Lymphangioma of the Head and Neck in Children: A Report of Six Cases

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ABSTRACT

Lymphangiomas are uncommon congenital malformations of the lymphatic system that involve the skin and subcutaneous tissues. They are regarded as malformations that arise from sequestrations of lymphatic tissue that fail to communicate with normal lymphatic system during embryogenesis. They are rare vascular malformations. We therefore reported six cases that occurred in the head and neck region that were treated in our Centre.

Keywords: Lymphatic malformation, Lymphangioma, vascular, neck, cystic hygroma.

INTRODUCTION

The lymphatic system is the network of vessels responsible for returning fluids to the venous system. Lymphangiomas are benign hamartomatous malformations arising from the sequestration of lymphatic tissue that fail to communicate with the normal lymphatic system during embryogenesis (Grasso et al., 2008 & Weiss et al., 2008). Majority of them are superficial, but a few may extend deeply into the connective tissue. The direct cause of lymphangioma is a blockage of the lymphatic system as a fetus develops, although symptoms may not become visible until after the baby is born.

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Lymphangiomas have marked predilection for the head and neck region. It had been reported that about

50-70% of the lymphangiomas occurred in the head and neck region (Guarisco et al., 1991 & Rathman et al., 2005). The occurrence in other parts of the body such as axilla, groin, liver, mediastinum and leg had been documented (Fernandez et al., 2008 & Matsunmoto et al., 2010).

However, intraoral lymphangiomas occur more frequently on the dorsum of tongue, followed by palate, buccal mucosa, gingival, and lips (Jeeva et al., 2005 & Uguru et al., 2011). Lymphangioma are frequently found in the neonatal period and childhood but rarely in adulthood (Uba & Chirdan 2006). It had been estimated that lymphangioma occurs in about 1 in 4000 births (Olajide-Seyi et al., 2020).

These malformations are either congenital or acquired. Congenital lymphanigomas are often associated with chromosomal abnormalities such as Turner syndrome, although they can also exist in isolation (Naidu et al., 2004). Lymphangiomas are commonly diagnosed before birth using fetal ultrasonography. Acquired lymphangiomas may result from trauma, inflammation, or lymphatic obstruction. There are three distinct types of lymphangiomas: which are capillary, cavernous and cystic, each with their own distinct symptoms. They are distinguished by the depth and the size of abnormal lymph vessels, but all involve a malformation of the lymphatic system. They present as bulging masses which occur deep under the skin, typically on the neck, tongue and lips, and vary widely in size, ranging from as small as a centimeter in diameter to several centimeters wide (Mirza et al., 2010). In some cases, they may affect an entire extremity such as a hand or foot (Olajide- Seyi et al., 2020). Although they are usually painless, the patient may feel mild pain when pressure is exerted on the area. They present in different colors such as: white, pink, red, blue, purple, and black; and as the pain lessens the lighter the color of the bump (Kocher et al., 1995).

Histopathological features consist of multiple intertwining lymph vessels in a loose fibrovascular stroma. Histologic features of the lesions may be of three types: capillary, cavernous, or cystic lymphangioma. Capillary lymphangioma composed of solid proliferation of small, capillary sized lymphatic channel. Lymphatic vessels display attenuated layer of endothelium, thin, adventitial layer surrounds lymphatic spaces. Lumen of lymphatic spaces may be filled with clear proteinaceous fluid containing small lymphocytes. Lymphoid aggregates present in stroma. Cavernous lymphangioma is characterized by grossly distended lymphatic channels, hemorrhage into lumen may simulate cavernous (Carpenter et al., 1996).

Surgical excision is the treatment of choice for lymphangiomas. The prognosis is good for most patients, although large tumors of neck and tongue may result with airway obstruction and death. We therefore reported six cases of lymphangiomas: five occurred in the neck and one in the dorsum of the tongue, they were treated in Maxillofacial Unit of Barau Dikko Teaching Hospital, Kaduna, Northwest, Nigeria.

Table 1: Cases Reported with Age and Sex

Cases	Age	Sex	Location of tumour	Co-existing pathology	Histology	Treatment	Complications
1.	6years	F	Posterior Neck	None	Capillary	Excision	Recurred
2.	5month	M	Posterior Neck	None	Cystic	Enucleation	None
3.	4month	M	Anterior &Posterior Neck	Haemangioma	Cystic	Enucleation	None
4.	3month	F	Posterior Neck	None	Cystic	Enucleation	None
5.	4month	F	Posterior Neck	None	Cystic	Enucleation	None
6	6years	F	Tongue	None	Capillary	Excision	None

Case no. 2

History and clinical examination

A 5-months-old male infant was referred to the Maxillofacial Clinic of Barau Dikko Teaching Hospital, Kaduna from the Paediatric Outpatient Clinic of the same hospital with complaints of right sided neck swelling, which was present since birth but has been gradually increasing in size. The child weighed 5.8kg. On examination, there was a large swelling in the cervical region on the right side of neck measuring 15x9 cm^2 (Figure 1). The swelling was non-tender, fluctuant, easily compressible and transluminant, extending into both anterior and posterior triangle of the neck.

Investigations and Differential Diagnosis

Aspiration biopsy yielded yellow coloured fluid. Ultrasonography of neck region showed cystic swelling, which measure 29.0cm^3. There were significant internal echoes within it and has septations. It is avascular and appears separate from the thyroid gland. The great vessels on the right side are laterally displace but not compressed. There were small adjacent lymph nodes with largest measuring 1.5cm x 0.8cm. The left side of the neck was preserved. A diagnosis of cystic hygroma was made. Magnetic Resonance Imaging (MRI) showed multiple cystic lesions within left masseter muscle and extending to

anterior aspect of the neck and into subcutaneous fat plane. Blood investigations showed haemoglobin concentration of 11.5g/dl, and chemistry showed urea 1.1mmol, sodium 138mmol/l, potassium 4.1mmol/l, chloride 98mmol/l, bicarbonate 20mmol/l and creatinine 20mmol/l. Differential diagnosis include: branchial cleft cyst, haemangiomas, dermoid cyst and thyroid mass.

Surgical Management

Endotracheal intubation was done and under general anesthesia the cyst was exposed through submandibular incision and by careful dissection, the cyst was enucleated (Fig. 2). Post-operative period was uneventful. The result showed cyst wall lined by predominantly flattened epithelium, focally columnar and papillary. This overlies a fibrocollagenous wall with areas of marked lymphocystic aggregation. Serous salivary glands noted. The diagnosis was given as cystic lymphangioma (hygroma) (Figure 4).

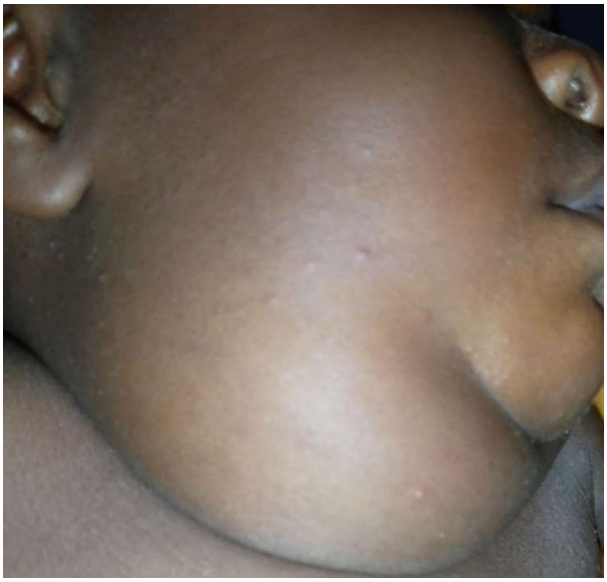

Figure 1. Gross appearance of swelling: pre-operative clinical photograph, showing large swelling in the right neck.

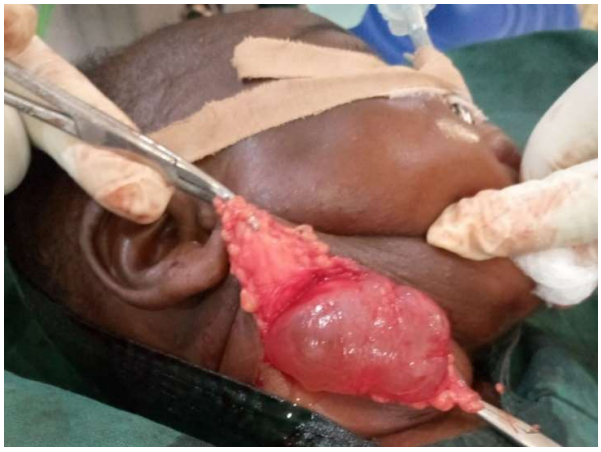

Figure 2. Intraoperative photograph of cystic hygroma showing Ballooning appearance

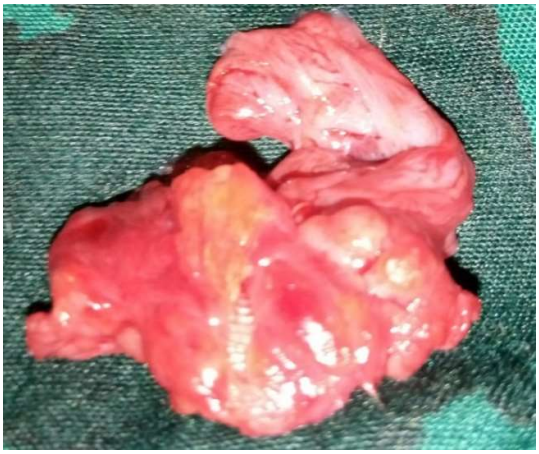

Figure 3: The cyst wall was submitted for. histopathology examination.

DISCUSSION

Lymphangiomas are benign, hamartomatous tumors of lymphatic vessels. They represent developmental malformations that arise from sequestrations of lymphatic tissue that do not communicate normally with the rest of the lymphatic system. They are usually filled with a clear protein-rich fluid containing few lymph cells. It can also occur in association with hemangioma. Case 3 in this study was 4-months old male child with bilateral cystic hygroma on the neck and hemangioma in the floor of the mouth (Figure 5). Aspiration biopsy of the swelling in the neck yielded yellow colored fluid and the floor of the mouth yielded frank blood. Diagnosis of lymphangioma co-existing with hemangioma was made in case .

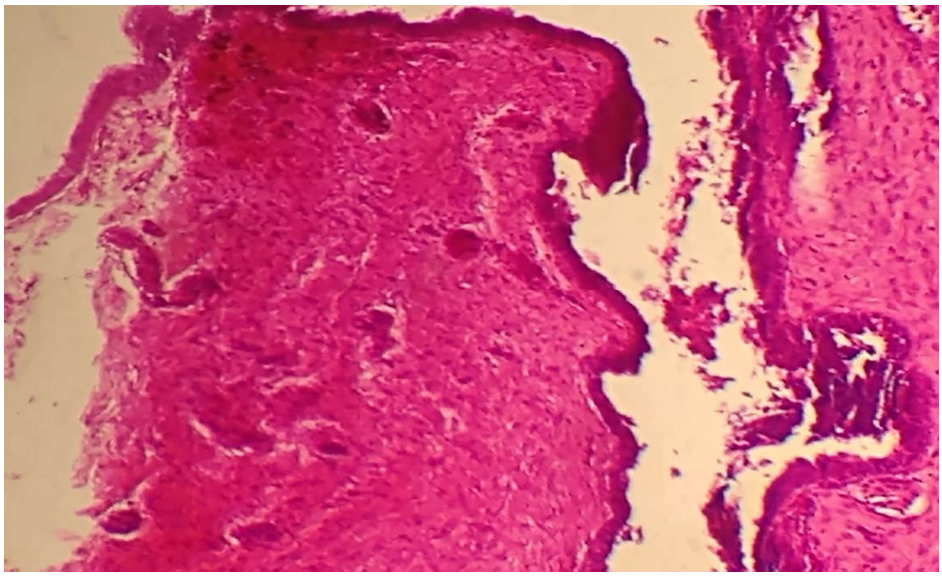

Figure 4: Microphotograph of cystic hygroma.

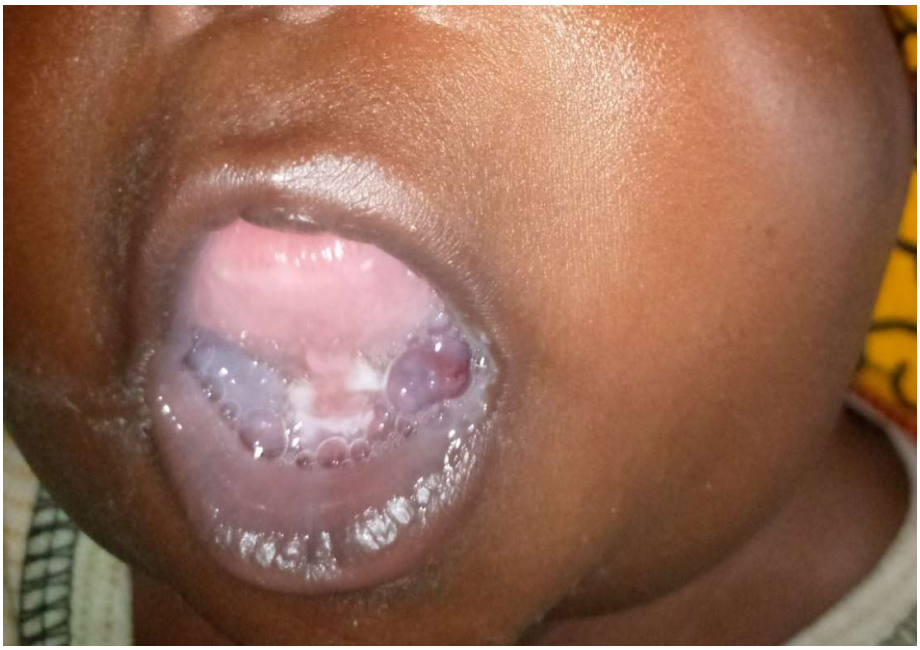

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Figure 5: A 4-months old male child with hemangioma in the floor of the mouth and bilateral cystic lymphangioma in the neck

Three types of lymphangiomas had been described in literature based on their microscopic characteristics, and they include: lymphangioma simplex (capillary lymphangioma), cavernous lymphangioma and cystic lymphangioma (cystic hygroma) (Neville et al., 2009). This study reported four cystic lymphangiomas and two capillary lymphangiomas (Table 1). Several researchers had reported higher occurrence in males than females at the ratio of 2:1 (Naidu et al., 2004). In contrast, this study showed higher occurrence in females than males at the ratio of 2:1 (Table 1). This may be due to small size of the study population. The age range of cases in this study was from 3-months to 6-years, which showed that the malformation occurred mostly in children. This was in agreement with other researchers (Uguru et al., 2011; Uba et al., 2006).

Capillary lymphangiomas in this study were composed of small, capillary-sized lymphatic vessels and one case was located in the deep in the neck and was adherents to jugular vein, therefore it was not excised completely at surgery, and one occurred in the dorsum of the tongue which presented with features of macroglosia and V- shaped excision was done to excise the lesion and shape the tongue. However, all the cystic hygromas in this report occurred in the neck and it can be misdiagnosed with other cystic neck swellings like: branchial cleft cyst, hemangiomas, dermoid cyst, thyroid mass and other childhood tumours. Cystic hygromas in this study were large masses in the

anterior and posterior triangles of the neck, and aspiration of the tumour yielded yellow coloured fluids (Figure 2). Histology of cystic hygroma showed cyst wall lined by predominantly flattened epithelium, focally columnar and papillary. This overlies a fibrocollagenous wall with areas of marked lymphocystic aggregation (Figure 4). In the intraoral lymphangiomas, lymphatic vessels are located just beneath the epithelium, often replacing connective tissue papillae. Case 6 occurred in the dorsum of the tongue just beneath the surface epithelium.

Moreover, lymphangiomas may be described based on the location in relation to the hyoid bone whether present above or superior to the hyoid bone (suprahyoid), below or inferior to the hyoid bone (infrahyoid), and whether the lymphangiomas are on one side of the body (unilateral) or both (bilateral) (Shan et al., 2020). In this study the cystic hygromas were situated below the hyoid bone, these locations were determined with ultrasonography. Majorities (5 cases) were unilateral and case 2 only occurred bilaterally on the neck (Figure 5).

Moreover, in prenatal cases, lymphangioma is diagnosed during the late first trimester or early second trimester, using an ultrasound. Other imaging methods such as CT and MRI scans are useful in treatment planning; it delineates the size of the lesion, and determines its surrounding vital structures. T2-weight MRI is useful to differentiate lymphangioma from surrounding structures due to its high T2 signal (Mirza et al., 2010). Ultrasonography was done for all our neck lesions to determine the closeness of the lesions to major neck vessels such as carotid artery and jugular vein.

Surgical excision is the treatment of choice for lymphangiomas. However, because of the non-encapsulated and infiltrating nature of the lymphangioma, complete removal is difficult, and hence recurrence is common. Moreover, other various treatment modalities have been considered for successful treatment of cystic hygromas, this includes: injection of sclerosant agents, repeated aspiration, injection bleomycin, radiotherapy and surgical exploration (Oxford et al., 1995). Ibrahim et al., 2009 reported non-surgical successful management of a huge life threatening cervico-mediastinal cystic hygroma with combination of fluid drainage and sclerotherapy. Surgical enucleation under general anesthesia was successfully done for all our cases with neck massess (Figure 3). Surgical procedure may be difficult and challenging owing to multicystic nature of the lymphangioma, especially the thin wall and proximity to vital neurovascular structures, therefore many non-surgical options have been advocated such as sclerotherapy before surgical excision have been advocated for lesions extending to major vessels or vital structures. Draining lymphangiomas of fluid provides only temporary relief, so that they can be removed surgically (Uguru et al., 2011). Follow up of our cases was difficult as most of our cases did not come for reviews. Nevertheless, it could be concluded that lymphangiomas could be treated. However, respiratory challenges most encountered was life threatening conditions, which necessitated close monitoring of the patients and oxygen therapy until the patient was stable.

CONCLUSION

Midwives, Gynaecologists and birth attendants needed to carefully examine the neck and oral cavity of all newborns for early detection of lymphangiomas.

Declaration of patient consent.

The consent to use patient images was obtained from the parents.

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