because post-operative oral feedings were unsuccessful, accompanied with sustained vomiting and dysphagia, we additionally performed an esophagography. The initial esophagography showed mild nasoopharyngeal reflux with aspiration of contrast from the oropharynx to the nasopharynx and signs of dysfunction of the soft palate without a tracheoesophageal fistula (Fig. 2A). The feeding was supplied entirely by a feeding tube.

After discharge, a series of esophagography were done for nasoopharyngeal reflux. Two months after operation, follow-up esophagography still visualized minimal aspiration in the side of trachea (Fig. 2B). However, the final follow-up esophagography at 5 months after operation showed no evidence of nasoopharyngeal reflux, and the patient was tolerable to the oral feedings (Fig. 2C). In addition, echocardiography, neurosonography and abdominal sonography showed no specific findings.

Discussion

Lymphangioma is a rare benign congenital tumor of the lymphatic system, which is classified based on the size of the endothelial cell-lined lymphatic sinuses histologically: Capillary lymphangioma consist of small capillary-sized lymphatic vessels, cavernous lymphangioma contain large dilated lymphatic vessels and cystic lymphangioma contain large macrocystic spaces. Our case was diagnosed as cystic lymphangioma on the ground of tissue pathology and on the radiologic finding of cystic mass occupying space.

Lymphangioma can appear in any site, but the cystic lymphangiomas usually appear as large soft tissue masses with translucency on the exposed areas of the neck and can be easily discovered early in life even though main parts was deeply seated in loose connective tissues. However, involvement of the upper airway is rare, and only few cases are reported in retropharyngeal space or isolated to the larynx. In our case the lymphoma involved the upper airway, more specifically the retropharyngeal area. Lymphangioma presents in variable sizes. Small and unnoticed lymphangioma is only presented later with secondary symp-

![Fig. 1. (A) A cervical CT scan shows a large retropharyngeal cystic mass with a low attenuation density, measured at 3.1×2.9×1.1 cm and mainly located from anteroinferior aspect of clivus to anterior portion of C4 body with compression of retropharyngeal and pharyngeal airway. (B) Postoperative cervical CT shows resected lesion of the cystic mass with air shadow and postsurgical fluid collection.](image-url)
toms depending on their size or locations. In the case of infection, trauma or hemorrhage, the cyst can rapidly grow in size in several days, however, it usually grows slowly in proportion to the growth of the infant. Respiratory obstruction and swallowing difficulty, which are the second and third most common symptoms take place when mass extends to the oral cavity, pharynx and larynx. Also isolated tongue involvement induce symptoms of macroglossia, dysphagia and airway obstruction. Stridor, cyanosis, apnea, dysphagia with failure to thrive, these symptoms appear when cysts extend into thoracic inlet, mediastinum \(3, 4, 6\). Dyspnea occurs in 11% to 27% of the patients \(7\). When cystic hygroma is located in the retropharyngeal space, it is not easily detected initially, but may later be detected by secondary symptoms like respiratory obstruction and feeding problem \(3, 4\). Involvement of the upper airway may be a factor determining prognosis because of the difficulty of complete excision \(1, 3, 4, 6, 7\). Even though the new treatment modalities such as sclerotherapy, diathermy, percutaneous aspiration, radiotherapy and systemic cyclophosphamide, the treatment of choice at present is complete radical resection of the mass in order to prevent the recurrence of lymphangiomas \(1, 3, 4, 6\). The recurrence rate depends upon the complexity of the lesion and completeness of excision. For completely removed lesions the recurrence rate was approximately 10–27%, whereas for partially resected lesions, 50–100% \(3\). If possible, early surgery is recommended before developing ambiguous boundary with adjacent tissue. Only neonates with large cervical cystic hygromas causing tracheal compression and respiratory distress require urgent resection \(1, 3, 4\).

In transoral surgery, the rate of postoperative complication reveals in approximately 45% which consist with wound infection (7.4%), infection or leakage of cerebrospinal fluid (11.1%), lower cranial nerve dysfunction (11.1%), palatal problem (14.8%), swallowing problem or speech difficulty (18.5%) and nasopharyngeal reflux (11.1%) \(9\). However
nasopharyngeal reflux almost related with palatal extension of transoral procedure and has rarely been reported in the neonate\(^9\), \(^10\). In the barium study, nasopharyngeal reflux is defined as passage of contrast medium from the oropharynx or hypopharynx into the nasopharynx of the nose in the course of swallowing\(^11\). After transoral procedure, it mainly occur due to velopharyngeal insufficiency, as a result of soft or hard palate injury, structural changes in the posterior pharyngeal wall or large dead space in the posterior pharynx\(^9\), \(^10\). In our case, nasopharyngeal reflux was noted after transoral procedure without palatal extension, not from any anatomic abnormality such as tracheal–esophageal fistula but from dysfunction of soft palate. Spontaneous regression of postoperative nasopharyngeal reflux is mentioned in report of choanal atresia\(^11\), as was found in our case.

We present a rare case of retropharyngeal lymphangioma in a 14-day-old infant. The baby initially was asymptomatic but later manifested stridor and respiratory difficulty, and eventually treated with excision of the mass. After resection, nasopharyngeal reflux which caused postoperative feeding difficulty persisted for a while, it was resolved gradually over 5 months after surgery.

### References