LYMPHEDEMA

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ALTHOUGH lymphedema is not a common cause of swelling of the extremities neither is it rare, and occasionally the family physician is consulted by an anxious adolescent girl who complains of progressive swelling of the legs. Too often the physician knows little or nothing about the condition and is inclined merely to sympathize with the patient whose symptoms indicate eventual "piano legs" or elephantiasis. It is true that once the end stage of intractable, fibrous edema is reached, treatment is generally unsatisfactory; however, initiated early in the disease it is usually successful, and will in most cases, keep the edema under control and avoid the adverse psychologic patterns often encountered in a woman with unsightly legs.

General unawareness of the entity of lymphedema stems from the fact that little has been written on the subject and consequently it is passed over lightly in medical schools and textbooks. In recent years the excellent works of Drinker and Field[1] on the anatomy and physiology of the lymphatic system, and Allen[2] on the classification, etiology and differential diagnosis based on 300 cases at the Mayo Clinic, have done much to clarify and simplify the previous confusion concerning lymphedema.

Definition. Lymphedema is an abnormal accumulation of lymph in the soft tissues. It is characterized by swelling, usually of the extremities, in the early stages, and by so-called elephantiasis, the result of continued edema and fibrosis, in the late stages. It is generally conceded that the term elephantiasis should not be used synonymously with lymphedema since it is merely a descriptive term which aptly describes the end result of a variety of conditions.

Mechanism. The accumulation of lymph in the tissues is due to obstruction of lymph flow caused by noninflammatory or inflammatory processes. Homans, Drinker, and Field[3] have shown that, as the lymph collects, there is a steady increase in its protein content (to 3 or 4 per cent in 5 patients). As the concentration begins to approach that of serum there is an active proliferation of fibroblasts and eventually increasing fibrosis of the deep layers of the skin and subcutaneous tissues. The fibrosis causes further stasis and the whole process becomes a vicious cycle. The stagnant lymph is subject to recurrent attacks of acute inflammation. This results in thrombosis of lymph vessels and further stasis. The terminal result is a proliferative, intractable swelling (elephantiasis) of the extremity.

Classification. The largest series of cases of lymphedema studied was reported by Allen[2] in 1934. He made a detailed study of 300 cases seen at the Mayo Clinic over a 10 year period and offered a classification of the disease which has been generally accepted as simple and accurate. His work has been

instrumental in clarifying the clinical picture of lymphedema. The disease is divided into the noninflammatory and inflammatory types as follows:

A. Noninflammatory:
   1. Primary—
      a. Praecox.
      b. Congenital (Simple or Familial).
   2. Secondary—
      a. Malignant occlusion.
      b. Surgical removal of lymph nodes.
      c. Pressure.
      d. X-ray and radium therapy.

B. Inflammatory:
   1. Primary.
   2. Secondary—
      a. Venous stasis.
      b. Dermatophytosis.
      c. Systemic diseases.
      d. Local tissue injury or inflammation.

Noninflammatory Lymphedema

Lymphedema Praecox. This is the most common type of lymphedema. It affects young women in about 90 per cent of the cases. It may begin in childhood but most often appears during puberty. Painless swelling occurs in one lower extremity in the majority of cases, affecting the foot first and slowly progressing up the leg. The edema may remain mild, increase slowly or, rarely, rapidly involve the whole leg. Occasionally both legs are affected at the beginning, but more frequently the second leg is affected weeks to months after the first. The edema is soft at first, pits on pressure, and is more pronounced during warm weather and during menstrual periods. At the beginning it disappears when the patient retires and is increased by prolonged standing and walking. During this early period treatment is most effective and results in either control of the edema or prevention of progressive enlargement. In the absence of treatment and, in a few stubborn cases, the edema becomes intractable and the irreversible progression to elephantiasis makes the skin hard, rough, occasionally scaly and resistant to pressure. There is no pain but the limb becomes heavy and uncomfortable. In a small number of cases, 13 per cent in Allen's series, one or more attacks of acute lymphangitis occurs.

The cause of lymphedema praecox is unknown but because of its incidence in young women, its usual onset during puberty and its accentuation during pregnancy, it is believed that the female reproductive organs influence the etiology.

Congenital Lymphedema. Congenital lymphedema exists in two forms: (a) hereditary lymphedema, most commonly known as Milroy's disease, and (b) simple congenital lymphedema. Firm, diffuse edema is present at birth

and affects one extremity, usually a leg. The hereditary type is extremely rare and several members of the same family may be affected. Milroy originally reported the disease in 1892. In a follow-up report[4] in 1928, he had traced the disease through 6 generations of 42 persons. Twenty-two had lymphedema, 21 of whom had the condition at birth. Various reports of Milroy's disease have appeared in the literature, but in most cases the edema appeared in children or in young adults. Allen[2] feels that these incidents represent lymphedema of some other type, probably lymphedema praecox, with a family predilection.

Simple congenital lymphedema has been shown pathologically to be due to lymphangiectasis, by Middleton,[5] Allen,[6] and others. The picture is typical in that normal subcutaneous fat is replaced by large, dilated lymph spaces and fibrous tissues. There is no evidence of inflammation or thrombosis of lymph vessels or blood vessels. It is probable that the cause of the lymphangiectasis is a developmental anomaly.

Secondary Noninflammatory Lymphedema. Secondary lymphedema is due to a variety of causes. Most familiar is the surgical lymphedema of an upper extremity following radical surgery for carcinoma of the breast. Any condition producing lymphatic obstruction or destruction can result in lymphedema. Malignant disease is a common cause, either primary as in the lymphoma group, or from metastatic invasion of the lymph nodes by carcinoma originating in the breast, prostate, female pelvic organs, and probably other organs. For this reason it is of primary importance to eliminate any possibility of malignant disease by careful examination in all cases of lymphedema. Radiotherapy and extrinsic pressure on lymph channels have been reported to have caused secondary lymphedema.

Inflammatory Lymphedema

Inflammatory lymphedema is characterized by single or recurrent attacks of cellulitis and lymphangitis followed by varying degrees of chronic lymphedema. If more than one attack occurs there is an increase in the degree of edema following each attack until a far advanced, chronic lymphedema results. When the cellulitis and lymphangitis occur without an apparent precipitating cause, the resultant swelling is referred to as primary lymphedema. When it follows dermatophytosis, filariasis, pregnancy, and various systemic and localized infections, it is known as secondary lymphedema.

Whether primary or secondary, a typical attack occurs suddenly with chills, fever, malaise and occasionally nausea and vomiting. The temperature rises to 102 F. or higher and prostration is usually pronounced. A painful, tender, warm red area appears in the extremity and the lymph nodes draining the area become enlarged and tender. The process spreads until the entire limb is swollen, hot and tender. The fever continues for several days to a week and the acute inflammation in the leg slowly subsides within 1 to 2 weeks. There is residual swelling due to thrombosis of lymphatic vessels which is usually persistent and becomes more severe after each attack. The responsible organism

has been shown to be streptococcus. If only one attack occurs the lymphedema generally subsides slowly as collateral lymph channels are developed. The edema will become chronic, however, if it persists long enough for fibrosis and the resultant additional stasis to occur. Lymphedema often follows an infection or injury without any evidence of cellulitis or lymphangitis. Allen believes that the lymphatic infection in these cases is subclinical.

Diagnosis

Diagnosis of lymphedema is not often difficult. When the swelling is not severe and advanced skin changes have not occurred it must be distinguished from lipodystrophy. Both conditions occur more frequently in women and are similar in general appearance. The swelling of lipodystrophy is rather soft and spongy and either does not pit or pits slightly on pressure, and is always bilateral. Lymphedema is usually unilateral, is firmer in consistency and usually pits readily on pressure. Lipodystrophy is not progressive, does not disappear on rest and elevation of the legs, and occurs in obese women. Lymphedema is progressive and diminishes noticeably after a periodic elevation of the extremities. The advanced, chronic changes of lymphedema are characteristic, and are not seen in other conditions.

When bilateral, lymphedema must be distinguished from the edema caused by heart failure, kidney disease and hypoproteinemia, and when unilateral from chronic venous insufficiency and arteriovenous fistula. Acute lymphangitis may be confused with acute thrombophlebitis but the septic course and clinical evidence of lymphatic involvement is not observed in thrombophlebitis.

Treatment

The treatment of the early stages of lymphedema is generally satisfactory. It should be re-emphasized that the condition is not hopeless and the victim must not be allowed to become discouraged regarding the outcome. Treatment is exceedingly simple and if diligently pursued early in the disease will keep the edema at a minimum and prevent deformity. The patient should remain in bed, with the foot of the bed elevated to 10 to 12 inches, until the maximum reduction in size occurs. This may take only 2 or 3 days but occasionally a week is required. When there is no longer any decrease in the swelling the patient is provided with a well-fitted elastic stocking and allowed to be ambulatory. The stocking should be fitted only when the edema is at a minimum, preferably the first thing in the morning, and should always be put on before the patient gets out of bed. The limbs should never be dependent unless a good support is worn. Pure rubber roller bandages make an excellent support but because they are hot, cumbersome, and give a poor appearance most women refuse to wear them. Elastic stockings come in several weights and are made of either cotton or nylon; however they are fairly expensive and must be replaced several times a year. The patient should continue to sleep with the foot of the bed elevated. During the day the legs should be elevated at every opportunity. When the edema seems to be controlled the support may be

discontinued for a day or so but should be resumed if edema reappears and worn for another month before a further trial without support. Acute attacks of lymphangitis and cellulitis respond well to antibiotics which shorten the duration of the attack significantly. This treatment should be combined with the use of massive, hot, wet packs. In the primary type recurrent attacks are difficult to prevent but chronic infections, including dermatophytosis, should be eradicated if possible.

There is no satisfactory treatment for lymphedema following surgery for carcinoma of the breast, but occasionally elevation of the arm will reduce the swelling somewhat. The lymphedema caused by obstruction is usually the result of malignant disease and treatment should be directed toward the primary cause. In most instances the malignancy is far advanced, and except in rare cases of Hodgkin's disease and lymphosarcoma, which respond to radiotherapy or nitrogen mustard, treatment is merely symptomatic.

Surgical treatment of lymphedema is reserved for those far advanced cases of elephantiasis in which the extremity is massive in size, cumbersome and unsightly. The modified Kondoleon operation as advocated by Ghormley[7] and Pratt[8] is the procedure of choice. The subcutaneous tissue and fascia is completely removed so that the skin becomes attached to the muscle and the lymph is drained off through the muscle lymphatics. The limb can be reduced to normal size but the lymphedema recurs in a fairly large percentage of patients if an elastic support is not worn for an indefinite period postoperatively.

References

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