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# Giant Cystic Hygroma in a Child: A Rare Case Report and Literature Review

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**Abstract:** Cystic hygroma is a benign congenital malformation that results in a cystic lesion, predominantly in the head and neck. Although benign, they can be a cause for concern as these cysts can cause compression and invasion of surrounding structures resulting in respiratory difficulty, dysphagia etc. Clinically these lesions are soft, non-tender, transilluminant unless complicated by infection or hemorrhage. Definite diagnosis and treatment planning is done with the help of an ultrasound, computed tomography and magnetic resonance imaging. Here we present a case of a 2 year 10-month old boy with large swelling on the right side of neck. Complete surgical excision was achieved without any damage to vital surrounding structures. Follow up showed good healing of wound and no recurrence.

**Keywords:** Cystic, Hygroma, Giant, Child

## INTRODUCTION

Cystic hygroma is a congenital benign malformation that results from abnormal development of lymphatic system. Cystic hygroma is more accurately termed as lymphangioma as it denotes the tissue of origin (1). Abnormal communication between developing lymphatics and draining veins have been proposed as the reason for its development (2,3). It is more common in children, although rare incidence in adults has been reported (4). They are more common in neck and cervical region (75%) and can be unilateral or bilateral (4,5). Majority of cystic hygromas are asymptomatic swellings but can be complicated by secondary infection, rupture, hemorrhage in the cyst and invasion of surrounding structures (3,6). Diagnosis with an ultrasound in prenatal period has been well documented (3). Apart from ultrasound, computed tomography and magnetic resonance imaing can play an important role in their diagnosis, differentiation from other lesions, establishing relation to surrounding structures and effective planning of further treatment (7,8). Although other treatment modalities have been used, complete surgical excision has remained the mainstay of treatment for management of giant cystic hygroma (3,6,9).

## CASE REPORT

Here we present a case report of a 2 year 10-month-old boy who presented to us with right lower neck (cervical) swelling since six months. Swelling was progressively increasing in size. He had no respiratory distress or dysphagia at the time. Clinical examination revealed a

10*8*6 cm lobulated, soft, cystic swelling in the posterior triangle of the neck. The swelling was non tender and transilluminant. His other systemic examination and blood tests were within normal limit.

Ultrasonography showed a multilocular cystic lesion. Magnetic resonance imaging (MRI) showed Multiloculated cystic lesion in right lower neck and supraclavicular region, with features suggestive of a macrocystic lymphatic malformation likely cervical cystic lymphangioma (Figure 1). The mass was extending to medial supraclavicular region posterolateral to right carotid sheath. There was no extension in to the thoracic cage (Figure 2).

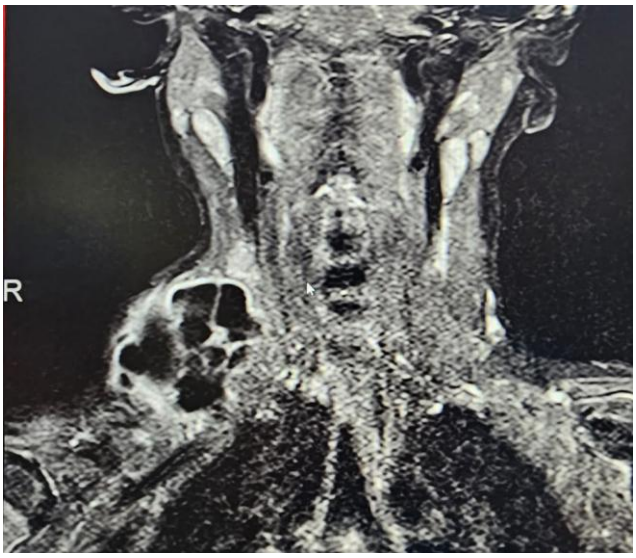


**Figure 1: MRI showing giant cystic lymphangioma in right lower neck**

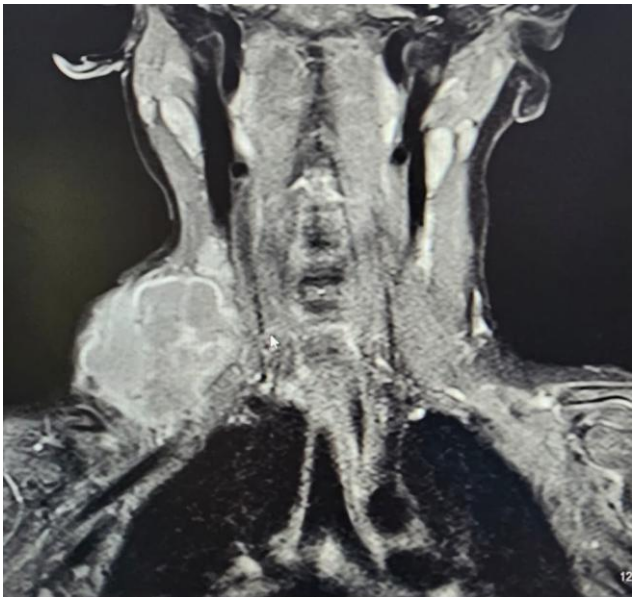

**Figure 2: MRI with contrast showing giant cystic lymphangioma of right lower neck with early compression of surrounding structures**

The child was operated through a right lower transverse cervical incision. The cystic swelling was removed completely through careful dissection ensuring safety of surrounding nerves and blood vessels (Figure 3). Patient had a good postoperative recovery and was discharge in good condition.

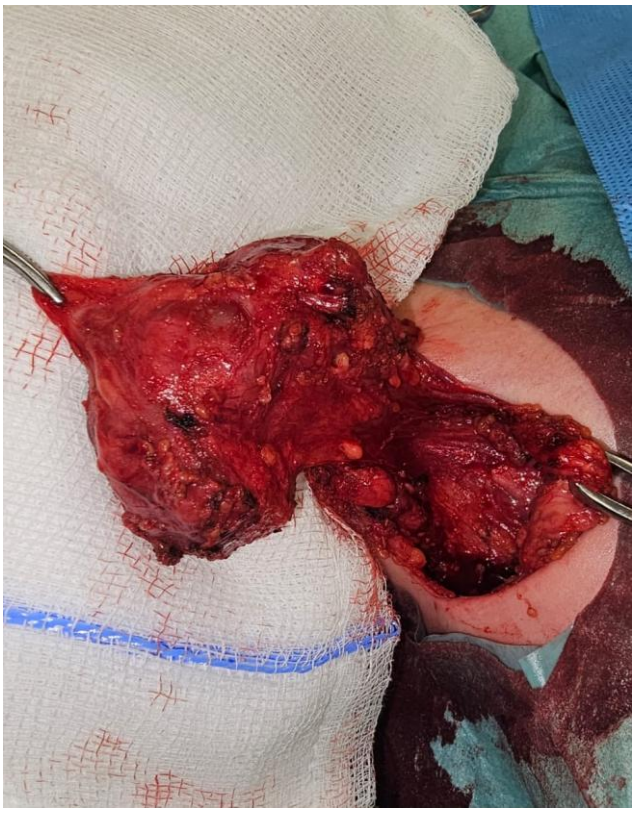

**Figure 3: intraoperative picture showing surgical excision in-toto preserving vital structures**

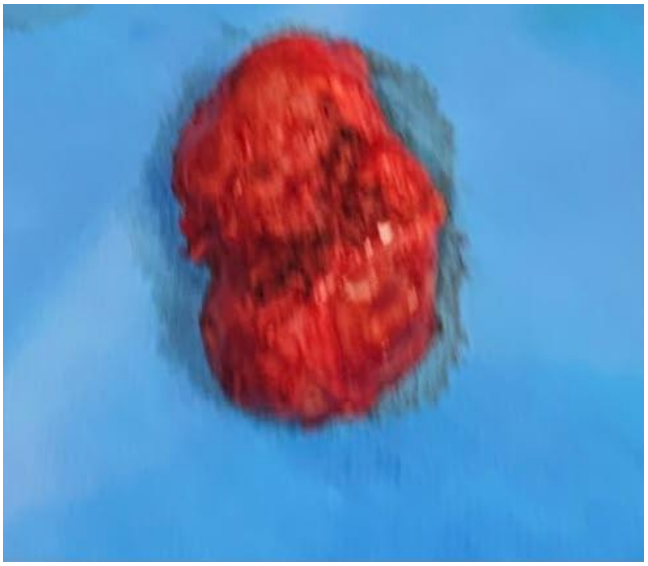

**Figure 4: Excised specimen**

The histological findings grossly revealed a single reddish brown mass that showed multiple cysts with haemorrhage to myxoid in appearance (Figure 4). Microscopy revealed thin walled, dilated lymphatic vessels lined by flattened epithelium. Lumina containing eosinophils, proteinaceous fluid surrounded by lymphoid aggregates diagnostic of Cystic Lymphangioma.

He was followed up after an interval of two weeks, one month and six months with satisfactory healing of wound, acceptable scar and no recurrence.

## DISCUSSION AND LITERATURE REVIEW

The word hygroma originates from Greek, meaning a water containing tumor (6). Cystic hygroma is a benign congenital malformation that occurs at sites of lymphatic venous connection and can be single or multiple fluid filled cysts (2).

During eighth week of gestation six lymphatic sacs can be identified in the developing embryo, two jugular sacs, two iliac, one at the base of root of mesentery and one posterior to abdominal aorta. These sacs establish communication through a network of lymphatics (3,6). There are a number of mechanisms that have been proposed to explain their development. One of them is embryological sequestration of lymphatic tissue from lymphatic sacs during development. These tissues fail to establish communication with lymphatic or venous system. Dilatation of these lymphatic tissues results in the development of cystic hygromas (3,6). The cyst walls are lined by a single layer of flattened endothelium supported by connective tissue stroma. The fluid within the cysts is serous, being clear or straw colored and occasionally blood stained. (11).

Lymphangiomas can be classified into three categories based on type. (1) Lymphangioma simplex composed of capillary sized thin walled lymphatic channels (2) cavernous lymphangioma composed of dilated lymphatic channels (3) cystic lymphangioma, being commonly referred as cystic hygroma (3,12). They can also be classified based on size of cysts as microcystic, macrocystic and mixed. Microcystic lymphangiomas are less than 2 cm in size, macrocystic are more than 2 cm in size while mixed lymphangiomas have cysts of variable sizes (3,13).

The most common location of cystic hygroma is cervicofacial region (especially in the posterior triangle of neck), axilla, mediastinum, groin and oral cavity. More than 95% of cystic hygromas occur in head and neck with the remaining 5%, located in various sites including the abdomen (3,6,14).

The most common presentation of cystic hygroma is an asymptomatic swelling in the posterior triangle of the neck. Other modes of presentations are related to effect or complication of the cyst such as respiratory difficulty, feeding difficultes, fever, increase in the size and infection of the cyst (3,6,11,13). On clinical examination, they are soft, cystic compressible and non-tender masses. They are transilluminant unless complicated by infection or hemorrhage (3,6,9).

The role of ultrasonography as a diagnostic tool has been well established in the prenatal period. Postnatally clinical examination combined with ultrasound can be used for diagnosis. For more complex lesions computed tomography or magnetic resonance imaging maybe required to accurately assess the extent of these lesions. Magnetic resonance imaging

has been reported to be particularly useful in evaluating extension of these lesions in the neck and mediastinum as well as establishing their relationship with the surrounding neurovascular bundles and soft tissue structures. These modalities are particularly useful in planning surgical management of these lesions (3,6,7,8,15).

The treatment indications for cystic hygromas are enlarging size, respiratory distress, swallowing difficulties, infection, spontaneous rupture and hemorrhage in the cyst (3,6,10).

The most preferred treatment advocated for cystic hygroma is complete surgical excision (3,6,9,10,16) . Other treatment modalities which have been described for treating lymphangiomas are; simple drainage, aspiration, radiotherapy, laser excision, steroids and sclerotherapy (Bleomycin, OK-432) (2,3,6). Management of large cystic hygroma which involves head and neck, oral cavity and deeper structures is always with surgical excision. Surgical excision is advised sooner rather than later as excision is technically easier before the lesion has invaded the surrounding structures and before infection has resulted in fibrosis and scarring. Care has to be taken during surgical excision to avoid damage to surrounding neurovascular bundles and vitals structures. Cystic hygroma is a benign pathology and damage to vital structures should be avoided. It is good practice to follow certain principles during surgical excision to avoid injury to vitals structures such as adequate exposure of the lesion, minimal bleeding, avoiding rupturing the cyst to preserve delineation of cyst anatomy, and an attempt to ligate draining lymphatics which might not always be feasible (2,6,9,10,16). In giant cystic hygromas complete excision might not be possible at times and it is sensible to plan a staged procedure. Postoperative complications include recurrence, seromas, infection, scar hypertrophy and nerve damage. Recurrence rates vary depending on size, complexity and completeness of excision. In completely excised complex lesions recurrence has been observed in 10% to 27% of patients, while in partial resections, recurrence has been observed in 50% to 100 % of patients (12,17). In our case complete surgical excision was done with good postoperative recovery. Follow up at one month and six months' interval showed satisfactory scar healing and no recurrence.

Among other non-surgical treatment options sclerotherapy with OK-432 and bleomycin has shown good results in modern times. A few side effects such as fever, surrounding erythema and swelling have been noted after this therapy (2,18). Surgical excision is still the preferred treatment modality for giant cystic hygromas with satisfactory results especially lesions with propensity to cause respiratory compromise, dysphagia and other associated complications (3,9,16)

## CONCLUSION

Giant cystic hygromas can be managed successfully with complete surgical excision. Complete surgical excision results in minimal chances of recurrence. Careful preoperative planning can improve chances of complete surgical excision and avoid damage to vital structures. Our aim is to enhance the existing scanty literature regarding this condition and emphasize on the effective surgical management of the same.

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