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Case Report

Giant cystic hygroma of the neck in a 3-year-old child: Clinical case and literature review

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Abstract

Giant cystic hygroma of the neck often represents a pathology in children with severe potential complications and considerable technical difficulties in surgical therapy, which often makes it necessary because the alternative therapies provided are not available. A case of a cystic hygroma with a maximum diameter of 10 cm, located in the neck of a 3-year-old girl, with sudden rapid growth and an underlying inclusion cyst compressing the internal jugular vein, is described. The case was resolved with radical surgical therapy, without complications, without recurrence, and with acceptable aesthetic results.

Introduction

Lymphatic malformations of the neck are classified into simple lymphangioma, cystic lymphangioma and cystic hygroma. Since this is a disontogenic pathology of the lymphatic system, its manifestation is present in childhood, and often, if of conspicuous size, it can represent a considerable aesthetic and functional damage to the neck. These malformations can also increase the the risk of compression on nerve-vascular structures or on the upper respiratory tract, especially if intracistic haemorrhagic complications occur with sudden volumetric increase in mass.

The most effective treatment can be radio intervention, with aspiration of the cyst and injection of sclerosing substances, or surgery. In emergency conditions, in case of abnormal and sudden volumetric increase, incision and drainage is often required.

The clinical case of a 3-year-old girl with right laterocervical cystic hygroma operated in deferred urgency for rapid volumetric increase in mass and compression of the underlying structures is described.

Case report

A 3-year-old girl arrived in the emergency room due to a sudden volumetric increase in a mass that involved the laterocervical region of the neck in its entirety (Figure 1), already studied at another hospital with ultrasound and magnetic resonance, and classified as cystic lymphangioma. The child was conspicuously suffering from evident signs of acute inflammation of the lesion, and of the overlying skin. The mass on Ultrasound examination showed a mass of over 10 cm in maximum diameter. The mass was multiloculated and numerous lymphoglands were present and increased in volume up to 2 cm in longitudinal diameter. The little girl complained of difficulty breathing even though the saturation was normal.

The little girl underwent, under local anesthesia, first topically with anesthetic cream and then with lidocaine infiltration, a small incision (about 5 mm) and drainage of the cyst, with leakage of serum-hemorrhagic fluid, which was sent for cytological examination and bacterial cultivation. A small laminar drainage was left in place for a few days, until the acute inflammation subsided and the swelling disappeared. The girl was discharged after 5 days with the planning of a possible treatment of the lesion with radio interventional method at another hospital.

About 2 weeks after the discharge, the mass presented a new and sudden volumetric increase. At the ultrasound examination it appeared again with a diameter of 10 cm, and below the lymphangioma there was a new oval lesion, apparently cystic, with a diameter of 2 centimeters, which compressed the internal jugular vein reducing its diameter by 50% (Figure 2). The patient underwent a second surgical therapy.

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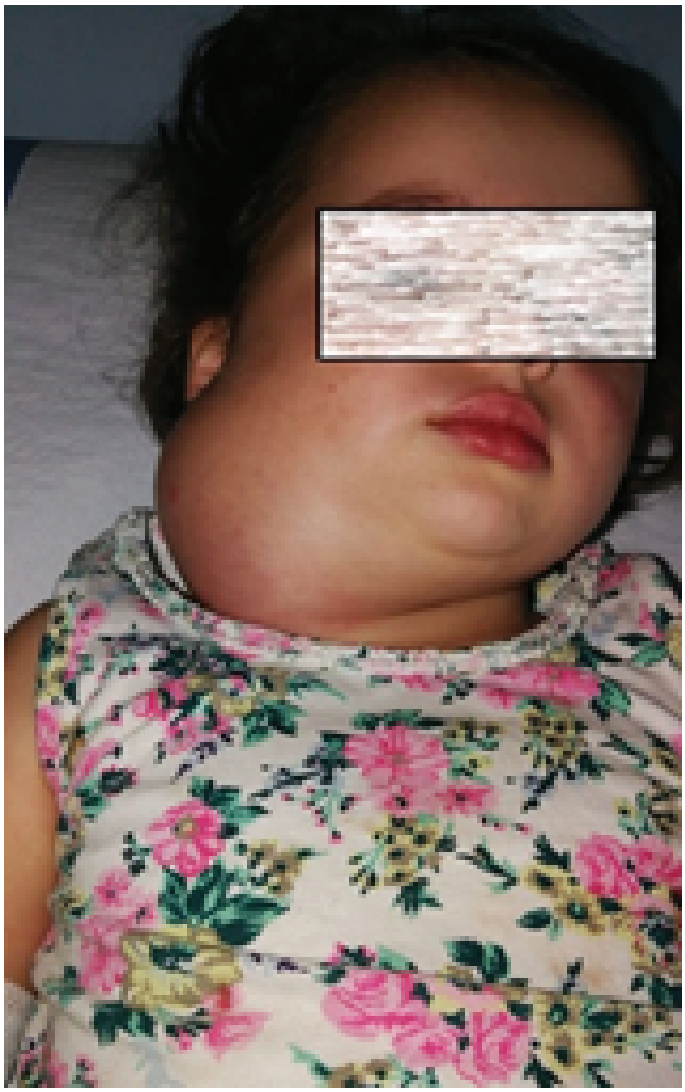

Figure 1. *The child before the operation.*

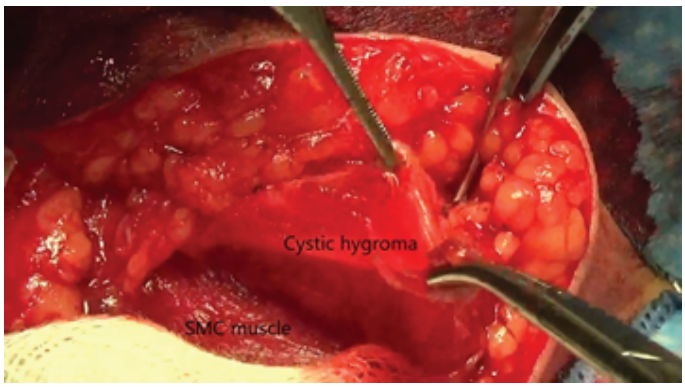

Figure 2. *Intraoperative aspect of the hygroma*

Surgical procedure: general anesthesia with oro-tracheal intubation. Supine position of the patient, with cervical spine in hyperextension and rotated 30 ° to the left. Right laterocervical incision along the anterior border of the sternocleidomastoid muscle. Isolation of the voluminous mass, which extends from the jugulo-digastric to the supraclavicular fossa, and difficult release of the cyst from the surrounding structures, with identification and isolation of the internal jugular vein, the hypoglossal nerve andthe mandibular branch of the facial nerve. Removal of the mass and three lymph nodes of stations II and III; isolation of the cystic lesion described by ultrasound, tenaciously adhering to the anterior wall of the internal jugular vein, and its removal en bloc (Figure 3).

The histological examination confirmed the presumptive diagnosis of cystic lymphangioma; there were reactive inflammation of the lymph nodes; the lesion was labeled as a cyst containing pigmented macrophages.

At the three-month follow-up, the patient showed no signs of recurrence, and the internal jugular vein had normal caliber and flow.

Discussion

Lymphangiomas are benign, hamartomatous, congenital malformations of the lymphatic system. They are usually classified as lymphangioma simplex, also known as capillary lymphangioma, cavernous lymphangioma and cystic lymphangioma. Cystic lymphangioma is commonly referred to as cystic hygroma. Lymphangiomas may also be classified on the basis of sizes of their cysts as microcystic, macrocystic and mixed lymphangiomas. Microcystic lymphangiomas consist of cysts which measure less than 2 cm in size, macrocystic lymphangiomas consist of cysts which measure more than 2 cm in size, and in mixed lymphangiomas, the cysts are of variable sizes.

Cystic hygromas are relatively rare lesions and approximately 50–75% of them are evident at birth. Up to 90% of cases are diagnosed in children less than2 years old, with most being diagnosed between the ages of 3 and 5.

Considering all cystic hygroma lesions and due to presence of extensive lymphatic system 75-90% of them are situated in the head, neck, axilla and extremities.. Other sites which also include the abdomen have been found to be affected only in 5% of the cases, 20% are axillary, and the remaining are inguinal, retroperitoneal and thoracic.

Embryologically, lymphangiomas are believed to originate from the sequestrations of lymphatic tissues and vessels that fail to establish a connection with normal draining vessels during intrauterine life. The exact embryonic origin is unclear; however, they are believed to be the result of developmental defects or cystic malformation of dilated lymphatic channels. It is understood that they arise from the remnants of embryonic lymphatic tissue that retains the ability to proliferate. They have been found to be associated with certain conditions, such as chromosome aneuploidies, hydrops fetalis, and intrauterine death. This failure of communication between lymph and venous pathways ultimately causes an accumulation of lymph, resulting in the formation of a cystic structure [1-3].

Due to infiltrative nature of hygromas within the soft tissues of the neck, these may extend from the posterior cervical area into the anterior compartment of the neck, may cross the midline and reach into the cheek or down into the mediastinum and axilla.

Clinically, cystic hygromas are large, soft, cystic, and non-tender masses. Transillumination is positive, unless it is complicated with haemorrhage or recurrent infections.

Although lymphangiomas are benign lesions, they can compress and infiltrate the vital structures or they can exist with complications such as intracystic haemorrhages, cyst

rupture or infections. Giant cystic hygromas of the neck and oral cavity may present with respiratory distress and dysphagia.

Cystic hygroma vary from 1 to 30 cm in size, the mean size in Stromberg's series was 8.0 cm. The skin overlying the cystic hygroma is often normal, and they are usually painless on palpation. An infected cystic hygroma is likely to present in a similar fashion; however, they may be exquisitely tender and have erythematous skin overlying them [4,5].

In general, symptoms depend on the anatomical location of the mass, and on their size and site; they vary from one mere presence of a painless, hoarseness, enlarging mass to respiratory compromise, dysphagia and difficulty in feeding with regurgitation. Very rarely, massive hygroma may present with symptoms of neural encroachment (brachial plexuses and recurrent laryngeal nerve). These symptoms can occur following compression of major structures within the neck, such as the larynx, trachea, esophagus, or great vessels. Patients may or may not also complain of limitation of neck movement.

If the patient has developed an abscess, this may cause a raised white cell count with raised inflammatory markers. Aspiration of the lump in combination with imaging is the gold standard of diagnosing cystic hygromas. Complications can arise due to the rapid enlargement of the cystic hygroma, causing infiltration of the neck. This can result in complete or incomplete airway obstruction, dysphagia, and obstructive sleep apnoea. Other complications include hemorrhage within the cystic hygroma leading to infection and later abscess formation.

The prenatal diagnosis of a cystic hygroma, which is made by using ultrasound, has been well documented in the literature. They are most commonly found in young infants or on prenatal ultrasound, and depending on the anatomical site, have the potential to obstruct the airway.

Post-natally, the diagnosis of a cystic hygroma is usually obvious and it is made by doing a physical examination and it can be confirmed with the ultrasonography (USG) of the swelling.

Various imagingprocedures like USG, computerized tomography (CT), or magnetic resonance imaging (MRI) are used to confirm the diagnosis. USG is the least invasive of all and typically shows multicystic lesions with internal septations. CT and MRI are valuable in the differential diagnosis, and are also helpful in further delineating the lesion and for planning surgery as they help to illustrate the involvement and proximity to neighboring structures. In this regard, MRI is superior to visuale the skull base extension, since it is not hindered by bony artefacts.

Aspiration is likely to produce serous, serosanguinous or straw-colored fluid.

Respiratory distress, infection, and aesthetic reasons are the main indications for treatment.

Theoretically, being cystic hygromas benign lesions, if asymptomatic, they do not necessitate treatment. Indications for treatment include disfigurement, large sizes, leakage of lymphatic fluid (rupture), recurrent infections, etc. Respiratory distress and dysphagia may be the reasons for seeking early/emergency treatment for lymphangiomas which involve neck and oral cavity.

The mainstay of management is surgical; however there are other options. Nonsurgical management of cystic hygroma has been advocated by some authors in order to avoid tedious and risky task of surgical resection. Especially in children, nonsurgical treatment options are suggested for lesions located over the parotid region in order to avoid the complications of surgery. In these cases, the other modalities which are used for treating lymphangiomas are; simple drainage, aspirations, steroids, sclerotherapy (OK-432, monoclonal antibody, bleomycin), radiation, laser excision, radio-frequency ablation and cauterization.

Radiotherapy (RT) which was used in the past is avoided for its ineffectiveness and potential for delayed carcinogenesis. Lymphangiomas are radioresistant.

Aspiration of hygromas is useful only to decompress, when the airways are compromed.

Sclerotherapy with bleomycin and OK-432 have been used in the treatment of cystic hygroma with some success, especially in children. Sclerosing agents such as boiling water, sodium morrhuate, alcohol, and 50% dextrose are also used, but the disadvantages of these agents are their unpredictable results and extensive sclerosis, which may make future surgery extremely difficult when it is required.

Cystic hygromas that turn into abscesses will require antibiotics, antipyretics, and analgesia, with or without subsequent surgical management. Surgical treatment, if needed, is usually withheld till the next three months after the antibiotic course.

Therefore, surgical excision still remains the ideal treatment. The objective in surgery of cystic hygroma is relief of vital structures obstruction, a good cosmetic result, good judgement must control the extent of the operation .

Surgical advocates, claim that these tumors may grow relentlessly, producing unacceptable cosmetic distortion of the face and neck, may compromise trachea and brachial plexus. The infiltrative hygromas as more risky while operating with the potential for incomplete excision, surgical sequel of neural injury, persistent lymphoedema, lymphocele and lymphorrhoea as being worse than the presence of the tumor itself.

Many times, it may not be possible to completely excise the lymphangiomas in a single sitting and it may require more additional operations. Recurrences are also reported to occur in 5-15% of the cases and additional operations may be required for the excisions of the recurrent lymphangiomatous cysts.

Giant cystic lymphangiomas of neck, head and oral cavity can also be successfully managed by surgery, with satisfactory results, although extreme precaution must be undertaken during complete excisions [6-9].

Conclusions

As lymphangiomas are benign lesions, the vital structures should not be sacrificed during their surgical resections.

Provided that the mass is completely excised, an excellent prognosis is of great possibility.

In the scientific literature there are not numerous cystic hygromas of the neck of considerable size, treated in children up to 3 years of age. The singularity of the case in question is due to the rapid volume increase of the lesion, and to the coexistence of an underlying cystic lesion compressing the internal jugular vein. In accordance with the data already reported in the literature, the chosen treatment was early and surgical; it was possible to completely excise the lymphangioma during first operation, without sacrificing any vital structures.

A two-month follow-up after the surgery showed no recurrence and there was only a good post-operative scar that was cosmetically acceptable.

Moreover,a multidisciplinary collaboration between pediatric surgeon, otolaryngologist, neurosurgeon, anesthetist, nurse, radiologist, physiotherapist is important for the management of these pathologies. The nurses are also a necessary member of the interprofessional group as they monitor the patient's vital signs and assist in the education of the patient and family.

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