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Editorial

Large Cervical Cystic Hygroma in Infants: Report of Two Cases

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Abstract

Cystic lymphangiomas are also called as cystic hygromas and are benign, congenital malformation of the lymphatic system, and reported to occur in the head and neck regions in 90% of the cases. This is a report of two cases of large cystic hygromas involved at the neck in both the cases, while in the second case also involved the face along with the neck. Case one was 45-days male and he was admitted with a large swelling at the right side of the neck, while the second case was 3-months-old female and she was admitted with a large swelling involved the right side of the neck and face as well, and she also had mild respiratory distress. Both were investigated with Ultrasonography (USG) and Computed Tomography (CT) scans of the swellings, which showed multiloculated, cystic hygromas. In both the cases complete surgical excision of the cystic hygromas was successfully accomplished. Immediate post-operative period in both the cases were uneventful. In first case, follow-up done 3 months after surgical excision of the cystic hygroma was good and he was doing well. In second case, follow - up was done after a month of surgical excision of cystic hygroma and she had features of neuropraxia of mandibular branch of right facial nerve, and was expected to correct with time.

Introduction

Lymphangiomas are benign, congenital malformation of the lymphatic system with marked predilection to head and neck regions and reported to occur in 90% of the cases at head and neck area [1,2]. In some of the cases a large cystic lymphangiomas involving the neck area may obstruct pharynx / esophagus and may present with dysphasia, or it may obstruct and compresses the air way and leading to the respiratory distress and that necessitates an immediate / early surgical intervention [3,4]. Post-natal investigation for lymphangiomas includes; USG, CT scan, and magnetic resonance imaging (MRI) of the swelling [1-6]. Surgical excision remains a standard treatment for the cystic lymphangiomas, although other modalities like sclerotherapy and others are also used with their own merits and demerits [1- 8].

Case Reports

Case One

A 45-days-old boy was admitted with a history of a large size swelling involving his right side of the neck since birth. He was full-term normally delivered. Antenatal period was irregular and USG was not done. Clinical examination of the swelling revealed that it was single swelling of size 10 x 8 cm, occupied anterior, lateral and posterior aspect of the right side of the neck (Figure 1a). It was soft to firm, non-tender, and cystic. Other systemic examination was within normal limits and there were no any associate other anomalies detected during clinical examination.

USG of the swelling revealed that is was multicystic in nature and reported as cystic hygroma of the neck. His CT scan of neck area revealed that there was a large multiloculated, cystic lesion measuring 7.8 x 3.8 x 7 cm, involving the right posterior cervical space, anterior and lateral aspects of neck, and also showing thick septations (Figure 1b). It was also displacing the right sternomastoid muscle towards laterally and displacing the trachea, oropharynx, and other right great vessels towards medially. The CT findings were consistent with large cervical cystic lymphangioma.

Complete surgical excision of the lesion was achieved in single sitting without any difficulties (Figure 1c and Figure 1d). His post–operative period was uneventful. Excised specimen was submitted for histological examination and was reported as that the cysts size varying from 0.5 to 5 cm in maximum diameter. Cut section of cysts was filled with clear serous fluid and wall thickness varying from 1 mm to 3 mm. The histological picture was suggestive of lymphangioma. Follow-up done 3-months after surgical excision of the cystic hygroma, he was doing well.

Case Two

Three-months-old girl was admitted with a large swelling involving her right side of neck and face since birth. She was full term normal delivered girl. Her antenatal period was normal, although USG was not done during antenatal period. Clinically she also had mild respiratory distress. Clinical

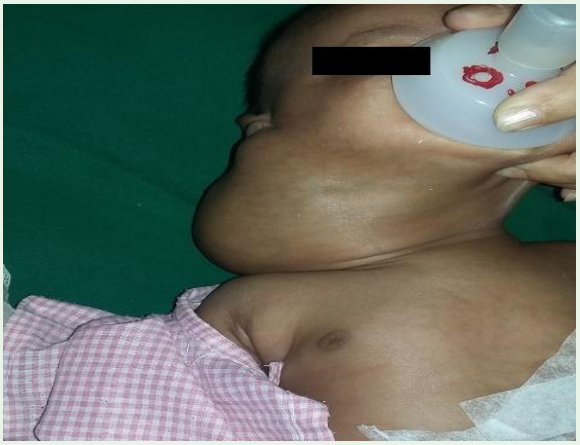

Figure 1a: Pre-operative clinical photograph showing swelling at right side of the neck.

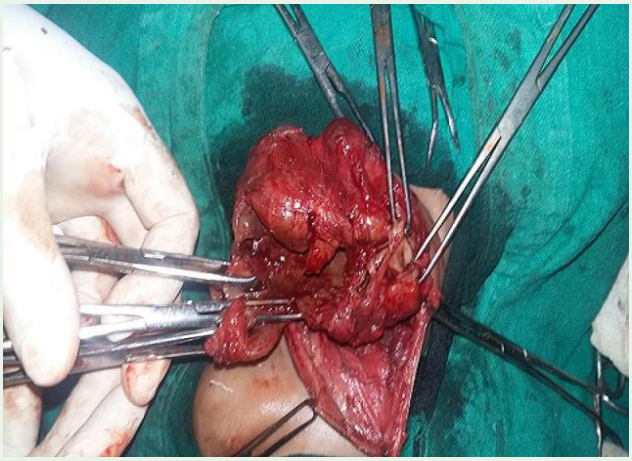

Figure 1c: Operative photograph showing partially excised cystic hygroma.

examination of the swelling revealed that the swelling occupied right side of the neck and face (Figure 2a and Figure 1b). It was soft, cystic, non-tender, and trans-illumination test was also positive (Figure 2c).

Her USG of neck area showed multicystic cystic hygroma. CT scan of the neck revealed that there was a large multiloculated, cystic lesion measuring 9.1 x 9.8 x 8 cm, occupying the right posterior cervical space, sub mandibular space, pre-vertebral space, and retro-pharyngeal space, and also had thin septations (Figure 2d). It was also displacing the right sternomastoid muscle postero lateral, and displacing the trachea, oropharynx, tongue, and right great vessels towards medially.

She was posted for surgical excision of the lesion after complete work up, but postponed twice due to failure to intubate. She also had mild respiratory distress and decided to partially aspirate the cystic fluid. Under sedation 250 ml of yellowish, clear fluid was aspirated (Figure 2e), which was reported as lymphatic fluid showing lymphocytes. This aspiration of cystic fluid helped her in relieving the respiratory distress.

After a week she was again posted for surgery and at this time it was possible to intubate easily. Skin incision was given parallel to the right mandible and at the most prominent part of the lesion. During surgical excision of the lesion, the cystic fluid was partially aspirated out. Complete surgical excision of the cystic lesion was achieved without any difficulties. Named major blood vessels or nerve fibers were not sacrificed during surgery (Figure 2f, Figure 2g and Figure 2h). Her immediate post-operative recovery was excellent. Excised specimen was submitted for histology examination which was reported as single grayish tissue already cut open, measured 7 x 5.5 x 2 cm. The cysts varying in size from 0.5 to 2.5 cm in maximum diameter with cysts wall thickness from 1 mm to 3 mm. The histological findings reported as fibro collagenous stroma showing multiple medium to large size thin walled vessels lined by flat endothelium, stroma showed scattered lymphocytes and lymphoid aggregations. The histological picture was suggestive of lymphangioma / cystic hygroma (Figure 2i). Follow - up was done after a month of surgical excision of the cystic hygroma. She had features of neuropraxia / paresis involving the mandibular branch of the right facial nerve, and the same was expected to resolve with time as there was no obvious nerve injury observed during the surgical excision.

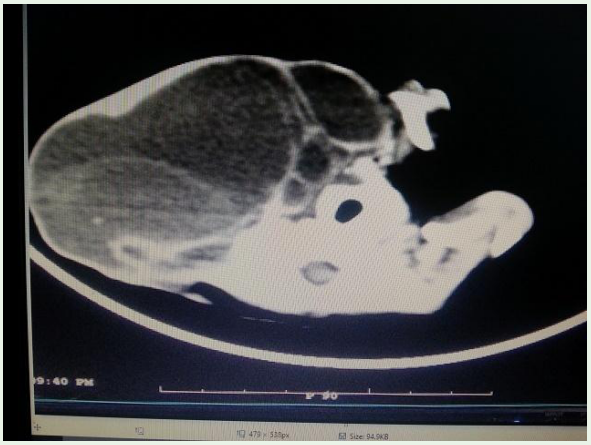

Figure 1b: CT scan of neck showing multiloculated, cystic lesion with septations at right side of the neck.

Figure 1d: Operative photograph showing total excision of lesion.

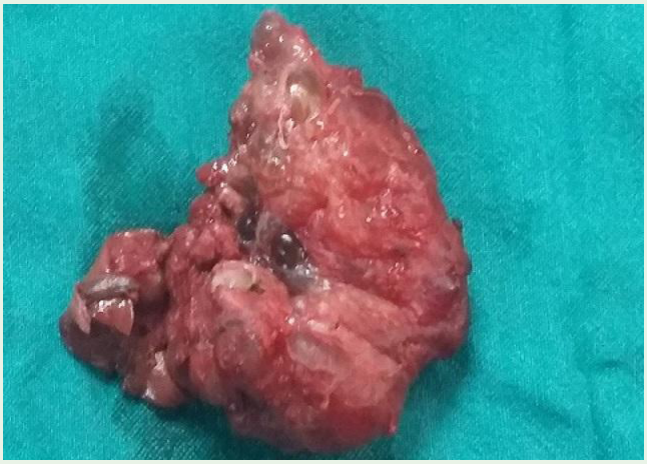

Figure 1e: Photograph showing excised specimen.

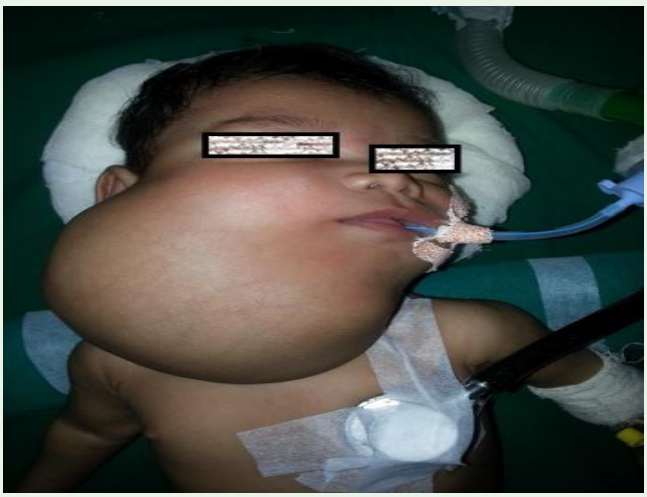

Figure 2a: Pre-operative clinical photograph (front view) showing large swelling involving right side neck, and face.

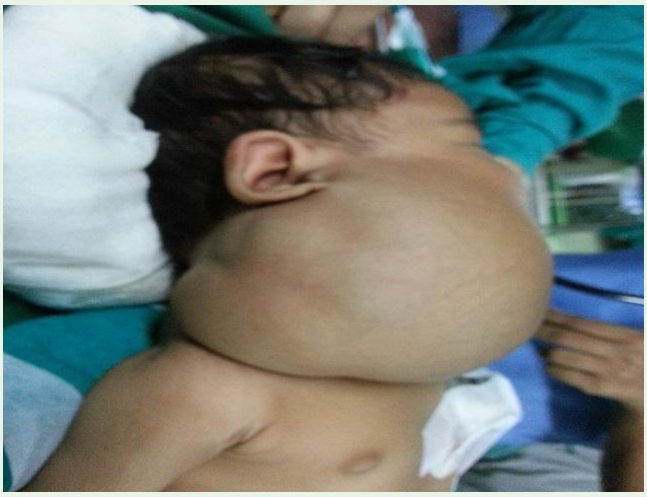

Figure 2b: Pre-operative clinical photograph (lateral view) showing large swelling involving right side neck and face.

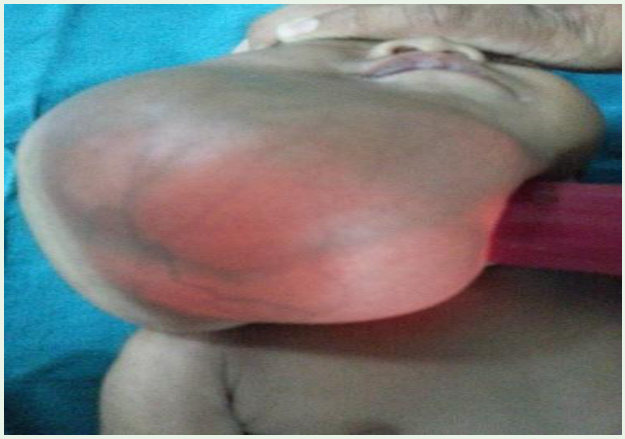

Figure 2c: Pre-operative clinical photograph showing positive transillumination test.

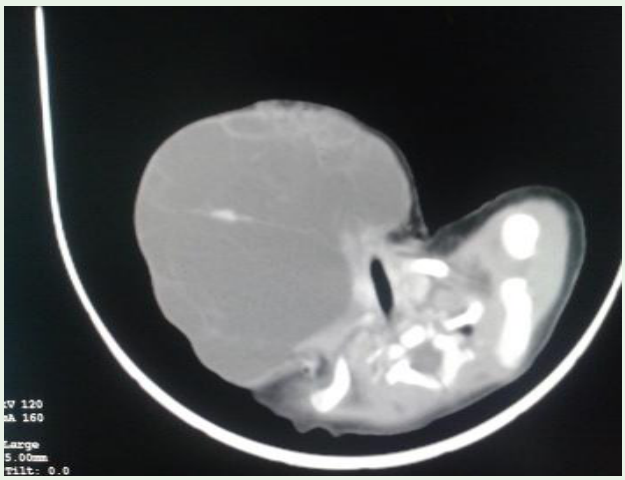

Figure 2d: CT scan of swelling (right side of neck) showing multicystic lesion with thin septations.

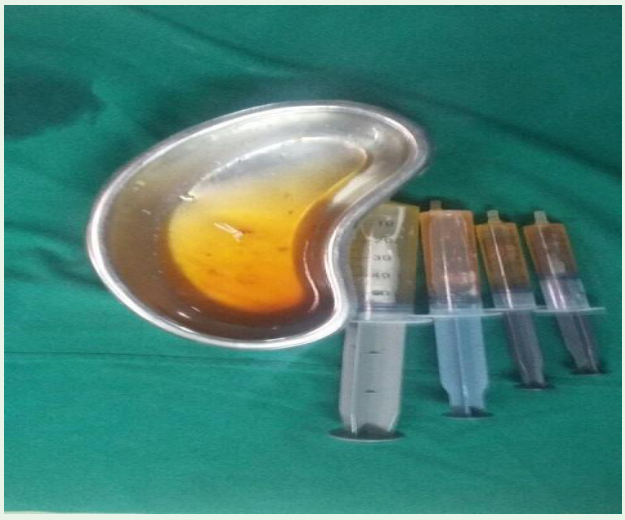

Figure 2e: Clear yellowish fluid aspirated from the cystic hygroma.

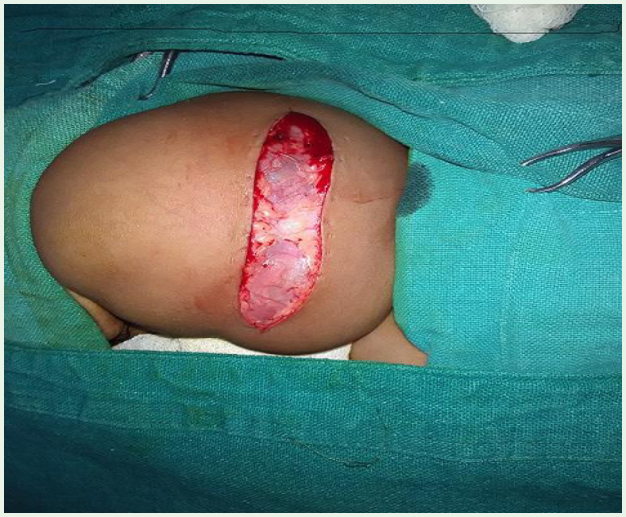

Figure 2f: Operative photograph showing skin incision.

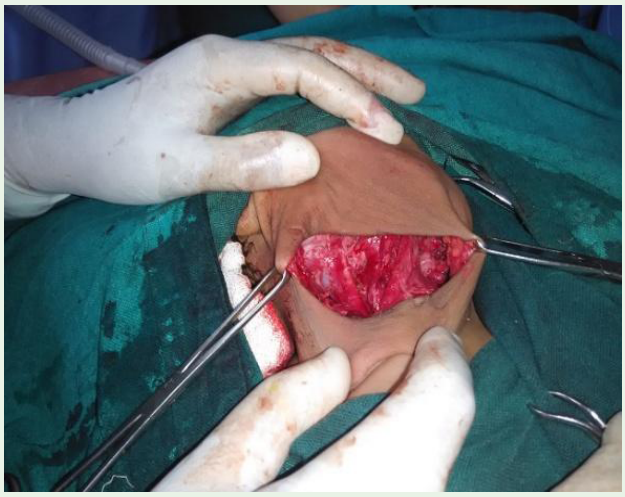

Figure 2h: Operative photograph showing complete excision of the cystic hygroma.

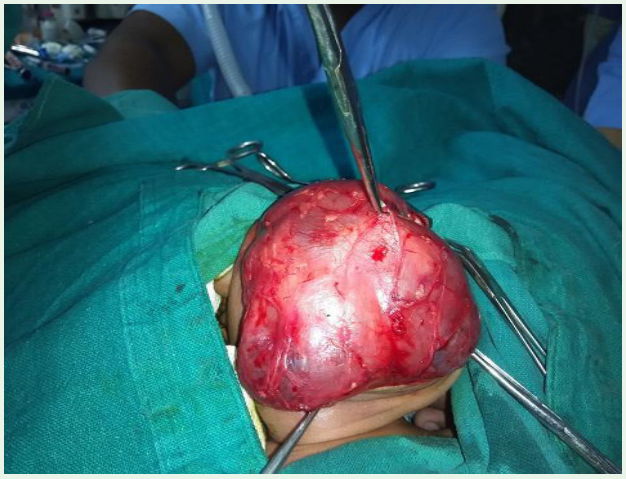

Figure 2g: Operative photograph showing large lesion.

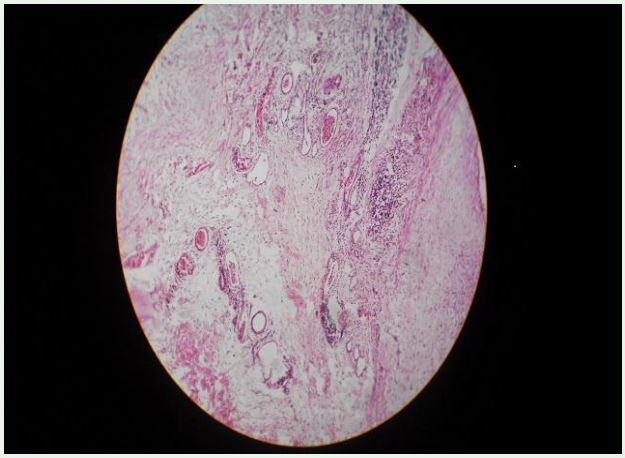

Figure 2i: Histology of excised specimen consistent with cystic lymphangioma.

Discussion

Lymphangiomas are benign, hemartomatous congenital malformation of the lymphatic system. Hygroma is a Greek word meaning "water containing tumor". Lymphangiomas has marked predilection for head and neck regions, especially towards posterior triangle of the neck. Other sites for cystic hygromas are axilla, mediastinum, groin, chest, and oral cavity. Ninety percentages of the cystic lymphangiomas involved the head and neck area [1,2,7]. The most acceptable theory to explain the occurrence of lymphangioma is that it originates from sequestrations of lymphatic tissues / vessels that fail to establish connections to the normal draining vessels during intra uterine life [1,4,5,7].

Lymphangiomas are best classified as lymphangioma simplex (capillary), cavernous lymphangioma, and cystic lymphangiomas. Cystic lymphangiomas are also referred as cystic hygromas. They are also classified based on the size of the lymphatic cysts incorporated within it as micro-cystic (capillary), macro-cystic and mixed lymphangiomas. In the cases of micro-cystic lesions the lymphatic cysts are less than 2 cm, in the case of macro-cystic the cysts are more than 2 cm and in the case of mixed lymphangiomas the sizes vary [1,2,7,8].

Clinically cystic hygroma involving neck area may present as swelling, disfigurement or with its complication and are bleeding, rupture, infection, etc, and above all are the indication of the therapy as well. They are mostly large, cystic, and non-tender and are also having positive trans-illumination test [1-8]. Some of the large hygromas involving the neck area are large enough to compress the airways and may present with respiratory distress, and also demand an urgent surgical intervention [1,4,5]. It is possible to detect, document and diagnose the cervical lymphangiomas on antenatal USG as early as in the last phase of the first trimester of the pregnancy [5]. Postnatal investigations of the cervical lymphangiomas include USG, CT scan and MRI. MRI not only delineates the details of the swelling, but also its relationship with great vessels, and other important structures in more details [1-7].

Surgical excision remains a best treatment option for large cystic

hygromas involving the neck. It is possible to completely excise the cystic hygromas in majority of the cases. As it is a benign anomaly so that destructive surgical resection must be avoided, and resection of cystic hygromas in stages is a better option, if required [1,3,4,6-8]. Surgical excision is not free from the complication and may lead to facial nerve palsy, Horner's syndrome, etc [4,6]. Other modalities for treating lymphangiomas are steroids, sclerotherapy, lasar, bleomycin, radiation, OK 432, with their own merits and demerits [1,2,5,8]. Recurrences are also reported in 5-15% of the cases, and may require another surgical excision or other modalities for treatment.

Present report, both the infants had large cystic hygromas involved neck, and face was also involved in second infant. Second infant also had mild respiratory distress. Surgical excision was performed successfully in both the infants. At the follow-up of one month after the surgical excision none had recurrence, but second child showed features of neuropraxia / paresis involving the mandibular branch of right facial nerve and was expected to resolve with time as there was no obvious nerve injury observed during surgical excision.

Conclusion

Large cystic hygromas at neck region are not unusual findings and may present with respiratory distress. Surgical excision remains a best option to manage large cystic hygromas. Prior to surgical excision, sometimes it is necessary to aspirate the cystic fluid to prevent or relive the obstructive symptoms.

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