CASE REPORT

Non-surgical treatment of a relapsed cystic hygroma in an adult

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SUMMARY

Lymphatic malformations, also known as lymphangiomas or cystic hygromas, are benign masses that typically affect newborns and infants and involve the head and neck regions. They are, however, rare in adults and even rarer in the axillary region. Although surgery is considered to be the treatment of choice, we present a rare case of a recurrent cystic hygroma 32 years after the first surgical operation. Due to the cosmetic concerns and the risks of a surgical approach, non-surgical therapy with percutaneous sclerosants was performed, with a good outcome after a 2-year follow-up period.

BACKGROUND

Lymphatic malformations are rare benign tumours caused by abnormal growth of lymphatic vessels and are typically diagnosed in newborns and infants. They occur in the head and neck regions, but also in the mediastinum and axilla. Since they are thought to be a failure in the development of lymphatic tissue, they usually occur during childhood, and their diagnosis is very rare in adults. The old terminology, such as lymphocele, lymphangioma and cystic hygroma, is still used today. Historically, the treatment of choice was surgical excision. Although long-term follow-up is recommended, the rate of recurrence, even on incomplete surgical removal, is very low. However, some authors highlight the importance of non-surgical management, especially by means of percutaneous sclerosants.¹

CASE PRESENTATION

A 33-year-old woman was admitted to our hospital presenting with a lump in the left supraclavicular region and left armpit, which had been growing over the last week. She also had swelling of the left arm. She had no fever, cramps, numbness, tingling, weakness or changes in arm temperature. On examination, the mass was painless and non-tender.

At the age of 8 months, the patient had a lymphangioma in the left armpit, which was surgically removed. Over the course of follow-up throughout her childhood, no evidence of recurrence of the mass was found.

INVESTIGATIONS

Routine blood tests were normal, and an ultrasound examination revealed a multilocular cystic mass located in the left supraclavicular region. Vessels were surrounded by the mass, but were not compressed by the lesion. A chest CT scan showed

a polylobulated, multicystic mass of fluid density, along the supraclavicular and axillary regions, measuring 9×3.5×5 cm (figure 1). This cystic, non-invasive mass was surrounding the subclavian artery and vein, the lumina of which were normal. Thrombosis, mechanical compression and infiltration were ruled out. An MRI was performed to delineate the lesion and rule out involvement of critical structures, nerves and vessels (figure 2), in case of further surgical procedures. Several days later, the cytological samples were reported as isolated lymphocytes within a proteinaceous background, which might prove its lymphatic origin. Pathologists reported the samples as benign cystic lesion.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses of a supraclavicular mass should include haematoma, abscesses, lipoma, neurofibroma and several types of soft-tissue sarcomas. Loculation and cyst formation within the mass can be helpful in assessing the lesion. Given the patient's surgical history and the radiological features of the lesion, lymphangioma was strongly suspected as the first choice. However, this entity is extremely rare in adults, so other choices should also be taken into account, especially to rule out enlarged lymph nodes or malignant masses.

TREATMENT, OUTCOME AND FOLLOW-UP

The patient was then referred to our vascular surgery department, where surgeons explained the treatment options. Although surgical therapy was preferred by the surgeons, the patient chose a non-surgical treatment with percutaneous sclerosants.

Therefore, by using ultrasound guidance and a syringe, the cystic fluids were aspirated from the supraclavicular area and bleomycin was injected intralesionally (15 mg, dissolved in 10 mL of sodium chloride for injection, 0.9%) in the supraclavicular area. Only 10 mL were aspirated and the tip of the needle was maintained within the cyst lumen for the bleomycin injection. The patient remained under observation for 24 hours and was discharged afterwards. She did not report any side effects. One month later, she was called for follow-up. The swelling had disappeared and on physical examination, the mass in the supraclavicular region was hard to feel. A chest MRI was performed to assess the extent of the mass (figure 2) 6 months after the discharge. The mass had not completely

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Figure 1 Axial plane of a chest CT showing the mass near the left axilla and the anterior chest wall, encircling the left axillary artery (white arrow).

disappeared, but as the patient was asymptomatic, we decided to carry out periodic follow-up. The patient also preferred this option. This follow-up has been done every 6 months over the past 2 years. The patient has remained completely asymptomatic and she leads a normal, active life. To date, no additional therapies have been required.

DISCUSSION

Lymphangiomas, or cystic hygromas, are lymphatic malformations affecting newborns and children, found in the head and neck regions, and less frequently in the chest, axilla and mediastinum. Approximately 90% of these benign tumours are diagnosed in newborns and infants before the age of two.² Lymphangiomas are rare, congenital benign tumours, which are probably due to a failure of the developing lymphatic tissue.³ When removed, lymphangiomas are unilocular or multilocular cysts containing yellowish or milky fluid. Lymphatic malformations

are typically classified into several subtypes, according to the size of the lymphatic channels and the size of the cysts (microcystic, macrocystic, mixed). As such, cystic hygroma, cavernous lymphangioma and cystic lymphangioma are sometimes used interchangeably.³

Patients with lymphangiomas have enlarged masses in the head, neck, anterior chest wall or armpit, usually with cosmetic deformity. These benign masses can sometimes compress critical structures such as vessels, nerves or the airway.³

When imaging studies are performed, lymphangiomas are seen as large, cystic masses. Ultrasound is used as the first diagnostic workup and can classify the subtype of lymphangioma and assess the size of the cysts. CT and MRI can assess the features of the mass and the involvement of critical, vital structures, the extent of the tumour,⁴ and can yield an accurate preoperative assessment.

However, lymphatic malformations in adults are rare. Several case reports have been published regarding adult-onset lymphangiomas. Naidu³ published a case-study paper with 91 patients along the English literature from 1913 to 2000, with lymphatic malformations in the head and neck regions. Later, Huang⁵ reported a case study of four adult patients with axillary cystic hygroma, who were treated with surgical excision. Two recent case reports described adults with spontaneous large lymphangiomas in the chest wall and axilla,^{6,7} both of which were surgically removed.

Recurrence after surgical excision is also rare.⁸ However, our patient was presenting with a recurrence more than 30 years after the removal. The mass became apparent when the patient noticed an enlarged lesion in the supraclavicular region, that is, when the tumour became symptomatic. We cannot consider this as a case of spontaneous adult onset, but rather a late recurrence of a previous disease, since the patient had a history of a cystic hygroma during childhood.

The patient was very concerned about the cosmetic outcome and the risks of surgery, and surgeons therefore offered her a non-surgical approach. The traditional treatment of these malformations is the surgical removal of the lymphatic tissue. However, when critical structures are involved, such as nerves or vessels, complete excision is not possible as this may increase the risk of recurrence. Several non-surgical approaches have therefore been proposed.¹ The use of percutaneous sclerotherapy with drugs such as OK-432, Ethibloc or bleomycin may yield promising outcomes, with a high level of effectiveness. It involves a sclerosant drug that irritates the endothelial cells of the lymphatic malformation, causing a non-specific inflammatory reaction.⁴

Bleomycin is an antineoplastic drug that can cause fibrosis and scarring when instilled into the cysts. This drug therapy can cause reduction in the size of the cyst. When the cysts have a partial response, the procedure can be repeated. The maximum cumulative dose allowed is 5 mg/kg of body weight.¹ Our patient was given 15 mg in a single dose. A reduction in size may facilitate a further surgical approach, if required. According to some case studies,^{1,4,9} an excellent or good response can be achieved in almost 95% of patients treated with sclerosants.

Regarding the risk of pulmonary toxicity, it is worth mentioning that sclerosis of the cysts is achieved by local action of bleomycin, of which a small dose is injected. However, when used for oncology purposes, bleomycin is given intravenously at higher doses (exceeding 400 mg). No pulmonary toxicity has been reported as a complication of intralesional bleomycin. The immediate complications include local swelling, erythema and fever.

Figure 2 Slides (A) and (B) show coronal and axial planes of fat-suppressed inversion recovery sequences; the white arrow is pointing to the lesion. Slides (C) and (D) show how the reduction in size was achieved after sclerotherapy; the MRI was performed 2 months after the injection of the sclerosant drug.

Patient's perspective

I cannot remember the surgical intervention, as I was an infant when it happened. My parents were very afraid as I was too young to have a tumour, even if it was benign. The outcome of the operation was good, and I have been healthy since then, except for a little white scar in my left armpit. When a lump began to grow in the lower region of my neck, I was afraid it was an enlarged lymph node or some kind of malignant condition. Fortunately, it was a benign mass, apparently a recurrence of the forgotten disease I had when I was a child. Surgeons offered me the option of undergoing aggressive surgery, but I was very reluctant because of the aesthetic consequences. Nevertheless, they offered me an alternative therapy, in the form of percutaneous sclerotherapy. It has been 2 years since this non-surgical procedure, and although the mass has not completely disappeared, I have no symptoms at all. I can freely move my left arm, I do not have any pain, and most importantly, I do not have any scars. As a young woman, it would be a terrible blow to have ugly scars. I am so grateful to my surgeons.

Learning points

- ▶ Lymphatic malformations are rare diagnoses in adults, and a strict differential diagnosis workup regarding a cystic mass in the head and neck should therefore be performed in order to rule out malignant conditions, enlarged lymph nodes or abscesses.
- ▶ Due to cosmetic concerns or high surgical risk, percutaneous sclerotherapy may be an appropriate therapy for this kind of lymphatic malformation. Intralesional therapy is both safe and effective for the treatment of lymphatic malformations, and has a high success rate. Some authors consider sclerotherapy to be the first-line treatment approach, rather than surgical excision.
- ▶ Bleomycin dose used as a sclerosant is much lower than the doses given for oncology purposes. This drug can prevent a surgical resection and, although the response may be partial, indicating persistent or recurrent disease, the mass may improve, patients may remain asymptomatic and no further therapy would be considered.

In our patient, bleomycin was only instilled once, and although the lesion remained in the subsequent follow-up, she was completely asymptomatic, as the mass had become smaller. We therefore consider that the success of percutaneous sclerotherapy was due to a sufficient reduction in the mass, allowing the patient to remain asymptomatic.

To our knowledge, this is the first reported case of a recurrence 32 years after the excision, and although a full reduction was not achieved, the patient remained asymptomatic, and no further surgical treatment or sclerotherapy was considered over a 2-year follow-up period. This case may contribute to the knowledge of the role of lymphatic malformations in adults and the role of bleomycin in the treatment of these lesions.

Contributors RGC drafted and reviewed the manuscript. Images were edited using The GIMP. BR-M and OV-G made interesting suggestions and helped in the translation of the manuscript into English.

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