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Case Report

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Retropharyngeal Lymphangioma: A Case Report

Farrokh Heidari, Firouzeh Heidari, Amirhossen Yadegar, Kayvan Aghazadeh and Ebrahim Karimi*

Otorhinolaryngology Research Center, Tehran University of Medical Sciences, Tehran, Iran

***Correspondence to:**
Dr. Ebrahm Karimi, MD
Professor of Ear, Nose and Throat
Otorhinolaryngology Research Center
Amir Alam Hospital, North Sadi Ave
Tehran, Iran
Tel: 02166760269
E-mail: karimient@gmail.com

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Abstract

Lymphangioma is presented as a painless mass with a soft consistency in the head and neck. The involvement of retropharyngeal space in lymphangioma is rare. Here, we presented the case of retropharyngeal lymphangioma and discussed its manifestations, imaging, and treatment. An 11-year-old female was presented with shortness of breath, stridor, fever, dysphagia and drooling. Clinical examinations revealed a retropharyngeal swelling and a hot-potato speech. MRI revealed the multicystic lesions extending from the base of the skull to the level of T4 in retropharyngeal space. The mass resection was done under general anesthesia and 5 ml dehydrated alcohol was injected as sclerotherapy. The pathological analysis yielded cystic hygroma. The six months post-surgery follow up showed no recurrence, and the patient remained symptoms free. Retropharyngeal lymphangioma should be considered as a differential diagnosis of retropharyngeal abscess. Their treatment in primary and acute conditions is the same, including drainage, intravenous antibiotics therapy, and airway care. However, diagnosis of retropharyngeal lymphangioma is important because it needs farther evaluation and treatment. Few cases of retropharyngeal lymphangioma were reported, and this was a challenging case with a wide area of involvement.

Keywords

Retropharyngeal mass, Head and neck lymphangioma, Cystic hygroma

Introduction

Lymphangioma is a congenital malformation due to a failure of communication and lymph drainage into the venous system [1]. Lymphangiomas are most commonly presented as painless masses with soft consistency in the neck, clavicle and axillary areas [2]. They also can be seen in the limbs, axillary, and mediastinum [3]. The involvement of retropharyngeal space is rare, and only few cases were reported [4-6]. Airway obstruction, dyspnea, and stridor in patients with lymphangioma is caused by external compression or intrinsic involvement of upper airways, which could lead to emergency tracheostomy [7]. Here, we present a case of retropharyngeal lymphangioma and, discussed its manifestations, imaging and treatment.

Case Report

History and examination

An 11-year-old female was referred to our department due to the failure of antibiotic treatment and drainage for suspicion of retropharyngeal abscess.

At the time of admission, the patient had a fever (38.8 °C), dyspnea, stridor, dysphagia, and drooling. On examination, retropharyngeal swelling and hot-potato speech were observed. Antibiotics (Ceftriaxone and Clindamycin adjusted with weight) and steroids were given intravenously. Cold humidification and oxygenation were administered. The patient was admitted into the intensive care unit (ICU) for closed monitoring and better airway care. After 48 hours the general condition of the patient improved. Symptoms of dyspnea and fever disappeared, and the patient tolerated enteral feeding. She transferred to the otolaryngology ward and magnetic resonance imaging (MRI) was requested. According to MRI results, indicating retropharyngeal lymphangioma, the patient was discharged with the continuation of oral antibiotic therapy and scheduled for elective surgery next month.

Imaging

On MRI sequences, a multicystic lesion was seen from the base of the skull to the level of T4 in retropharyngeal space, which extends into the right parapharyngeal space (Figure 1). On T1W1 MRI sections, the lesion was seen hypointense to isointense in comparison to adjacent muscles as shown in figure 2. Likewise, the lesion was presented as hyperintense from the surrounding tissue on T2W1 MRI sections (Figure 1). After injecting the contrast agent (GAD), the internal lesion space remained hypointense and not absorbing, but the walls and septations enhanced completely (Figure 2). On CT scan with IV contrast, the hypodense mass was seen in retropharyngeal space (Figure 3).

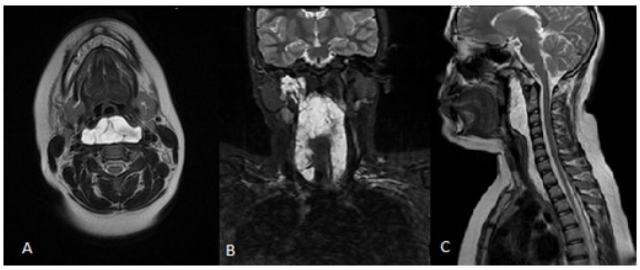

Figure 1: Preoperative T2W1 MRI of retropharyngeal mass; the lesion was presented as hyperintense from the surrounding tissue. A: Axial section, B: Coronal section, C: Sagittal section.

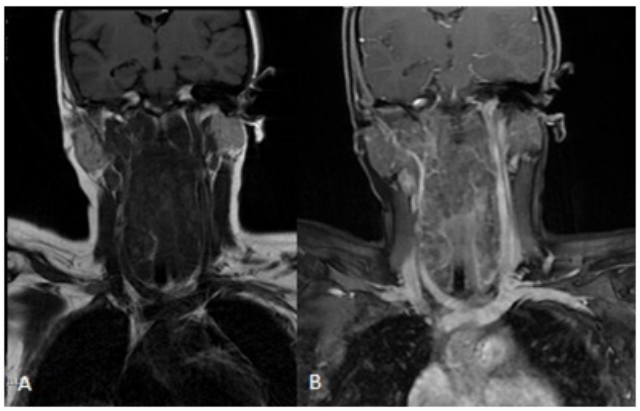

Figure 2: Preoperative T1W1 MRI of retropharyngeal mass. The lesion was seen hypointense to isointense in comparison to adjacent muscles. A: Without contrast, B: With GAD IV contrast.

Operation

During surgery, the vertical incision parallel to the anterior border of sternocleidomastoid (SCM) muscle was made on the right side of the neck skin. SCM muscle was retracted laterally. Jugular vein and other components of carotid sheath were retracted up and medial. Then, alar fascia under the larynx and esophagus were cut. Lymphangioma was removed by blunt dissection (Figure 4). To access the upper mediastina through the neck incision, the patient's neck was extended. To facilitate the complete removal of the lesion, lymphangioma was aspirated before resection. Then, at the site of surgery, 5 ml dehydrated alcohol was injected as sclerotherapy. The pathology examination reported cystic hygroma. In follow up, CT imaging six months post-surgery showed no recurrence, and the patient remains symptoms free without any significant findings in examination.

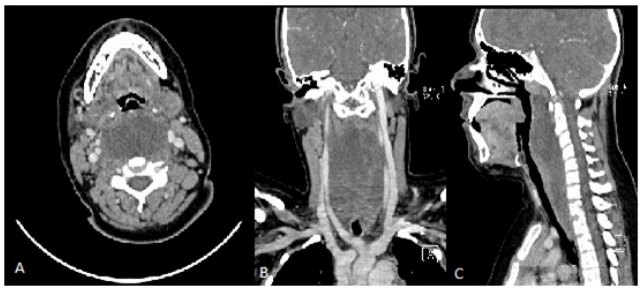

Figure 3: Preoperative CT-scan with IV contrast of retropharyngeal mass; the hypodense mass was seen in retropharyngeal space. A: Axial section, B: Coronal section, C: Sagittal section.

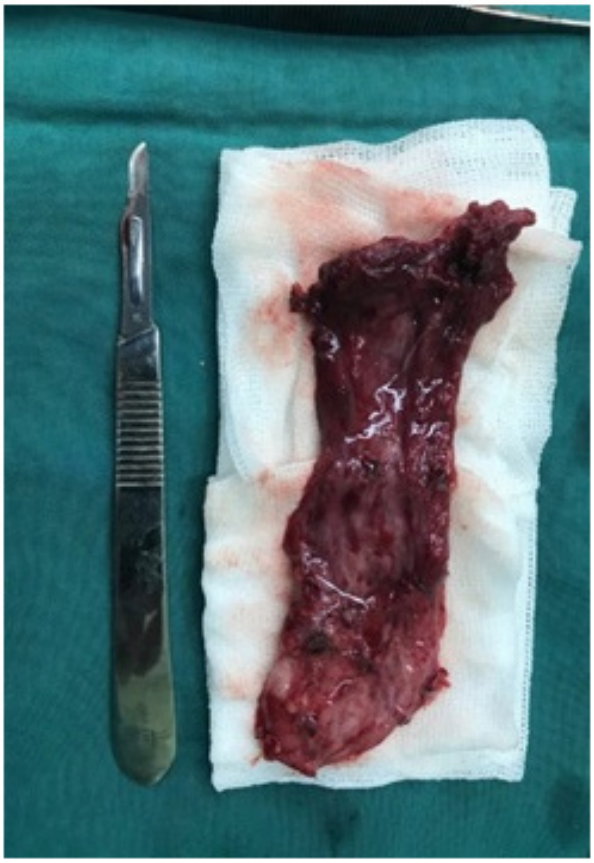

Figure 4: Resected retropharyngeal cystic lesion.

Discussion

Lymphangioma includes about 6% of the benign childhood head and neck mass. It is usually symptomatic at birth and in 90% of cases under the age of 2 years [8]. Some few cases have been diagnosed at older ages and even in adulthood [3]. At older ages, lymphangioma is mostly diagnosed following its complications. One of the complications of lymphangioma is its rapid growth due to infection. Growing mass can cause airway obstruction when intrinsic involvement of upper airways (such as trachea) or external compression (neck spaces involvement) is presented, which can be life threatening. In these cases, drainage, appropriate antibiotics and steroids therapy, and airway care (if needed, tracheostomy) was recommended. Complete surgical resection (choice treatment) can be postponed until the patient becomes stable [7].

In our patient, retropharyngeal lymphangioma got symptomatic following an infection that caused airway obstruction symptoms. By appropriate treatment without the need for tracheostomy, the patient became symptom free and was prepared for surgery.

Choice of treatment in head and neck lymphangioma is a complete surgical resection [3, 9], which sometimes encounters difficulties due to vital structures involvement. If the lesion is not completely resected, the percentage of recurrence is very high (40 to 100%) [10]. Radiotherapy and laser therapy are alternative therapies. As lymphangioma is a non-malignant disease and due to high side effects of radiotherapy, it is not used widely. Another therapeutic method that is most commonly used is sclerotherapy, which is less effective than surgery and sometimes associated with systemic side effects of sclerosing substance [9]. It seems that intralesional dehydrated alcohol injection as a sclerosing fluid after surgery is associated with a better outcome and reducing the prevalence of recurrence. Dehydrated alcohol is known as the strongest sclerosing substance without systemic side effects that induces fibrosis without promoting an inflammatory response. It's mechanism remain unknown [1, 11].

In this patient, after surgery and complete removal of the lesion, a dehydrated alcohol is used at the side of surgery. It should be noted that alcohol injection is very painful and injection under anesthesia could be much more tolerable.

Conclusion

If lymphangioma involves uncommon spaces such as a retropharyngeal space, it is mostly diagnosed following the complications. One of the complications of lymphangioma is a rapid growth due to infection. Therefore, it should be considered as a differential diagnosis of retropharyngeal abscess. Their treatment in primary and acute conditions is the same, including drainage, intravenous antibiotics therapy, and airway care. Diagnosis is usually made with imaging (often MRI). The choice of treatment in retropharyngeal lymphangioma is complete surgical resection, and it's better to be done with proper preparation in tertiary centers.

Disclosures

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None.

Conflict of Interest

None declared.

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