CASE REPORT

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Cystic hygroma of the neck – case report

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Abstract

Cystic hygromas (CHs) are benign congenital malformations of the lymphatic system mainly diagnosed in small children aged less than two years old. They may give a multitude of local, sometimes severe complications. The most used method of treatment is surgical removal. In this paper, we present the case of a CH of a 13-year-old boy, localized in the right lateral region of the neck, diagnosed through magnetic resonance imaging (MRI), with excellent results of the surgical treatment.

Keywords: cystic hygroma, lymphangioma, congenital malformations, surgical therapy.

Introduction

Cystic hygroma (CH), also known as lymphangioma, has its origins in the embryonic life, being an abnormality in the development of the lymphatic system [1–3]. It represents 5% of the benign congenital malformations occurring during childhood [4].

CHs are usually observed in children under two years old [5]; they are quite rare in adults and are thought to occur due to the proliferation of lymph vessels in response to trauma and/or head and neck infections [1, 6, 7].

Most commonly, CH presented in the head and neck region (75–80%), clavicle and axillary areas, as a soft, benign, and painless mass [8, 9]. The most common site for the tumor is the posterior triangle of the neck and it may involve vital structures, such as the sympathetic chain, carotid sheath content, and branches of the hypoglossal, lingual, and the facial nerves [2]. The lesion is the most often unilateral, and doughy on palpation.

In children, cervical lymphangiomas are potentially life threatening due to direct localization or external compression on the airway [10].

Aim

We present the case of a 13-year-old boy who insidiously developed a tumor in right lateral region of the neck, which turned out to be a hygroma.

Case presentation

A 13-year-old boy presented to the Children's Ear, Nose & Throat (ENT) Department, within the No. 1 Emergency Clinical Hospital, Timişoara, Romania, following referral from his general practitioner (GP), with a main complaint of large swelling on the right side of the neck for the past three years. The swelling was small initially, subsequently undergoing a progressive increase, during each upper respiratory ways infection. His past medical and family history was not remarkable.

There was no weight loss, obstructive symptoms and dysphagia were absent. The patient single complaints were fullness on the right side of the neck and some discomfort on moving his neck on right side.

On clinical examination, there was a unilateral, diffuse, soft, non-pulsatile, painless swelling on the posterior cervical triangle. The large tumoral mass was not attached to the overlying skin (Figure 1, a and b). The transillumination test was positive.

There were no palpable lymph nodes in the neck and no other clear swellings elsewhere.

The patient underwent a full set of laboratory tests, including a full blood count, C-reactive protein, and an autoantibody screen, the results being in normal limits.

The magnetic resonance imaging (MRI) scans showed the extent of the tumor (72/38/33 mm) and multiples cystic lesions with definite boundaries. It compresses the right

internal jugular vein without thrombosis. The MRI scan confirmed the non-invasive nature of the lesion, with only compressive properties, thus reinforcing the suspicion of lymphangioma.

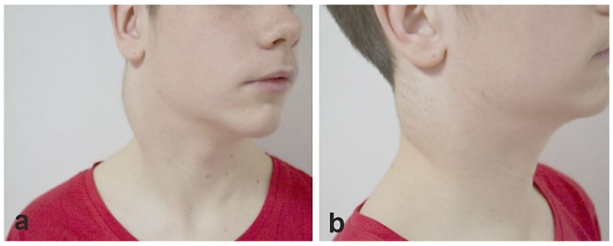

Figure 1 – ***Preoperative image of the laterocervical region of the patient, where we may observe the presence of a nodular, oval formation, localized behind and under the right sternocleidomastoid muscle, without affecting the upper skin layer: (a) Half-profile view; (b) Lateral view.***

In this situation, it was decided to perform a surgical intervention to remove the tumoral formation. Through a right cervical incision, under general anesthesia, the lesion was completely excised. The surgical sample measured 7/4/3 cm, appeared irregular in shape but was well circumscribed. Cut surface revealed multiple cysts like spaces separated by thin fibrous septa and filled with gelatinous material and few cystic spaces filled with blood.

The surgically removed piece was fixed in a 10% formalin solution with a neutral pH and sent to the Laboratory of Histopathology for a microscopic examination. After paraffin inclusion, there were performed 4 μm-thick serial sections, subsequently stained with Hematoxylin–Eosin (HE) and with Goldner–Szekely (GS) trichrome. For the immunohistochemical (IHC) study, we used the anti-D2-40 antibody (monoclonal mouse anti-human D2-40, clone D2-40, 1/100 dilution, Dako) for highlighting the cells of the lymphatic endothelium.

The macroscopic examination highlighted a well-limited nodular, elastic, soft structure, with a brown-yellow aspect, multicystic, with a "honeycomb" aspect at sectioning and with a gelatinous content. The microscopic examination highlighted a multicystic structure, with cysts of variable shape and size, completely separated or with fibrous conjunctive septa. The cystic spaces had a protein-like, mildly acidophilic content in the classical histopathological stainings.

The cyst walls had quite a varied thickness, from 5 μm to 50 μm, being structured from a fibroadipose conjunctive, well-vascularized, tissue, while in other areas there prevailed collagen fibers. In the adipose tissue, there were identified numerous small lymphatic ganglia, with a diameter of 1–4 mm, with minor reactive changes (clear germinative centers, moderate edema, and vascular congestion). All the cystic structures were lined up by a simple endothelium. The IHC examination showed that the endothelium was intensely positive to anti-D2-40 antibody (Figure 2, a–f).

Based on the clinical microscopic characteristics and the microscopic aspects, we set the diagnosis of CH.

The patient was hospitalized for nine days and was discharged in a satisfactory condition. Subsequently, no postoperative complications occurred.

The patient was symptom-free, with no recurrence during a four-year follow-up period (Figure 3, a and b).

Discussions

CHs are benign congenital malformations of the lymphatic system that manifest as macrocystic simple or multiple tumors, having a low communication with normal lymphatic vessels or venous structures. A very important percentage (about 90%) of hygromas is diagnosed in children less than two years old [11, 12].

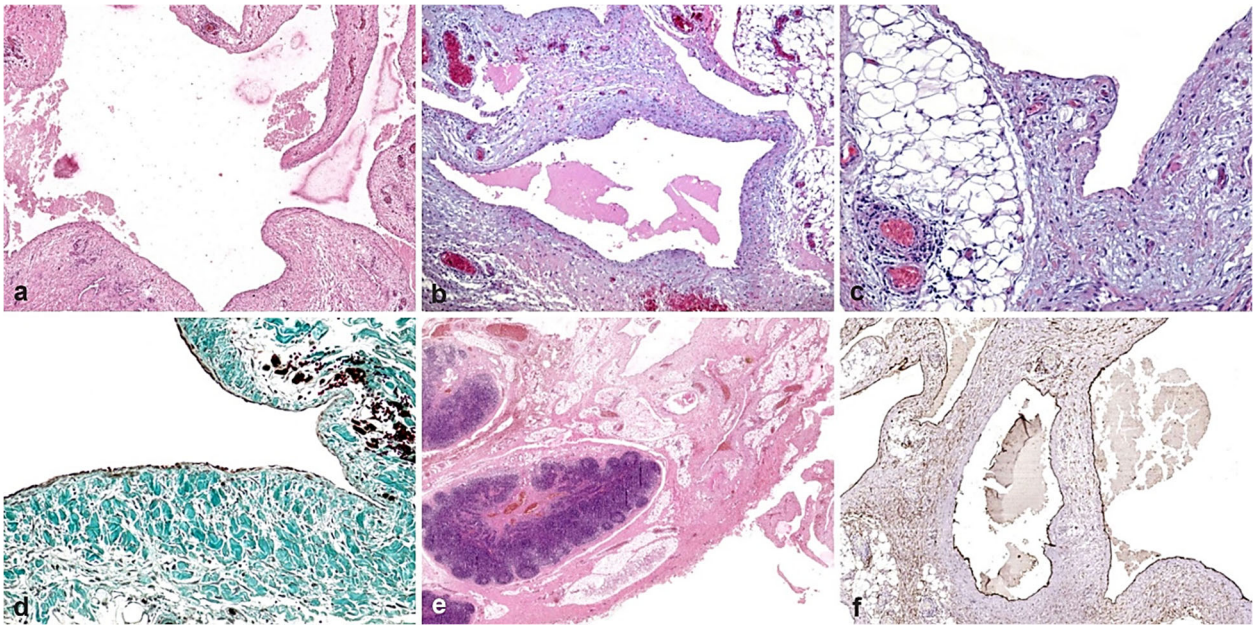

Figure 2 – ***(a) Microscopic overall aspect of the tumor highlighting numerous cysts of various shape and size, limited by multiple complete or incomplete conjunctive septa; (b) Cyst with a protein-like content, limited by thick conjunctive, well-vascularized septa; (c) Cystic wall mainly formed of adipose tissue, lined up by a thin endothelium; (d) Cystic wall mainly formed of collagen, limited by a simple endothelium; (e) Small-sized lymphoid follicles present in the cyst wall; (f) Overall image of cystic structures limited by the endothelium, positive to anti-D2-40 antibody. HE staining: (a, b and e) ×40; (c) ×100. GS trichrome staining: (d) ×100. Immunomarking with anti-D2-40 antibody: (f) ×40. GS: Goldner–Szekely; HE: Hematoxylin–Eosin.***

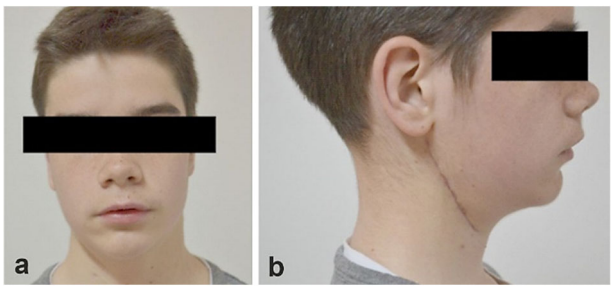

Figure 3 – ***Aspect of the right lateral region of the neck three months postoperative: (a) Front view; (b) Lateral view.***

The condition incidence is estimated at about one in 2000–6000 newborns. The symptoms are represented by pain and local swelling, especially during periods of regional infection and deformations of the anatomic structures [13, 14].

Our case presented various particularities regarding both the onset time and clinical symptoms and signs. The onset was late (after the age of 10 years old), with a slow progression, mostly associated with infectious episodes of the upper airways. Symptoms were low and were represented by the deformation of the cervical region and discomfort at neck rotation rightwards. Although there was a compression of the right jugular vein and the tumor reached a relatively high size (72/38 mm), the patient did not present any cardiovascular or neurological symptoms.

Up to now, there were reported numerous cases of CHs incorporated in the structure of other organs (larynx, trachea, esophagus, brachial plexus, and large vessels of the cervical region), associated with dysphagia, dyspnea, intense pain at neck movement [15–17]. Sometimes, hygromas may give major complications, such as infection, abscess, rupture, or hemorrhage.

As stated in many studies, most of the CHs develop in the cervical and facial regions (70–75%), axilla, mediastinum, groin, and more rarely in the liver, spleen, kidneys, intestines, mesentery, caul, adrenal glands, thoracic wall, limbs, etc. [18–20]. Until now, the exact pathogenesis of malformations in the lymphatic system is not completely known. There were proposed two hypotheses regarding the genesis of lymphatic malformations: an abnormal or insufficient network of the lymphatic system or a deficient connection between the lymphatic system and the venous system [21, 22]. The hypothesis of a deficient connection between the lymphatic system and the venous system is more probable for CHs of the head and neck, as the two vascular systems present some common stages and connections during the embryo development [22].

There are more proposals for classifying lymphatic malformations, based on the morphological aspects. A first classification divides lymphangiomas into microcystic, macrocystic and mixed. Microcystic malformations have a diameter less than 2 cm, mostly with an infiltrative aspect, they are more difficult to diagnose and do not present a good response to sclerosing therapy. In contrast, microcystic lymphatic malformations have a diameter over 2 cm, they are more easy to diagnose, they may be treated by surgical removal and have a better response to sclerosing therapy [23–26].

Nowadays, another classification was added to this widely used classification, according to the microscopic aspect: lymphangioma *simplex* (lymphangioma *circumscriptum*), characterized by the presence of thin-walled lymphatic vessels; cavernous lymphangioma, characterized by dilated lymphatic vessels; cystic lymphangioma (CH), with huge, macroscopic lymphatic spaces; and benign acquired progressive lymphangioma, in which the lymphatic channel dissects the dense collagen bundles [27].

At present, there are multiple treatment options: conservatory treatment by injecting sclerosing agents (Bleomycin, Rifampicin, corticosteroids, Dextrose, etc.), repeated content aspiration, radiotherapy, ablation by radiofrequency, surgical removal, etc. [25, 28]. When choosing the treatment method, there should be held in mind the lesion localization, the approach way, the patient's age, the presence of complications after treatment, etc. Also, there should be considered the multicystic nature of the lesion and the fact that it sometimes has a fragile, thin wall.

In the last years, intralesional injections with sclerosing agents showed good results. Of these, Bleomycin is more and more used. Bleomycin is an antineoplastic drug that may cause fibrosis and scaring of the tissues when injected in the cysts [29].

Our treatment option was surgical removal, due to the tumor localization and morphological structure. The results were favorable, as there were not recorded any immediate complications and the patient's monitoring for a period of four years showed no presence of local relapse.

Numerous studies showed that, in most cases of CHs, surgical removal remains the most common treatment method. It is indicated especially when there are complications of the cysts, such as recurrent infection, hemorrhage inside the cyst, dysphagia, compression of the airways, sudden growth of the cyst or deformation of the region where the cyst developed [25].

Conclusions

In our report, we presented a case of cervical CH in a 13-year-old boy, investigated by MRI, which was surgically treated. The objective in surgery of CH was preventing the obstruction of important anatomic structures, a favorable cosmetic result and lack of recurrences. The absence of immediate complications and local recurrence for four years after surgery show that we chose the best treatment method.

Conflict of interests

The authors declare that they have no conflict of interests.

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