

Giant cystic lymphangioma of the neck with retropharyngeal extension: A case report

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ABSTRACT

Introduction: Cystic lymphangiomas are rare benign congenital malformations of the lymphatic system.

Observation: We report the case of a 17 months-old male child who presented with a mass in the right side of the neck since birth. Imaging showed a cystic lymphangioma with retropharyngeal extension. Complete surgical excision was done. At follow up, no recurrence was observed.

Conclusion: Cystic lymphangiomas of the head and neck are benign lesions which are easy to diagnose. The best treatment is surgical removal. However others treatments especially sclerotherapy had shown good results.

Keywords: Cystic lymphangioma, cystic hygroma, neck mass, retropharyngeal

INTRODUCTION

Cystic lymphangiomas are congenital malformations of the lymphatic system that mostly affect the head and neck and usually diagnosed in infancy. Diagnosis is mainly based on clinical examination and imaging. The best treatment is surgical removal.

CASE REPORT

We report the case of a 17 months-old male child with no significant past medical history, full term and normally delivered.

He was admitted for a swelling on the right side of the neck, which was progressively increasing since birth.

Physical examination revealed a painless, elastic and mobile neck mass (figure 1).

No other congenital anomalies were detected.

Ultrasonography of the neck showed a cystic lesion 86.2 x 26.1 mm with internal septa, suggesting a cyst lymphangioma. MRI showed a cystic lesion, posterior to the sternocleidomastoid muscle and the carotid sheath and anterior to the prevertebral muscles, extending to retropharyngeal space hypointense on T1 sequences and homogeneously hyperintense on T2. There was minimal peripheral contrast enhancement (figure 2,3). The patient underwent extirpation of the mass; surgery was radical macroscopically (figure 4).

The postoperative course was uneventful and the patient was discharged on the third postoperative day.

The histopathological examination confirmed the diagnosis of cystic lymphangioma. At follow up, no recurrence was observed.



Figure 1 : Pre-operative image showing the lateral neck mass

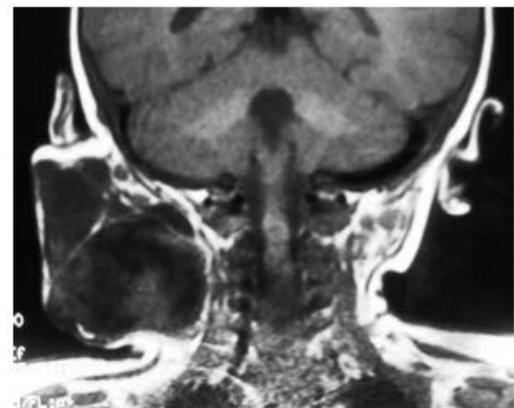


Figure 2 : Coronal view of T1-weighted MRI showing a large hypointense mass in the right side of the neck.



Figure 3 : Axial view of T2-weighted MRI showing a large hyperintense mass in the right side of the neck with retropharyngeal extension.



Figure 4 : Per-operative image showing the cystic lymphangioma the jugular vein, the carotid artery and the pneumogastric nerve with retropharyngeal extension.

DISCUSSION

Cystic lymphangiomas or cystic hygromas are rare benign congenital malformations of the lymphatic system. The pathophysiology of cystic lymphangioma remains unclear. Embryologically, these malformations are believed to arise from abnormal sequestration of proliferating lymphatic mesenchyme during the development of lymphatico-venous sacs. These sequestered tissues then failed to communicate with the rest of the lymphatic or venous system [1].

Lymphangioma can be seen anywhere in the body but are more commonly seen in lymphatic-rich areas, such as the head and neck, axilla, mediastinum, groin and below tongue, and are rarely seen in digestive system and limbs[2].

In 80%, lymphangiomas are located in the cervico-facial region (generally in the posterior triangle) [2]. These malformations are present in 65% to 75% at birth and 80% to 90% by the second to the third year of life [3], but it can appear in all age group [4]. There is no predilection for either sex.

Prenatal diagnosis is possible by ultrasound examination and can be isolated or associated with other anomalies such as Turner's syndrome, Down's syndrome, trisomy 18, trisomy 13, Noonan syndrome, etc. [5].

After birth, cystic lymphangioma appears as a painless mass that gradually enlarges, then remains static over a long period and can even shrink spontaneously [6].

Although these lesions are benign, other modes of presentations are possible due the complications and effects of cystic lymphangiomas such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion [2].

On ultrasound, a cystic lymphangioma appears as a multilocular cystic mass with septa of variable thickness [8], and on Doppler imaging, no or only a few signals are displayed [9]. The differential diagnosis of a cystic extrathyroidal neck mass included especially branchial cyst, thyroglossal duct cyst, abscess, resolving hematoma, lymph node.

MRI can be employed to describe the lesion, in a better way by showing extension and relation with other anatomical structures as our case were MRI revealed the retropharyngeal extension and to plan the surgical procedure. It appears that most lymphangiomas are hypointense in T1 weighted image and hyperintense reflecting the preponderance of fluid-filled cystic spaces [10].

Effective treatment for cystic lymphangioma remains complete surgical excision, but other therapeutic options are still possible such as sclerotherapy (injection of sclerosing agents: OK-432, doxycycline, ethanol, bleomycin, Ethibloc, etc) [11], laser therapy [12], systemic drug therapies, simple drainage, aspirations, radiation, radio-frequency ablation [13] and cauterization.

The indications for the treatment are recurrent bouts of infection, respiratory distress, dysphagia, hemorrhage, pain, sudden increase in the size of lesion and disfigurement [14, 15]. Sometimes a tracheotomy is necessary in case of severe respiratory distress [2, 15].

When surgical removal is performed, the vital structures should be conserved as lymphangiomas are benign lesions.



Main complications of surgical attitude are the damage to nerves and vessels, the scar, wound infections and the frequency of recurrences.

CONCLUSION

Cystic lymphangiomas of the head and neck are benign lesions which are easy to diagnose. The best treatment is surgical removal. However other treatments especially sclerotherapy had shown good results.

Conflicts of interest: Authors declared no conflicts of interest.

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