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Cystic Hygroma - A Rare Appearance in Adulthood

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Abstract

Cystic hygroma is a rare benign tumor commonly located in the head and neck. It is usually seen in children under 2 years of age and extremely rare in adults. In adulthood it usually presents as an incidental finding often as a cervical lump, apparent due to acute infection. Why these lesions remain dormant is still unclear, though trauma or upper respiratory tract infections have been suggested as possible triggers for onset.

Case Report: We present the case of a 32 year-old female patient, with a cervical mass located in the posterior border of the sternocleidomastoid muscle, with a recent increase in volume in the past 10 days. CT scan revealed a multilobulated cystic mass, located deep behind the sternocleidomastoid muscle, posterior to the vascular nervous pedicle and partially filling the posterior cervical space. Fine needle aspiration was compatible with cystic hygroma. The mass was surgically excised and eighteen months later the patient has no evidence of recurrence.

Discussion: Complete surgical excision is the treatment of choice in symptomatic patients. But it should be kept in mind that there are important complications of surgical therapy in the head and neck region such as cranial nerve injury. Nonsurgical treatment modalities for cystic hygroma such as interferon alpha, laser therapy, and intralesional sclerosing agents administration have been tested with good results.

Conclusion: Total surgical excision is important for the prognosis, since cyst remnants usually cause tumor recurrence. New therapies are in ongoing studies with promising results.

Keywords

Cystic hygroma, Cervical mass, Lymphatic malformations

Introduction

Cystic hygroma (CH) is a rare benign vascular tumor caused by a congenital malformation of the lymphatic vessels. It is commonly located in the cervicofacial region [1], especially in the posterior cervical triangle, though it may be found anywhere else in the body. More than 60% of CH are present at

birth and up to 90% become overt before the age of two [2,3]. Presentation in adulthood is a rare incidental finding, usually as a cervical lump apparent due to acute infection or trauma. The etiopathogenesis is still unclear, though trauma has been suggested as one of the possible causes [4].

Case Report

A 32 year-old female presented with a painless right-sided cervical mass, with a sudden onset and progressive growth in the past ten days. The patient reported a preceding upper respiratory tract infection, denying recent cervical trauma.

On examination a soft, smooth, compressible, painless and delimited 6 × 4 cm mass posterior to the sternocleidomastoid muscle was noticed.

Blood work revealed aleukocytosis of $12.37x10^{9}$/L and elevated reactive C protein of 2.46 mg/dL.

Ultrasound and CT scan showed a multilobulated cystic mass with 6.5x4.5x2.5 cm, located deep behind the sternocleidomastoid muscle, posterior to the vascular nervous pedicle and partially filling the posterior cervical space (insinuating in the depth of the anterior border of the trapezius muscle) (Image 1- 6). Fine needle aspiration cytology revealed a clear yellow fluidwith numerous small lymphocytes. These findings were suggestive of CH.

The patient underwent surgical exploration of the neck through a cervicotomy, with an incision in the posterior border of the sternocleidomastoid muscle (Image 7).“En bloc” resection was achieved by blunt dissection with easy detachment of the mass. The patient was discharged 24 hours after surgery and the post-operative period was uneventful.

Histological examination revealed multiple cystic spaces with a single layer of flattened epithelial lining, confirming the diagnosis of CH.

There was no recurrence after 18 months follow-up.

Discussion

CH is a rare congenital malformation of the lymphatic vessels, most likely related to the failure of blind clusters of lymph sacs to join the lymphatic system during fetal development [5]. Approximately 65-75% of the tumors are present before the age of 1 and about 90% of the lesions occur prior to the end of the second year of life [6]. Occasionally they may be present as late as the fourth or sixth decade [7]. The reason these lesions remain dormant for such a period of time is unknown, but it is speculated that local infection or trauma may precipitate the growth of a previously unrecognized lesion.

In contrast to children where greater variability in location is seen [8], in the adult a neck mass is the presenting symptom in 97% of the cases [9] and the differential diagnosis includes neck abscesses, branchial cysts, lymphatic metastasis and other soft tissue tumors.

The correlation between imaging techniques and fine needle aspiration cytology, ensures an accurate diagnosis in the majority of cases. The location in the posterior cervical triangle associated with the presence of lymphocytes in the smear of the fine needle aspiration fluid are the major hallmarks [10].

CH can be differentiated from other types of lymphangiomas, such as capillary and cavernous lymphangiomas, according to different macroscopic characteristics and histological findings. CH also

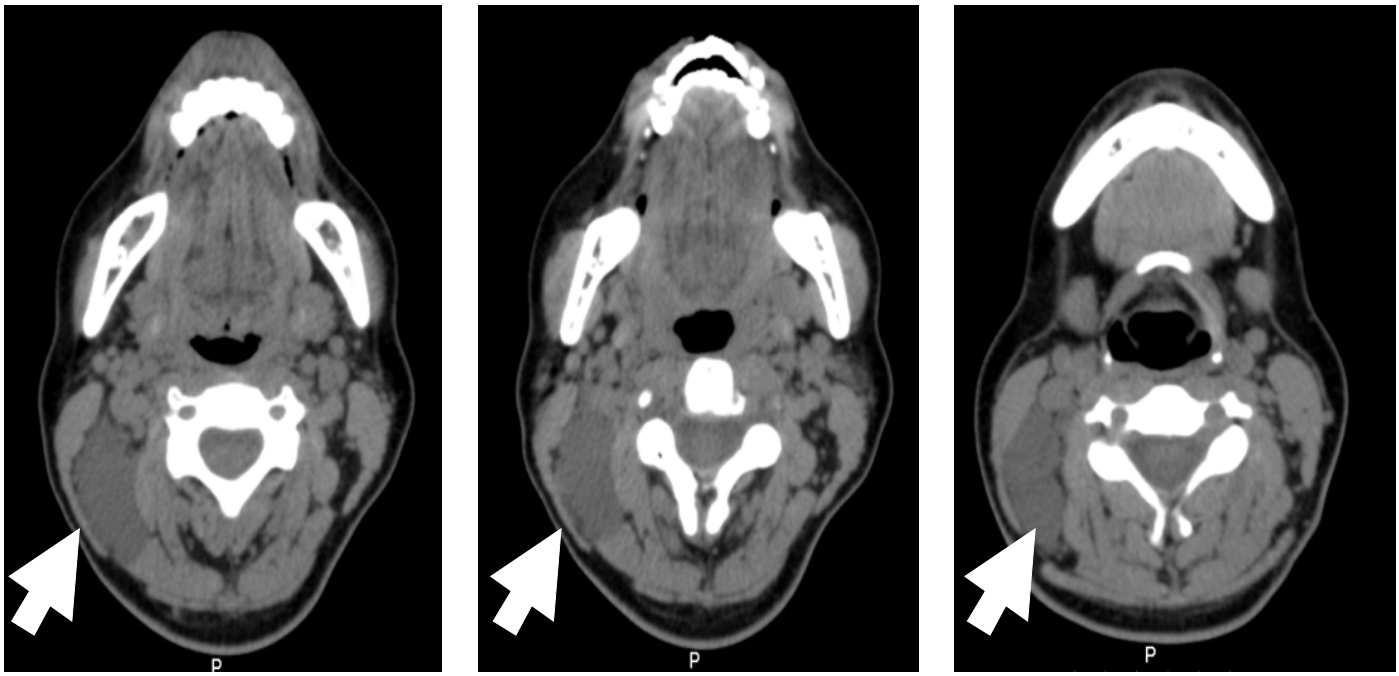
designated as cystic lymphangioma are cystic masses lined by a single layer of endothelium with a connective tissue stroma that grows and circumvents adjacent structures. Differently, capillary lymphangiomasarenon-invasive capillary-sized lymphatic channels and cavernous lymphangiomas are dilated lymphatic channels that in filtrate surrounding structures. [11]

Primary treatment for CH is total surgical excision [12]. However, CH´sinvasive characteristics and delicate walls can difficult "en bloc" resection especially in the head and neck region, where nervous and vascular structures can be easily injured. Care should also be taken to avoid leaving islands of tissue that can act as foci for recurrence, which has been reported to be as high as 20% [13]. Since recurrent lesions are more difficult to excise, several nonsurgical treatment options have been developed such as interferon alpha, laser therapy, and intralesional sclerosing agents [14]. Injection of sclerosing agents into the lesion such as OK432 (a lyophilized mixture of group A streptococcus pyogenes of human origin), 50% dextrose, triamcinalone, bleomycin, fibrin sealant and hydrocolloid impression material has had some success. Intravenous cyclophosphamide and intracystic injection of OK432 have been advocated [14].

Conclusion

CH are rather rare malformations in adults. However, they should be considered in the differential diagnosis of head and neck cystic masses in adults. Surgeons should ensure total resection of the tumor in order to avoid recurrence. New therapies in ongoing studies have shown promising results.

Figures



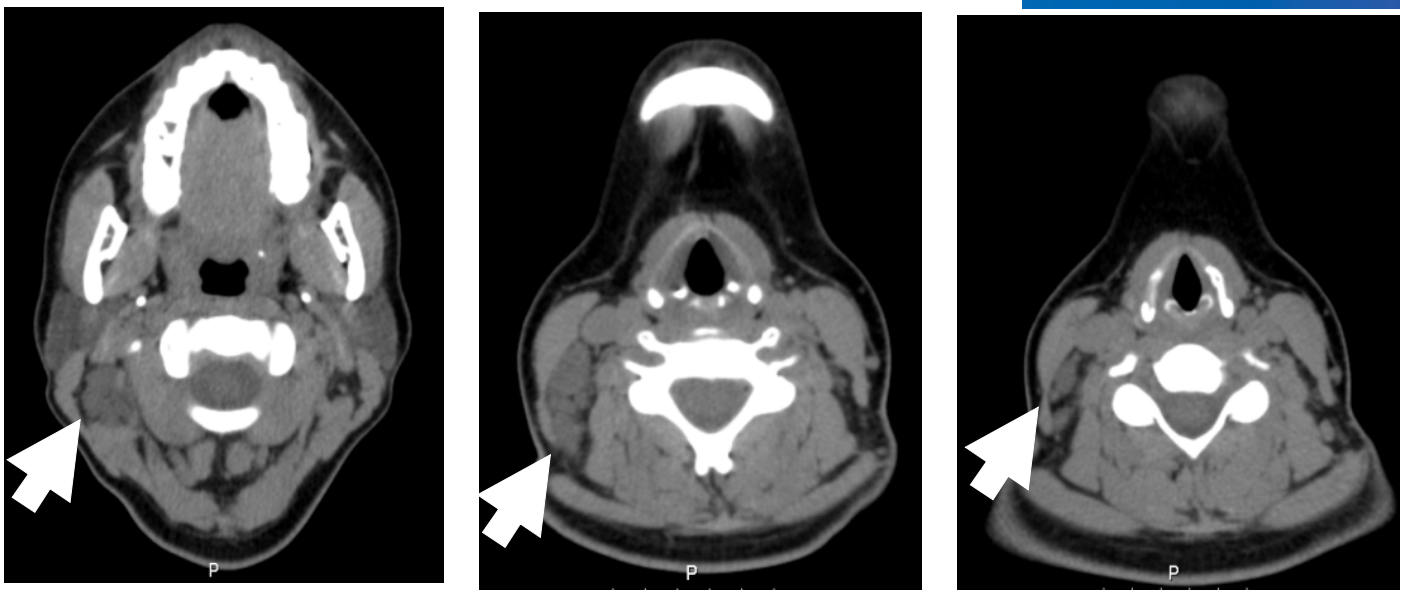


Figure 1-6: CT SCAN confirming a multilobulated cystic mass deep behind sternocleidomastoid muscle, posterior to the vascular nervous pedicle

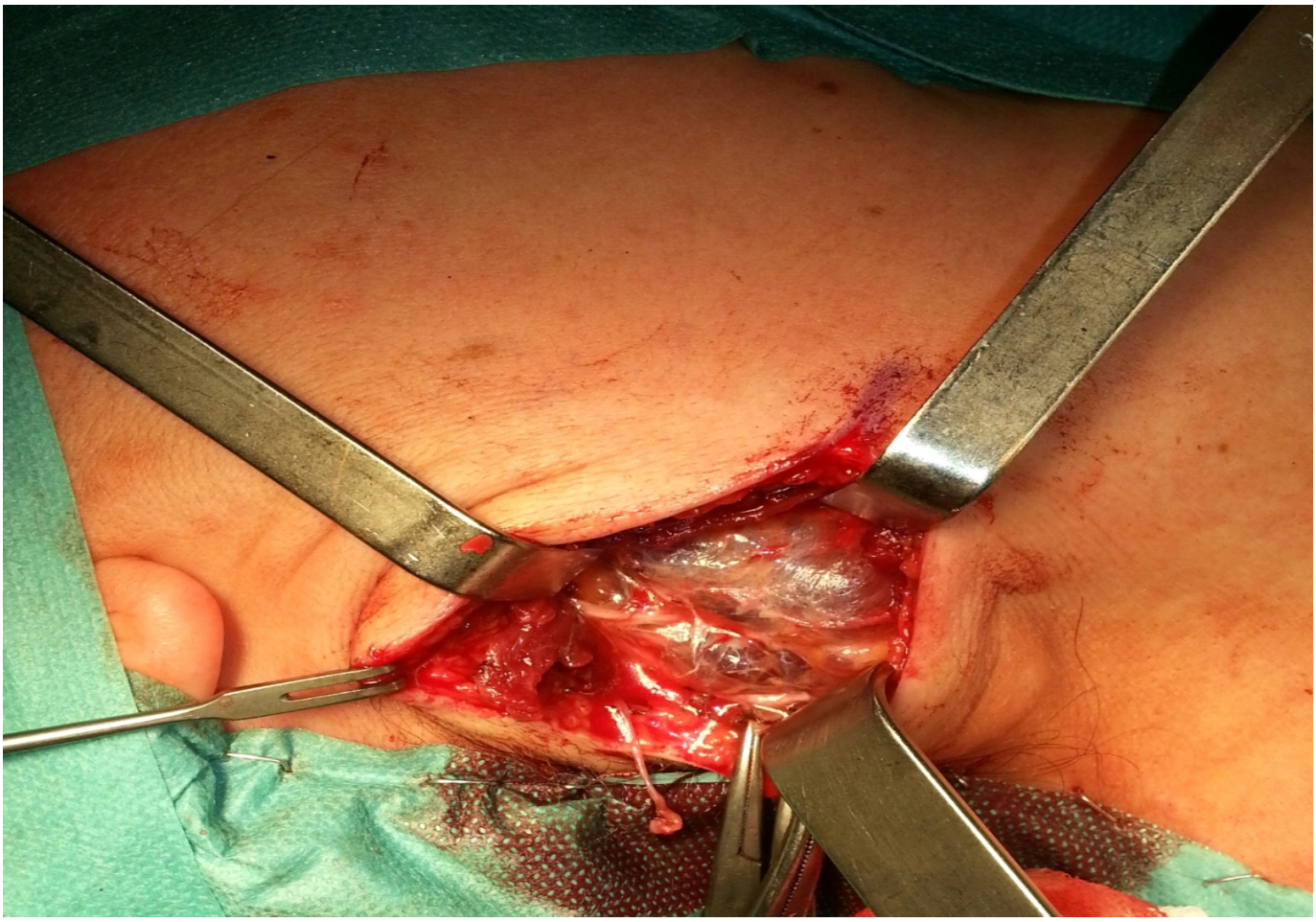


Figure 7: Surgical view with the characteristic multilobulated appearance of the cystic hygroma

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