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Case report

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Recurrent cystic lymphangioma of the neck. Case report

Cystic lymphangioma is a rare benign tumor commonly located in the head or neck. Approximately 100 adult cases have been reported in the literature. The etiopathogenesis is unclear, though trauma has been suggested as one of the possible causes.

CASE REPORT: *We report a case of recurrent cystic lymphangioma arising in an adult who had been successfully treated in our department, leading us to review the literature for a critical examination of our therapeutic choices and the pathological, clinical and therapeutic aspects. We performed a dissection of the cyst respecting anatomical structures.Three days after the surgery, the patient showed a recurrence under the previous site of the excision, We then decided to place a suction drainage under local anesthesia, The suction drainage was removed 25 days later.The post-operative course of the patient showed no signs of recurrence for twenty-four months.*

DISCUSSION: *Complete surgical excision is the treatment of choice in symptomatic patients. In our experience, postoperative suction drainage improved the outcome. Some authors have come out in favor of experimental, non-surgical methods, such as percutaneous sclerotherapy, which they claim offer the most promising results.*

CONCLUSIONS: *In our experience, totally excision of the mass was the treatment of choice but, placing just one suction drainage for weeks for new recurrence resulted in a surprising outcome. Infact, recurrence usually appears within the first nine months in about 10-15% of patients.*

KEY WORDS: Cystic lymphangioma, Disfigurement, Neck mass

Introduction

Cystic lymphangioma is a rare, benign tumor the pathogenesis of which is due to alterations, both congenital and acquired, of the lymphatic vessels. This disease appears within the first decade of life in 90% of cases, but can also rarely occur, apparently "*de novo*," in adults [1]. Cystic lymphangioma arises more frequently in the lateral neck region, where it appears as a slowly growing swelling, usually of soft, elastic consistency, which deforms and compresses the surrounding structures. Usually, patients show no hoarseness, dyspnea or dysphagia [2]. The main complications that are often associated with lymphangioma are infections and bleeding, which cause a rapid increase in size, with sudden compression of anatomical neurovascular structures and symptoms such as pain, dysphagia, dysphonia, and dyspnea, [2]. Although benign, lymphangiomas are often

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devoid of capsule and tend to infiltrate surrounding structures, making surgical excision difficult.[1]
The treatment of choice is radical surgical excision. Alternative techniques, such as needle-aspiration and percutaneous embolization with sclerosing agents, have been proposed, and have been indicated for cases in which radical surgical resection is impossible because the tumor invades important anatomical structures [3]. A case of recurrent cystic lymphangioma recently observed in our department led us to review the literature for a critical examination of our therapeutic choices and, more important, of the pathological, clinical and therapeutic aspects of this disease.

Case Report

In January 2010, a 50-year-old caucasian male presented to our department with the onset, for several months, of compressive symptoms, consisting of dysphagia for solid foods, and occasional wheezing, associated with the presence of massive swelling of the left lateral cervical area (about 13 cm in maximum diameter), which had arisen about 9 years before and had gradually increased in size. The patient reported that ten years previously he had undergone surgery excision of cystic lymphangioma. Three months after surgery the cystic lymphangioma reappeared, and was treated conservatively, four times, with fine-needle aspiration (FNA) of a large cyst. Regarding the etiology in our case, it seems that the patient reported a trauma in the left lateral cervical area of the neck about six months before the cystic lymphangioma appeared.
Clinical features: in our observation the patient showed a large mass about 13 cm in length in the left lateral neck area, covered with intact skin, and extending from the ear region to the ipsilateral clavicle, disfiguring the face (Fig. 1). It was not painful on acupressure, and was soft-elastic and deep. There were no signs of neurovascular distal damage. However, the patient did report paresthesia in the vicinity of the temporomandibular joint, conceivably a result of previous surgery.

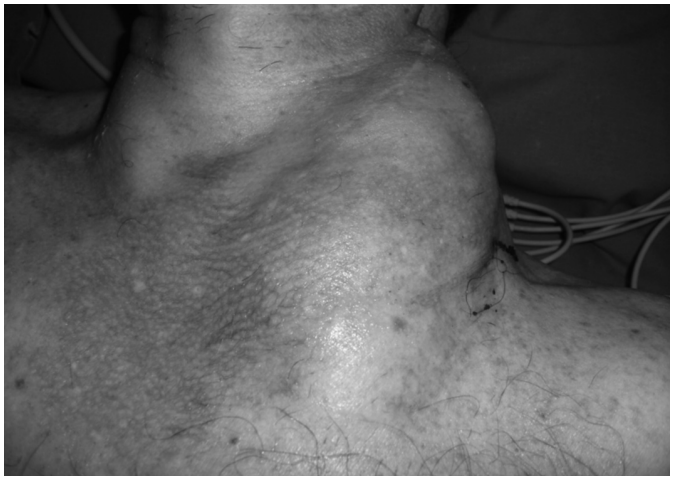

Fig. 1: Cystic limphangioma recurrent in the left lateral cervical area of the neck.

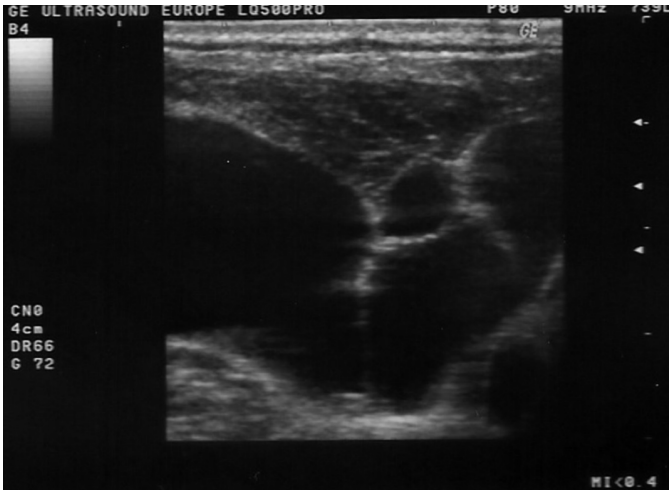

Fig. 2: Ultrasonography with a linear probe shows echogenic cyst with no flow in color-Doppler.

Diagnosis: after performing a routine serological test, which resulted normal, and a chest radiograph, we performed ultrasonography of the neck with a linear probe (MHZ) integrated with color Doppler (Fig. 2). It showed an echogenic cyst in the left cervical area that extended from the left ear region to the anterior mediastinum, involving the ipsilateral thyroid bed, and trachea, which was deviated contralaterally. The cyst was not vascularized; in fact, the color Doppler showed no flow inside the cyst.
However, if the ultrasonography allowed us to clarify the inherent characteristics of the new mass and, according to the literature [3, 4], should have allowed us to make a differential diagnosis with vascular malformations, it did not provide a clear correlation with deep structures. The neck ultrasound showed a multinodular goiter with dominant nodule which appeared hypervascularised at the color-Doppler. This nodule had been subjected to the fine needle aspiration twice with a indetermined result. In addition, the chest X-ray did not add any useful information, except for a slight inhomogeneity of the bronchial vascular pattern. The only way to clarify the correlation between the cyst and other anatomical structures was to perform a computed tomography (CT) scan of the neck and mediastinum with contrast enhancement (Fig. 3). The CT in the left cervical triangle showed a large, lobulated, soft-tissue lesion, with some internal septa containing fluid that was deep to the sternocleidomastoid muscle.
The mass was 11.4 cm x 7.9 cm x 13.2 cm in transverse, sagittal and coronal diameters, respectively.
It extended from C4 to D5, and was in the proximity of the neurovascular structures of the neck. It dislocat-

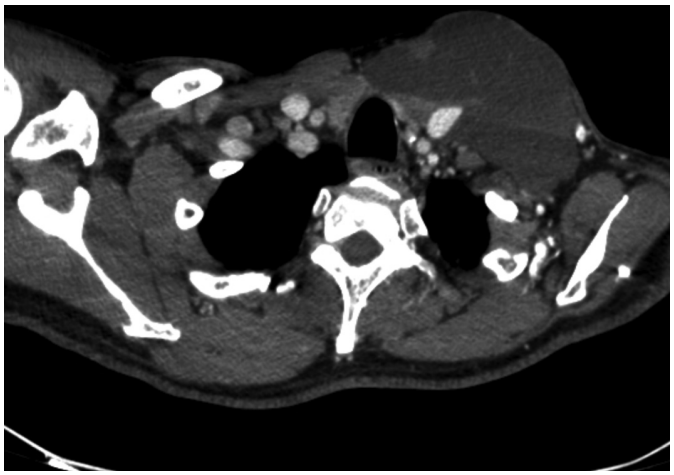

Fig. 3: The CT scan with contrast-enhancement shows a mass of 13 cm in coronal diameter in the cervical left area neighboring the sternocliei-domastoid muscle, pre-thyroid and omohyoid muscles, left thyroid gland, and jugular vein.

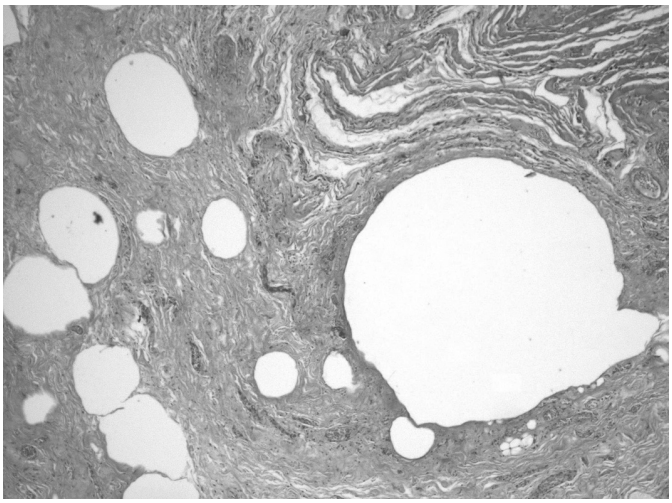

Fig. 5: The histologic section, stained in Emathoxilin-Eosin.

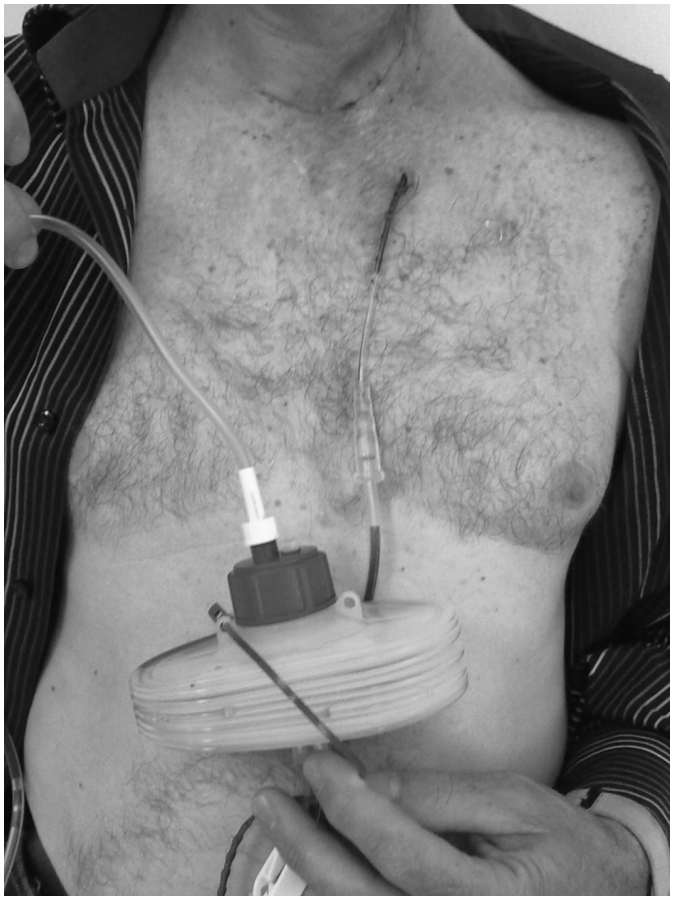

Fig. 4: Suction drainaige.

ed the internal jugular vein, which appeared curved, the external jugular vein laterally, and the sternocleidomastoid muscle medially.

Near the first cyst there were another two with the same characteristics, about 1.4 cm and 1.6 cm in diameter, respectively. Considering all these findings the patient was scheduled for surgical excision.

Surgical treatment: considering the presence of a voluminous multinodular goiter, the result of the ultrasound and indefinite cytology led us to also perform the thyroidectomy. A skin incision was made directly under the previous scar (which was eliminated) from the mandibular angle along the anterior margin of the sternocleidomastoid muscle, 2 cm. above the sternal notch and we extended the scar for 4 cm transversally in the neck.

Under the platysma muscle there was an encapsulated voluminous cyst. Extended to the clavicular region, sternum, anterior and superior mediastinum, it adhered to such surface structures as the pre-thyroid and omohyoid muscles, left lobe of thyroid gland, and jugular vein, so we proceeded to isolate the phrenic nerve, spinal accessory nerve and cervical plexus and perform a total excision of the cyst with total thyroidectomy. The cyst was strictly adherent to the jugular vein and carotid artery; medially contracted adhesion with subclavear vein; there were no relations with the thoracic duct. So we performed a meticulous dissection of the cyst respecting anatomical structures and we performed a total thyroidectomy. A TachoSil patch was applied to improve the hemostasis. Two suction drainages were placed, one in the thyroid bed, and the other in the left cervical area.

Following the treatment, the patient was monitored in the hospital for 24 hours.

Three days after the surgery, the patient showed a recurrence under the previous site of the excision, so we opted for two FNAs of the recurrent lesion. Four days after the FNA, there was recurrence once again without any clinical symptoms, but with important disfigurement of the neck. We then decided to place a suction drainage under local anesthesia, which drained about 50 ml a day of light fluid (Fig. 4). The suction drainage was removed 25 days later.

The post-operative course of the patient showed no signs of recurrence for fifteen months.

Histopathology: this revealed the presence of some large cystic spaces, and smaller channels covered by lymphatic cells. The histological section, stained in Emathoxilin-Eosin, showed fibro-adipous tissue with ectasic blood vessels and hemorrages. In the middle, some cystic formation, sourrounded by a monostratified flat epithelium lacking of atypia are sourronded by lymphoid tissue (Fig. 5). These features prompted the diagnosis of a cystic lymphangioma. A histological examination of the thyroid showed a multi-nodular goiter with follicular adenoma.

Discussion

Cystic lymphangioma is an uncommon benign tumor of lymphatic origin. It is a rare congenital abnormality of the lymphatic system that typically occurs in the pediatric age, usually observed in children within the first two years of life. It is caused by anomalies in the embryonic development of the lymphatic system, and typically arises at birth (approximately 90%). The other manifestations are in adolescence [3-11].

Cystic lymphangioma, also called cystic hygroma, occurs in 1/100,000 inhabitants [12]; spontaneous regression occurs in only about 6% of lymphangiomas [4,10].

Lymphangioma is most commonly located in the head or neck (75%) [5,7-9]; in 70% of cases it does not present as an extension to the oropharynx or mediastinum, and complete surgical resection is usually easy. Extension to the oropharynx occurs in about 20% of cases. Extension to the mediastinum is found in about 10% of cases. Respiratory distress is rare, but dangerous, and large surgical resection is necessary [8,10,11]. Less usual localizations include the mediastinum, scrotal area and retroperitoneum [8].

Approximately 100 adult cases have been reported in the literature [5]. No differences in terms of gender have been observed [8]. In their review [14], Ricciardelli and Richardson explain three indicators for severity and extent of disease: symptoms, size and growth pattern of the lesion, and the anatomical regions involved. They also show that these factors influence clinical outcome of the disease [13].

The etiopathogenesis of lymphangioma is unclear [5,8,9], though various studies have speculated that it results from inadequate drainage of the lymphatic vessel into the venous system, due to atresia or insufficiency of efferent channels [3,5,6,8].

So, obstructive or proliferative disorders of lymphatic embryogenesis are suspected as resulting in this type of tumor. The lymphatic system in human embryos has been described as an outgrowth of the venous system [5]. When lymphangiomas appears after the 30th week of gestation it is usually an isolated malformation, as when discovered during infancy or childhood [10]. Trauma has been forwarded as a possible etiology in the development of such lesions in adults. In fact, it has been suggested that local trauma can be a casual factor or trigger event in the development of these tumors in adults [3,5]. In our experience the trauma seems to be the triggering event, in fact the patient had a trauma in left area of the neck before the appearance of the cystic lympangioma. The etiology of lymphangioma in adults is likely due to delayed proliferation of residual cell. Wiggs describes how this proliferation occurs in older patients because of an instigating stimulus such as infection or trauma [6,9]. In this situation, the lymphatic channels invade the adjacent tissues and destroy the normal plane of dissection [9]. The lymphangiomas can be unilocular or multilocular, and usually contain serous or chylous fluid. They are often classified based on the gross appearance of the abnormal lymphatic tissue as capillary, cavernous or cystic [3,4,7,8,11,14]. Frequently, different histological varieties of this tumor coexist within one mass. [3,11]

A histological examination of the cystic lesion reveals a network of spaces varying in size, with some large cystic spaces and smaller channels. The spaces are lined by bland and flattened cells. These features are indicated in the diagnosis of a cystic lymphangioma [6].

The clinical significance of this entity depends on the size and localization of the tumor. In fact, in small lymphangiomas, without functional impairment or aesthetic disfigurement, therapeutic intervention is not necessary [3]. Although lymphangiomas are benign tumors with no malignant potential, they usually increase in size, progress and relapse (especially after prior incomplete excision), or are complicated by infections. The treatment of choice, therefore, is complete surgical excision, though according to the literature this should always be conservative, because such a benign condition does not warrant resection of vital structures.

Given this conservative approach, surgery is often incomplete, and the relapse rate is, therefore, high [8]. In fact, as already mentioned, this type of tumor is usually slow growing, and rarely regresses spontaneously, though it can sometimes expand rapidly, especially in cases of bleeding or infection. In cervical locations, it can cause compression disorders, such as respiratory distress and dysphagia. It can also induce skeletal deviations or bony erosion, aural or ocular involvement, and aesthetic disfigurement, all without functional impairment [3,8].

Ultrasound diagnosis, CT and magnetic resonance imaging (MRI) are the most accurate methods for diagnoses of lymphangiomas. Echotomography with color Doppler ultrasound can distinguish lymphangiomas from vascular lesions [3,4], thyroid nodules, lipomas, desmoid tumors, brachial cysts, and sebaceous carotid glomus tumors [2].

The treatment of lymphangiomas depends on surgical experience. In a recent review of pediatric lymphangiomas, Orvidans and Kasperbauer recommended meticulous surgical excision as the primary approach to treatment. They say that infection of the lymphangioma can

lead to total regression of the lesion, so they propose using injection of sclerosing substances into the cyst [11]. Indeed, approaches such as the use of diathermy [11], radiotherapy, percutaneous sclerotherapy [8,11], and FNA [14] have been tried to treat these lesions. Among these, sclerotherapy has given the best results, but multiple sclerosing substances have been used, and there is some debate over what the agent of choice should be [8]. Various sclerotherapeutic agents have been shown to have minimal effects on lymphangiomas [11]. Among the sclerosing substances, there are descriptions of OK-432 (picibanil), bleomycin and corticosteroids [3,4,14]. The infiltration of these sclerosing substances may help avoid surgery, according to some reports [6]. The sclerosant could involve the destruction of the wall of the cystic spaces, with a resulting decrease in fluid production, and collapse of the lesion [4,11].

OK-432 (picibanil) is the first sclerosing substance described in the literature, and as such is a new and promising agent for sclerotherapy. OK-432 is produced by incubating a culture of low virulence, SU strain of type III, group A Streptococcus pyogenes of human origin [11], treated with benzylpenicillin. When OK-432 is injected into the cystic spaces it produces a sclerosis that does not spread outside the lesion. The first results of intralesional injecton of OK-432 as a treatment of lymphagioma were reported in 1987; until then this substance was used for the immunosuppressive treatment of malignant tumors. Since then, reports have been published on the use of picibanil in the treatment of lymphangiomas, with good results. There have been no reports on scarring of the surrounding tissue and structures, or disturbance of function [4,11].

A number of study [6,11] are in favor of conservative treatments, such as laser therapy, administration of interferon alpha, and various sclerosing agents, such as steroids, hypertonic saline, ethanol, bleomycin, though some of these substances have proved to be unsuccessful. Moreover, an intracystic injection of sclerosing substances produces an inflammatory reaction, leading to the destruction of the epithelial lining, and subsequent scarring (sclerosis) of the lesion [11]. These agents may spread outside the thin-walled lesion and cause damage to the surrounding structures, making subsequent surgery difficult and leading to extensive scarring. Bleomycin is known to cause pulmonary fibrosis, though this is rare [4,11]. The use of alcohol has led to mediocre results, with subsequent scarring problems. Recently, good results have been reported on the use of Tissucol (fibrin sealant) [3,11] These modalities have been found to be ineffective, and associated with severe complications by some [3,4,] who consider total surgical removal of the lesion to be the treatment of choice. However, a more conservative surgical approach is advised by others. Most autors agree that surgery should never be excessive, and vital structures must never be sacrificed [3,9,10]. Another important consideration is recurrence. Though it seems there is complete resection in 80% of cases, recurrence is observed in approximately one of five cases [10]. Lesions involving the lip, hypopharynx, larynx, tongue and floor of the mouth have high rates of recurrence after surgery, and persistence of disease is much higher. So as a first approach, complete and meticulous excision is recommended [11]. Therefore, it seems appropriate to perform the surgical excision when the lesion is at an early stage, because the treatment often results in failure. However, complete removal of cervical cystic lymphangiomas is difficult because they have a thin wall of endothelium, which can easily be torn [14]. We esteem that the early recurrence we observed could depend on the incomplete removal of thin walls of cystic mass, difficult to identify. Subsequently, a postoperative lymph leak may have given. In this situation, we believe that the suction drainage, placed for a long time (about 3-4 weeks), may be useful in the treatment of nearly recurrences because of the obliterating lymphatic ducts and consecutively sclerosis. The suction drainage may be useful in the treatment of nearly recurrences probably due to persistence of the residual wall that are difficult to completely remove in case of recurrence.

Conclusions

Cystic lymphangioma is a benign tumor whose manifestation in adults is rare, but when it occurs, it manifests in more than 97% of cases as an enlarging neck mass, while in children a greater variability is seen [5]. According to one study [3], in small lymphangiomas without functional impairment or aesthetic disfigurement, therapeutic intervention is not necessary. With large tumors that interfere with deglutition, respiration, vision, hearing, regular posture of the head and neck, bone development, or aesthetic appearance, therapeutic intervention is obligatory. Our case was particularly interesting. It had a localization in mediastinum, which is a rare event (only 10% of cases), [10] and had also relapsed five times. In fact, the patient had undergone surgical excision approximately 10 years before, and was then treated conservatively with FNA biopsy four times. In some reported cases in adults, the triggering event was an injury or minimal cervical trauma occurring accidentally [3,5]. As suggested by the literature [3,5], trauma can lead to lymphangioma, and our patient had reported a local trauma in the left region of the neck one month before the first surgery. Complete surgical excision is considered the treatment of choice in symptomatic patients, either for curative or stabilization purposes [5,8]. It should not be forgotten that lymphangioma is a benign condition and treatment should, therefore, spare vital structures at all times. Some studies have come out in favor of experimental, non-surgical methods, such as percutaneous sclerotherapy, which they claim seem to show the most promising results [8]. In our experience, total exci-

sion of the mass was the treatment of choice but, placing just one suction drainage for weeks for new recurrence resulted in a surprising outcome. The suction drainage was placed in the neck of the patient for twenty-five days and, perhaps, with continuous suction of the fluid, the upper tissue adhered to the lower tissue, thus obstructing the formation of a cavity for another cyst. That drainage solved the problem of recurrent cystic lymphangioma is hypothetical, though the patient stills hows no signs of recurrence, while recurrence usually appears within the first nine months in about 10-15% of patients [9].

Riassunto

Il linfangioma cistico è una rara neoplasia benigna comunemente localizzata nella regione della testa e del collo. Approssimativamente 100 segnalazioni riferite a pazienti adulti, sono state riportate in letteratura. L'etiopatogenesi della malattia è poco chiara sebbene il trauma sia stato suggerito come una delle possibili cause.
Riportiamo un caso di linfangioma cistico recidivo insorto in un soggetto adulto trattato con successo nella nostra unità operativa. Questo caso ci ha condotto ad una revisione della letteratura per un riesame critico delle nostre scelte terapeutiche e per una messa a fuoco degli aspetti anatomo-patologici, clinici e di trattamento. Nel nostro caso, è stata effettuata un'accurata escissione della massa nel rispetto delle strutture anatomiche adiacenti. Tre giorni dopo l'asportazione, il paziente mostrò una recidiva in corrispondenza del sito chirurgico. Fu quindi deciso di inserire, in anestesia locale, un drenaggio in aspirazione. Il drenaggio venne rimosso in venticinquesima giornata. Il decorso post-operatorio non ha dimostrato segni di recidiva a 24 mesi dall'intervento,
L'escissione completa può essere pertanto considerata il trattamento di scelta nei pazienti sintomatici, completato, come segnalato nella nostra esperienza, da un drenaggio in aspirazione che ha migliorato il risultato finale. Alcuni autori segnalano risultati favorevoli di metodiche di trattamento non chirurgico, come la scleroterapia percutanea, che, nella loro esperienza, offre i risultati più promettenti.
Nel caso da noi osservato l'escissione completa della massa è stato il trattamento di scelta, ma l'impiego del drenaggio in aspirazione ha determinato un risultato positivo sorprendentemente durevole, dato che le eventuali recidive, segnalate nel 10-15% dei pazienti, sono generalmente precoci e si verificano generalmente entro i 9 mesi dal primo intervento

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