Recurrent lymphangioma: What are the treatment options?

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ABSTRACT

This is a report of a rerecurrence of extensive lymphangioma involving the neck, chest and both breasts of a female.

The patient has had recurrence of the lesion twice following surgical excision. Surgery alone seemed to be insufficient in this case. Could adjunct of sclerotherapy have made a difference at the first surgery? Faced with a third recurrence, what other options are left? We report this case to highlight the difficulties and dilemma in managing extensive lymphangiomas.

Key words: Lymphangioma, management, recurrent

INTRODUCTION

Lymphangiomas are among the surgically treatable diseases of the lymphatic vessels. Extensive involvement of the body poses a major challenge to treatment and recurrence is a known and worrisome complication. Presently, surgical excision is the major treatment modality for lymphangiomas. Sclerotherapy with the aim of obliterating the lymphatic channels is also being increasingly used.[1,2] However, reports of recurrences and morbidity suggest that the treatment options currently accepted for this condition may not be adequate.[3] We present this case to highlight the difficulties in managing extensive and recurrent lymphangiomas.

CASE REPORT

A 20-year-old female student nurse presented to us with a 6-year history of progressive swelling in the breast, chest and neck. The breasts became massively distended (more than double the size) when she sat up and became smaller when she lay down. There were no other associated symptoms and no similar swellings in any other part of the body. Despite treatment from many unorthodox and religious healers, there was no success. Embarrassed because of the huge breasts, she threatened to abandon school.

She had been diagnosed as having cystic hygroma of the neck at birth, for which she was operated on and had the swelling removed in a teaching hospital. A swelling reappeared on the chest at 8 years of age, which necessitated a second surgical excision at the same institution. The last swelling recurred at 14 years of age (6 years after the second surgery) and was much more extensive than the previous ones.

She attained menarche at 14 years and had been having regular 28–30 day menstrual cycles. She had never been pregnant before and had never been on hormonal drugs.

Examination revealed a well-nourished young girl in no obvious distress. Her weight was 48 kg and height was 1.4 m. The most significant finding was a diffuse cystic swelling with an emptying sign, which spanned the anterior chest wall, involving both breasts and extending into the anterior neck up to the angle of the jaw. Apart from two transverse scars noted on the swelling, the skin looked normal. The swelling was just below the skin. The breasts became markedly distended (left > right) and the neck and the upper chest swellings disappeared when the patient sat up [Figure 1]. Conversely, the breasts shrunk remarkably while the upper chest and neck swellings became worse when she lay down [Figure 2]. The nipples appeared thinned and stretched, the breasts measured about 20 x 20 cm (right) and 25 x 30 cm (left) in the sitting position and felt cystic and transilluminated.

The full-blood count, serum electrolyte/urea and creatinine, X-ray of the chest and neck and urinalysis results were within normal limits.

At surgery, the findings were extensive sponge-like subcutaneous tissues containing serous fluid and lying between the skin and the muscle layer [Figure 3]. There

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were no clearly defined macro- or microcysts. The breast tissues were scanty and appeared to be immersed in the lesion. Extensive dissection but incomplete excision of the spongy tissues was performed, leaving those interspersed in the breast tissues. She had a partial flap breakdown in the postoperative period, which healed spontaneously [Figure 4].

At the last follow-up visit, the patient was a lot happier with her outlook, although she complained of a slight increase in the size of her left breast.

DISCUSSION

Lymphangiomas are swellings resulting from abnormalities of the lymphatic vessels in which there is impaired flow of lymph with consequent formation of fluid-filled vesicles and cisterns.[4] The vesicles could be macrocystic (as in cystic hygroma), microcystic or mixed.[5] Lymphoid tissue showing germinal centres may also be present.[6] The vast majority of these lesions occur in the head, neck and chest regions.[7,8] Whereas few cases exhibit the phenomenon of spontaneous regression, recurrence following surgical excision is a major worry. Complete excision of lymphangiomas is not always feasible because of a number of reasons.[9-11] Firstly, these lesions are often traversed by vital structures, which stand a risk of injury during surgery. Secondly, extensive dissections may be defacing, as in our index patient and thirdly, the extent of the lesion is often very difficult to define at surgery. In our patient, the site of the first lesion at birth was in the neck. The second lesion (first recurrence) was in the right infraclavicular area. The third lesion (second recurrence) transcended the sites of the previous lesions. This feature of recurrence away from the original site of the lesion had been reported by Adeyemi,[9] who suggested the term 'Progressive Infiltrating Cystic Hygroma'. This term would suggest that this condition spreads out into the normal adjacent tissue. It is possible that the adjacent tissues may have been harbouring the affected lymphatic channels, which simply opened up



Figure 1: The breasts became markedly distended (left > right) and the neck and the upper chest swellings disappeared when the patient sat up



Figure 2: The breasts shrunk remarkably while the upper chest and neck swellings became worse when she lay down

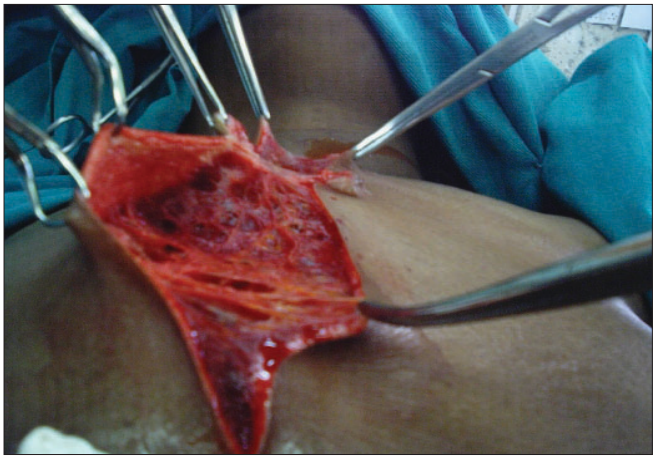


Figure 3: Extensive sponge-like subcutaneous tissues containing serous fluid and lying between the skin and the muscle layer



Figure 4: Partial flap breakdown in the postoperative period, which healed spontaneously

following excision of the dilated ones, or the cut ones regenerated afterwards. This to us seemed to be a more laudable explanation for the recurrences at sites away from the earlier ones.

The second recurrence in this report exhibited two interesting features: it transcended the sites of the previous lesions and there were no clearly demonstrable vesicles. The spaces were contiguous and fluid flow was free. Could it be that the vesicles had been so tensely distended that they ruptured with leakage of lymph into the tissues or that the earlier surgeries had eliminated the vesicles leaving open ends of lymph vessels to leak into the tissues?

As expected in such extensive lesions, the excision was incomplete to save the breast tissues. Having realized these shortfalls of surgery, obliteration of these channels by sclerotherapy is being attempted and evaluated by researchers as an alternative to surgery. Agents that have been used include bleomycin chloride,[12] OK-432,[13] doxycycline,[14] etc. but results are variable and acceptance is still generally low.

It seems that a step further in the management of extensive lymphangiomas should include sclerotherapy as an adjuvant to surgery rather than sclerotherapy as an alternative treatment modality. In view of the reported slight enlargement of the left breast at the last check-up visit, we wonder what treatment options we have not explored, should the breast enlargement prove another recurrence?

In conclusion, extensive lymphangiomas remain a daunting problem in the practice of surgery. Although complete excision is the treatment of choice, this is difficult to achieve in many cases. Sclerotherapy is not without pitfalls. We think that a combination of surgery and sclerotherapy may be an idea worth assessing as advancement in the future treatment of these lesions.

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