Massive Cervico-Lingual Cystic Hygroma

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Summary

Cystic Hygroma (CH) is an aberrant proliferation of lymphatic vessels creating fluid-filled sacs that result from blockage in the abnormal lymphatic system. These benign lesions are characteristic in their capability to grow to an enormously huge size has potential pressure effects on the neighboring structures. The massive expansion can also lead to gross disfigurement of the neck and face. In this paper, we present a rare case of cervico-lingual CH in a 17-year old Rwandese boy. Though the lesion had caused massive expansion of the tongue (12cm x 10cm) and led to gross deformity of the mandible, the boy had no signs of respiratory distress. His main concerns were inability to eat solid foods and compromised speech. The management challenges and literature review on cystic hygroma of the head and neck region are discussed.

Key Words: Cystic Hygroma, Cervico-Lingual

Introduction

Cystic Hygroma (CH) also known as cystic lymphangioma, or macrocystic lymphatic malformation is an aberrant proliferation of lymphatic vessels creating fluid-filled sacs that result from blockage in the abnormal lymphatic system. CHs can occur as single or multiple cysts that can be found in any anatomic site of the body but more often in the neck region (1). They may be present as a birth defect or be acquired and occur in1% of fetuses between the 9th and 16th week of pregnancy. In some instances they may progress in size to become larger than the affected fetus. The incidence of CH is estimated to be 1 case per 6,000-16,000 live births. CHs are benign lesions; however their capability to grow to an enormously huge size has potential pressure effects on the neighboring structures like compression of the airway and other vital structures like nerves and blood vessels. The massive expansion can also lead to gross aesthetic compromise or disfigurement of the neck and face (2,3). Management of this condition is mainly surgical, however, the use of various non-surgical interventions are also common.

Case Summary

A seventeen year old Rwandese boy presented at the dentistry department of the Kigali University Teaching Hospital (CHUK) with a tongue swelling that had been there since early childhood. The swelling was initially small painless and slowly expansile, however rapid growth had occurred in the three months preceding his presentation to the dental clinic. He was referred from the district hospital where treatment with antibiotics had been attempted. The boy was accompanied by his elder brother who gave his history due to speech difficulty occasioned by the massive tongue swelling. Despite the enormous enlargement of the tongue the boy had no breathing difficulty and showed no signs of respiratory distress.

His past medical history was non-contributory and his brother indicated that the boy had normal delivery as well as un-incidental childhood. He was attending school normally until the swelling became too large 3-months ago occasioning difficulty in feeding and speech.

Clinical examination revealed a boy with massive potato-like swelling bulging from the mouth and occluding the entire anterior oral opening but otherwise in satisfactory general condition. He was fairly hydrated, no pallor, no jaundice and no palpable cervico-facial lymph nodes. Detailed examination of the swelling revealed a distended fluid filled cyst-like mass involving the substance of the tongue ballooning out of the mouth and covering the upper and lower teeth. The swelling appeared translucent on shinning a torch light and measured about 12cm by 10cm making intra-oral examination impossible (Figure 1). Further examination revealed a second

swelling in the midline of the submandibular region of about 6cm by 7cm. This was soft and fluctuant with normal overlying skin (Figure 2)

Figure 1. Photograph of the boy with cervico-lingual cystichygroma at initial presentation

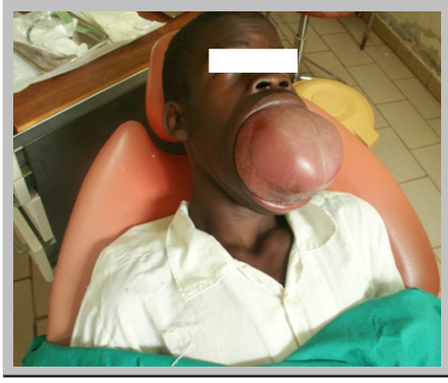
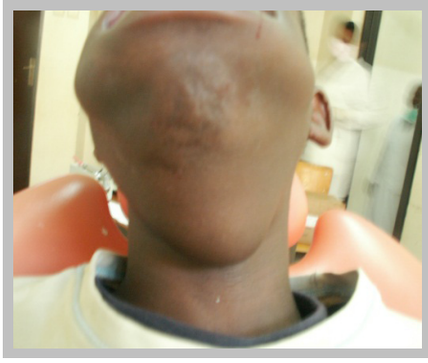


Figure 2. Neck extension of the lingual swelling of the Cystic Hygroma



Computerised Tomography (CT) scans revealed a well circumscribed cystic cavity in the tongue with contiguous cervical extension (Figure 3). The lesion had features of slow growth but had expanded significantly as was evident in the extent of jaw deformities which was well demonstrated in the 3D CT reconstruction (Figures 4).

Figure 3.Saggital CT scan demonstrating the cystic nature of the tongue lesion and its cervical extension.

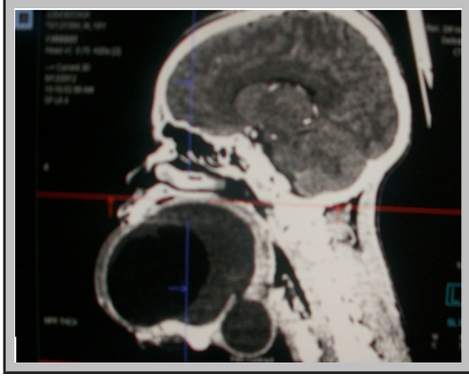
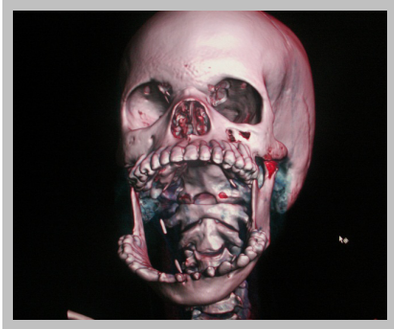


Figure 4.3D Reconstruction of the CT scan illustrating the extensive jaw deformity suggestive of the slow growing nature of the lesion.



An attempt to aspirate the contents was unsuccessful as the contents were too thick. Emergency incision was done and thick pus-like fluid expressed to decompress the lesion and relieve the pressure on the airway as well as facilitate improved feeding. He was then scheduled for elective tracheostomy in preparation for definitive surgery. However two days later, the swelling increased very rapidly in size, the patient was in severe pain and was experiencing difficulty in breathing. Another incision was done and more than 50mls of pus drained. Some thick cheesy material was also expressed through the sublingual incision. He was put on intravenous antibiotics cefuroxime and metronidazole. Tracheostomy was done immediately prior to the surgical excision of the lesion.

The cystic lesion was first dissected through the intra-oral approach; the capsule was separated from the surrounding connective tissue by a combination of blunt and sharp dissection (Figure 5). The lesion was tracked as far down as possible in the floor of the mouth and found to have split through the myelohyoid muscle into the submandibular region. An extra-oral incision was then made on the submandibular region, care being taken to ensure that the incision was low enough to spare the marginal mandibular nerve. The dissection was carried through the platysma muscle and the neck extension of the capsule exposed. The capsule was tracked cranially and found to be in continuity with the intra-oral cyst. Further blunt dissection enabled the trans-oral release of the capsule from the mouth through the submandibular incision (Figure 6). The feeding lymphatic channel was then tracked caudally to the supraclavicular area and ligated with 2/0 silk suture before excising the lesion completely. The surgery was uneventful and the diagnosis of CH or lymphangioma confirmed through histopathology (Figures 7&8).

Figure 5. Intra-oral dissection of the tongue lesion

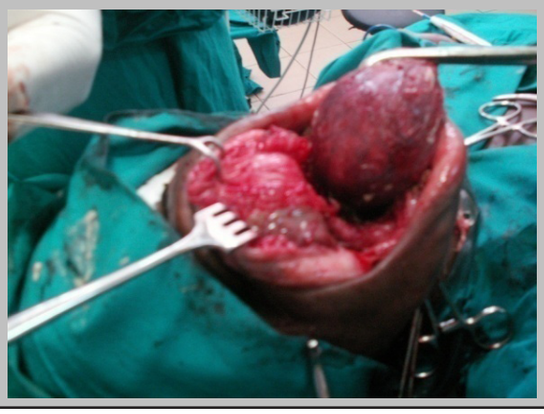


Figure 6 . Delivery of the whole lesion through the neck incision

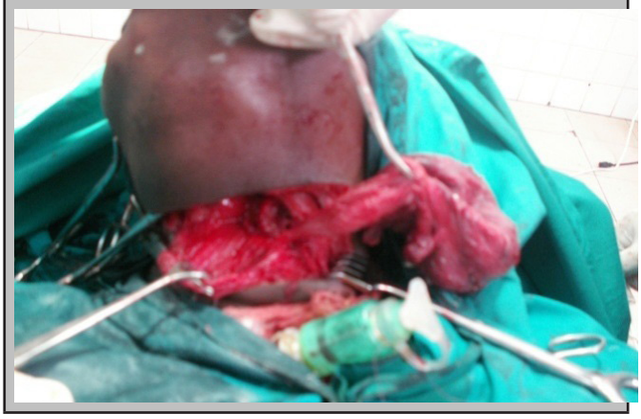


Figure 7. Histopathological appearance showing cystic spaces with endothelial lining

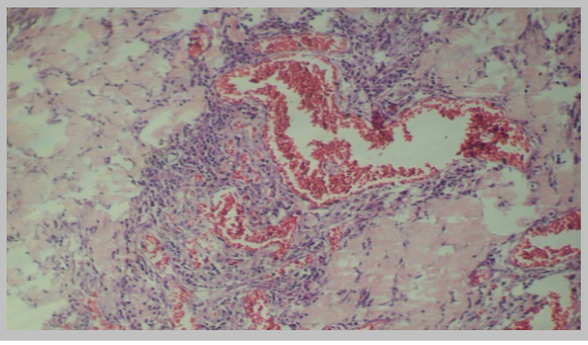


Figure 8. 3rd day post-operative appearance of the patient.

Discussion

CH can occur in isolation or be syndromic in association with other birth defects. Etiology remains largely unknown but various factors are thought to be responsible including environmental factors and genetic factors. Environmental factors that have been implicated in CH etiology include; maternal viral infections and maternal substance abuse, like alcohol consumption. Karyotype abnormalities are found in 25 -70% of children with CH. The Genetic syndromes reported with CH include; Turner's syndrome, Down's syndrome, Noonan syndrome and chromosomal abnormalities such as trisomy 13, 18, and 21. Isolated CH can also be inherited as an autosomal recessive disorder. Molecular studies suggest that defects in vascular endothelial growth factor C (VEGF-C) and its receptors may have a role in the development of lymphatic malformations (4). Acquired lymphangiomas can arise from trauma (including surgery), inflammation, or obstruction of lymphatic drainage. In this case the swelling lesion was probably present since birth, there were no features suggestive associated syndromes nor were there any member of the family with a similar condition.

There are both medical and surgical approaches to management. Although some schools of thought recommend watchful waiting in patients who are asymptomatic. Rapid enlargement over a short period of time has been reported and in our case the history indicates that the expansion might have been rapid in the precedent 3months. However, purulent infection set in, worsening the swelling when he was awaiting tracheostomy. This progression made emergency decompression by incision and drainage necessary (3,5).

The medical treatment of CH consists of the administration of sclerosing agents; including attenuated group A Streptococcus pyogenes (OK-432), bleomycin, absolute ethanol, sodium tetradecyl sulfate, and doxycycline. The use of interferon alpha 2a and fibrin sealant have also been reported in literature (6–8).

The mainstay of treatment is surgical excision. Acute infection if present should be treated with appropriate antibiotics prior to surgery. Attempt should be made to excise the entire lesion or to remove as much as possible while sparing vital neurovascular structures. Complete excision has been estimated to be possible in roughly 40% of cases (9). It is recommended that CHs are removed in one procedure since secondary excisions are complicated by fibrosis and distorted anatomical landmarks (10,11).

In emergency situations, aspiration with an 18-gauge or 20-gauge needle may obviate the need for an

emergency tracheostomy (12). A study by Burezq et al documented success with serial aspiration of CH (13). However, some authors contend that aspiration is often followed by recurrence, hemorrhage, or infection. Other non-surgical modes of treatment that have been tried include; Radiofrequency ablation in intraoral lymphatic malformations, especially microcystic lesions, magnetic resonance–controlled laser-induced interstitial thermotherapy.

Conclusion

Cystic hygroma can grow to a very massive size; when the lesion involves the cervicofacial region, securing the airway is the priority prior to surgical management.

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