Case Report

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A difficult case of cystic hygroma

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ABSTRACT

Cystic hygromas are uncommon congenital lymphatic malformations that commonly involve the lateral aspect of the neck. The treatment of choice of these swellings is surgical excision but at times, it becomes quite difficult to perform a complete removal because of the tendency of the tumor to infiltrate the surrounding structures. We encountered a similar case of cystic hygroma that was infiltrating the carotid sheath. We were able to achieve complete surgical excision without damaging the surrounding structures.

Keywords: Cystic hygroma, Lymphatic malformations, Lymphangioma

INTRODUCTION

Hygroma in Greek means water-containing tumor. Lymphatic malformations (lymphangioma and cystic hygroma) are uncommon congenital anomalies, and the precise embryonic origin of lymphatic malformations remains unknown. ¹⁻⁵ They are generally considered to be vascular malformations that result from sequestration of lymphatic tissue that fail to communicate normally with the lymphatic system. Notably, 80-90% of lymphatic malformation cases are diagnosed by the age of 2 years. ^{6,7} Lymphatic malformations occur throughout the body but are most common in the head and neck. The symptoms are related to the anatomical location of the lesions as well as the extent of involvement of the local anatomical structure.

Lymphangiomas are usually classified as capillary, cavernous or cystic lymphangiomas. They may also be classified more conveniently, on the basis of size of the cysts contained, as microcystic, macrocystic and mixed lymphangiomas. Microcystic lymphangioma consists of cysts measuring <2 cm in size, whereas the size of cysts in case of macrocystic lymphangioma is more than 2 cm. The mixed lymphangioma is characterized by cysts of variable sizes.^{6,8}

CASE REPORT

A 4-year-old girl presented with a complaint of swelling in the left side of the neck since last 2 years. The swelling had been aspirated twice at some private hospital, but both times it recurred.

On examination, there was a well-defined cystic swelling in the left side of the neck involving inferior aspect of the lower jaw and extending up to the supraclavicular region. On transverse axis the swelling was extending from the midline to the lateral border of the sternocleidomastoid muscle (SCM) (Figure 1).

Hemogram and other blood investigations were within normal limits. Magnetic resonance imaging (MRI) revealed a hyperintense T2 image showing the lobulated cystic lesion in the left side of the neck and abutting the carotid sheath and components.

Patient was taken up for surgery after arrangement of blood for any possible intraoperative blood loss. A transverse skin incision overlying the most prominent part of swelling, extending from the lateral border of SCM to midline along the skin crease was given, and subplatysmal flaps were raised. Intraoperatively a large cystic mass abutting the submandibular gland and encasing the carotid sheath was seen (Figure 2). With the help of sharp and blunt dissection the swelling was excised after removing all adhesions and preserving the vital structures (Figures 3 and 4). Adequate hemostasis was achieved. A suction drain was placed beneath the flaps and closure was done using monofilament nylon 4-0.

Post-operative period was uneventful; drain was removed on 3rd post-operative day and sutures were removed on 7th post-operative day. Histopathology report confirmed the diagnosis of cystic hygroma. Patient had no evidence of recurrence during the 3-month follow-up.

DISCUSSION

Lymphangioma is a benign congenital malformation of the lymphatic system. There are three histological subtypes: capillary lymphangioma (composed of small lymphatics), cavernous lymphangioma (composed of larger lymphatics) and cystic lymphangioma (cystic hygroma composed of large macroscopic lymphatic spaces with



Figure 1: Pre-operative image showing left lateral neck swelling.

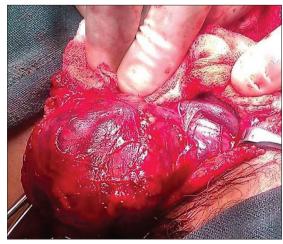


Figure 2: Intra-operative image showing the tumor with internal jugular vein hooked apart.

collagen and smooth muscle). Cavernous lymphangioma is the most common subtype. Cystic lymphangioma occurs approximately 1 in 12,000 births and 95% present by the 2nd year of life.^{6,7} Although the lesion can occur anywhere, the most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis.⁹

Lymphangiomas are best visualized by MRI; the high water content allows lymphangiomas to appear hyperintense on T2-weighted images. ¹⁰ The other imaging methods are Doppler ultrasonography and computed tomography. This disease may be associated with Turner syndrome, Noonan syndrome, cardiac anomalies, trisomy syndromes and fetal hydrops. ¹¹⁻¹³ Differential diagnosis includes hemangioma and meningomyelocele. ⁷ Infection within the cysts (usually caused by streptococcus or staphylococcus species) may occur. This complication can cause rapid enlargement that may result in airway obstruction. Bleeding into the cyst is another complication of the lymphangioma. These tumors do not resolve spontaneously. Percutaneous aspiration is not preferred because of the risk of bleeding, infection and recurrence.



Figure 3: Post resection image showing carotid sheath structures.



Figure 4: Resected specimen.

In our case, fortunately, infection was not there despite previous attempts of aspiration.

Injection of sclerosing agents like alcohol, bleomycin and OK-432 (a lyophilized mixture of streptococcus pyogenes and penicillin G potassium), with favorable results have been reported. Complete surgical excision is the preferred treatment. It can be performed under general or local anesthesia. Sometimes, this may be impossible due to the infiltrating nature of the hygroma within and around neurovascular structures, muscles, blood vessels. In our case also, the tumor was encasing the carotid sheath but with patience and careful, meticulous dissection, we were able to achieve complete excision. When complete excision is impossible, unroofing partial cystectomy and drainage of the cystic content can be performed. In this kind of treatment, the recurrence rate of 10-15% was reported.

CONCLUSION

Cystic hygroma is predominantly a disease of the prenatal period, infancy and early childhood (though cases in adults have also been reported). This is a benign situation which can be treated with different methods. The early recognition of the lymphatic malformation with wide en block excision while preserving vital structures and cosmetic function is the best approach to these lesions. Staged excision may be necessary to avoid mutilating surgical procedure in some selected cases.

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