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Case Report

Giant Cystic Hygroma Encapsulating the Internal Jugular Vein and Causing Severe Airway Obstruction: A Rare Presentation and Review of Literatures

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Abstract

Background: Cystic hygromas are benign congenital malformations of the lymphatic system resulting to abnormal dilatation of the lymphatic vessel. It is commonly located in the head and neck region with potential for local and systemic complications particularly those encasing and compressing vital structures.

Case Report: A three-year-old male who presented at the emergency room with a two weeks history of difficulty in breathing and progressively increasing right sided neck mass noticed after birth with associated fever, dyspnea, cough, nasal discharge and dysphagia to solids. He had a huge right sided neck mass measuring about 10x14 cm in diameter, fluctuant, warm to touch, non-tender and transilluminates well. The right lateral wall of the oropharynx was bulging and communicated with the mass. Radiological studies revealed a neck mass with marked compression and lateral deviation of the trachea, retropharyngeal extension with medial displacement of common carotid artery and internal jugular vein. He had emergency tracheostomy to relieve upper airway obstruction and was commenced on broad spectrum antibiotics, anti-inflammatory and antipyretics. He subsequently had an elective complete excision of the cystic sac which was found to be passing through and around the carotid sheath, encapsulating the internal jugular vein, separating it from the common carotid artery and vagus nerve.

Conclusion: Cystic hygroma when neglected can lead to severe complications with attendant increase for morbidity and mortality. Complete surgical excision when possible is the recommended modality of treatment with the lowest chances of recurrence. In so doing, extreme care including multidisciplinary collaboration should be undertaken to preserve the surrounding vital structures.

Keywords: Cystic Hygroma; Lymphatic Malformations; Surgical Excision; Carotid Sheath

Background

Lymphangiomas are benign congenital malformation of the lymphatic system resulting to abnormal dilatation of the lymphatic vessel. It is found most commonly in the cervicofacial region and axilla, but could be located in any part of the body [1-4]. They are commonly classified into capillary lymphangioma or lymphangioma

simplex, cavernous lymphangioma and cystic lymphangioma or cystic hygroma which constitutes approximately 90% and has a global incidence of 1:6000 live births [2,5-8]. Over 80% are evident at birth, in other cases though present, may be inconspicuous. Almost all cases will be obvious at 2years of age [4,9].

They are thought to arise from sequestration of lymphatic tissues from the major lymphatic sacs during embryological development of the lymphatico-venous system. These sequestered tissues fail to communicate with the remainder of the lymphatic or venous system, subsequent dilatation of the of these sequestered tissues results in cystic morphology of the lesion [2,5,8-10]. Certain chromosomal anomalies such as Down syndrome, Turner syndrome, Klinefelter syndrome, trisomy 18 and 13 have been linked to lymphatic malformations [1,11-13]. Where possible, karyotyping should be done to rule out these other associations. Intrauterine alcohol exposure has also been implicated [6,7].

Pattern of presentation is dependent on the location of the lesion [4]. They may present with sudden increase in size following infection or intralesional haemorrhage [5,10,14]. Pressure effect may result in dyspnea, noisy breathing (stridor), feeding difficulties with resultant failure to thrive or deformation of surrounding bony cranio-facial structures.

Surgical excision remains the mainstay of treatment as it offers the best opportunity for complete removal of the diseased tissue, and hence lower chances of recurrence [5,7,15-17]. Surgery may be multiple, complex and staged before complete excision is achieved, though subsequent surgeries are usually more difficult due to fibrosis [5]. Other treatment modalities which have been employed in other studies with variable results include simple drainage, serial aspirations, radio-frequency ablation and cauterization. Medical treatment involve administration of sclerosing agents such as bleomycin, OK-432, doxycycline, pure ethanol, sodium tetradecyl sulfate etc. [2,5,6,18]. Sclerotherapy is best when it is comprised of macrocysts.

Case Report

A three-year-old male admitted through the children emergency room (CHER) with a two-weeks history of difficulty in breathing on account of a progressively increasing right sided neck mass noticed after birth. The mass was initially the size of a table tennis ball, but rapidly increased recently to about the size of an orange. No swelling in any other part of the body. There was associated fever, dyspnea, cough, nasal discharge and dysphagia to solids. Pregnancy history was uneventful. He is the second child in a set of twins and the other sibling is well.

They had initially presented to a non-specialist when the swelling was first noticed but was erroneously reassured and sent home, with no follow-up and no proper referral.

On presentation, he was highly irritable, Febrile and in respiratory distress (tachypneic, flaring of ala nasi, intercostal and subcostal recessions, oxygen saturation was 92% on room air), noisy breathing (stridor).

He had a huge right sided neck mass measuring about 10x 14cm in diameter, fluctuant, warm to touch, non-tender and transilluminated well. The right lateral wall of the oropharynx was bulging antero-medially. Aspirate yielded a straw-colored fluid which was negative for malignant cells on cytological evaluation.

Neck ultrasonography showed a lobulated mixed echogenic mass on the right side of the neck extending from angle of the mandible to region of the clavicle, with cystic and solid components. The internal jugular vein and common carotid artery were displaced medially. Plain radiograph showed a right lateral soft tissue swelling with fullness of the prevertebral space, marked compression and lateral deviation of the trachea.

A cervical computed tomography scan revealed a non-enhancing, low attenuating right-sided neck mass that extends to the retropharyngeal region with associated obliteration of the adjacent airway. Hematological investigations showed leukocytosis ($12x10^3$/µl) with neutrophilia (78%) and elevated erythrocyte sedimentation rate (ESR) of 113 mm/hr.

He had an emergency tracheostomy to relieve upper airway obstruction and was commenced on broad spectrum antibiotics, anti-inflammatory and antipyretics which he responded very well to and became clinically stable. He subsequently had an elective complete excision of the cystic sac a month later; Modified Blairs incision was made and sub-platysma flap raised, investing fascia was divided and adjoining omohyoid, sternocleidomastoid and posterior belly of digastric muscles were carefully dissected and

reflected away from the cystic sac at the carotid triangle (Figure 1), where it courses through the carotid sheath and encapsulates the internal jugular vein, separating it from the common carotid artery and the vagus nerve (Figure 2). The Great vessels of the neck were carefully skeletonized and the cystic sac divided and completely excised (Figure 3 and 4). Surgical drain was left *in situ* for 48 hours. Tissue was sent for histology and confirmed the diagnosis of cystic hygroma (Figure 5). Two weeks postoperatively, the tracheostomy tube was downsized and subsequently decannulated. Patient had been followed up for a period of 8 months after discharge with no sign of recurrence.

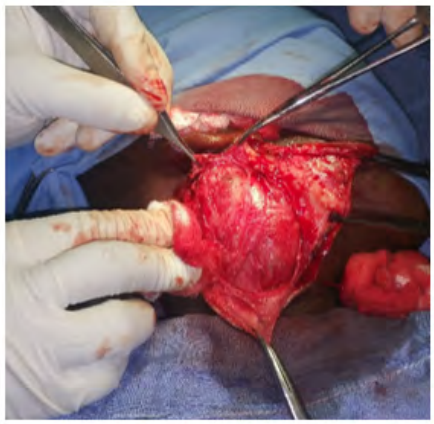

Figure 1: Cystic Mass *in situ*.

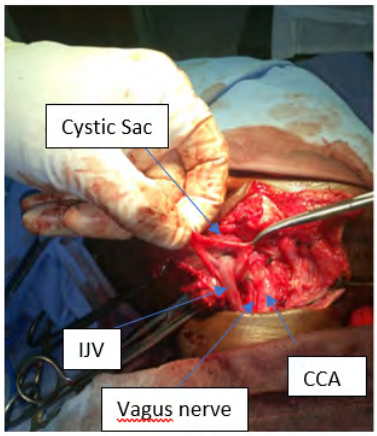

Figure 2: Cystic sac encapsulating the IJV and separating it from CCA and Vagus nerve.

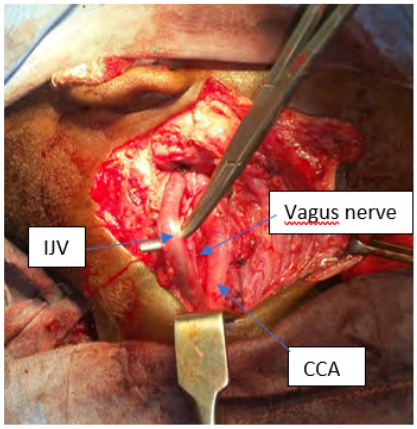

Figure 3: IJV, Carotid artery and vagus nerve left intact after removing the cystic sac.

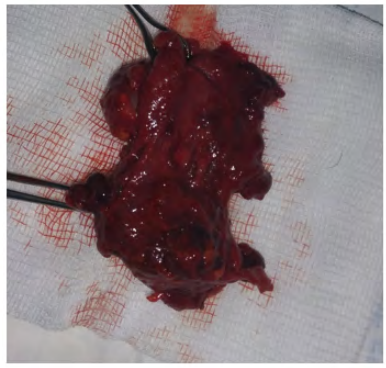

Figure 4: Cystic mass removed en-bloc.

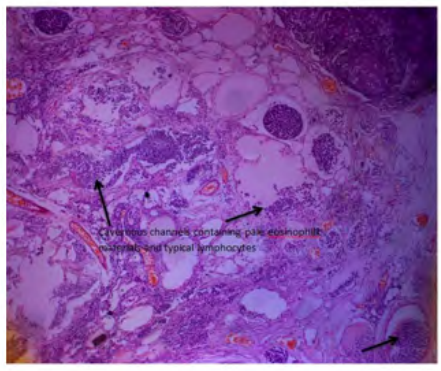

Figure 5: Section showing multiple cavernous vascular channels containing pale eosinophilic materials and lymphocytes. Also seen are secondary lymphoid follicles.

Discussion

Cystic hygromas are congenital benign lesions usually presenting as painless and asymptomatic swellings [9]. It usually increases gradually in size over time but could increase rapidly when infected or if there is bleeding into the mass with attendant pressure effect on surrounding vital structures [6-9,19]. This mass effect could result in dyspnea, stridor, cyanosis and dysphagia as observed in the patient above, thus necessitating emergency tracheostomy to relieve upper airway obstruction. Broad spectrum antibiotics, anti-inflammatory and antipyretics were administered to control infection and associated inflammation. Aspiration of the cystic hygroma can be performed as a temporary measure to reduce the size, and thereby, reducing its pressure effects on the respiratory and feeding passages [4,6,7,9]. A tracheostomy and feeding gastrostomy can be performed in extreme cases, especially in those with severe respiratory and feeding difficulties [9,16]. The primary focus of infection is usually from the upper respiratory tract via lymphatic spread or iatrogenic inoculation during fluid aspiration [7,9].

Lymphatic malformations could be difficult to manage in many occasions. Surgical excision of complex cystic hygromas involving deep and vital structures is such a herculean task and could prove a surgical dilemma especially when aimed at complete removal of the entire sac [9,16]. In the index patient the cystic sac was found passing through and around the carotid sheath, encapsulating the internal jugular vein (Figure 2 and 3), separating it from the common carotid artery and vagus nerve. These were not clearly shown by any of the radiological investigations done. We however proceeded to carefully and bluntly skeletonize the vessel, freeing the cystic sac looping around it, while maintaining the integrity of the membrane. The sac was then subsequently ligated, divided and delivered en-bloc satisfactorily (Figure 4), leaving the internal jugular vein, common carotid artery and vagus nerve intact. Studies suggests that outcome is better and recurrence is rare when all gross tissues are removed [2,6,7,9,15]. Some other potential complications during surgical excision includes injury to facial nerve, facial artery, thoracic duct and pleura [4].

Some authors have suggested watchful waiting for asymptomatic and indolent forms of lymphatic malformations [4,7,16]. Medical therapy involving the use of percutaneous sclerosing agents such as OK-432 (an inactive form of group A Strept pyogenes), bleomycin, pure ethanol, sodium tetradecyl sulfate and doxycycline etc. [2,5,6,18]. Gilony., *et al.* conducted a study in Israel to compare the outcome of patients with cystic hygroma treated with both medical and surgical therapy and discovered that surgical excision was associated with the lowest rate of recurrence [7]. This was the finding in some other studies [5,9,15].

Conclusion

Cystic hygromas when neglected can lead to severe complications with attendant increased morbidity and mortality. Early presentation, management and proper follow up by a specialist Head and neck surgeon could save the patient from these avoidable situations. Various modalities for treatment of cystic hygroma have been devised over time with variable success rates, however, complete surgical excision when possible is the recommended modality of treatment with the lowest chances of recurrence. In so doing, absolute care must be taken to preserve surrounding vital organs and neurovascular structures.

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