Lymphangiosarcoma is one of the rarest forms of soft tissue neoplasms. Lowenstein (1906) was the first to report this entity in a patient with chronic post-traumatic upper extremity lymphoedema. However, it was not until Stewart and Treves (1948) described the development of this malignancy in a post-mastectomy oedematous extremity that this syndrome, which carries their name, became widely recognized.

The tumour is highly malignant and usually arises in a lymphoedematous upper extremity of a patient treated for carcinoma of the breast. It has been well established that lymphangiosarcomas are not directly related to breast cancer (Aegerter & Peale, 1942). The common denominator is the lymphoedema and, therefore, other causes of chronic lymphoedema may lead to the development of this vascular sarcoma (Taswell et al., 1962). Indeed, this disease has occurred in congenital (Dubin et al., 1974), idiopathic, traumatic and filarial lymphoedema (Devi & Bahuleyan, 1977). The pathogenesis of these unusual tumours remain uncertain. Stewart and Treves (1948) believed that a systemic carcinogen was involved in the etiology. Martorell (1951) suggested that the lymphoedema may be tumour inducing while Schreiber et al. (1979) postulated that local immunodeficiency was the causative factor.

Histologically these tumours consist of large vascular cavities lined with plump, spindle-shaped endothelial cells containing large nuclei and prominent nucleoli. Some areas may exhibit solid sheets of spindle cells (Silverberg et al., 1971). Immunohistochemistry may prove to be quite useful; factor VIII-AG is the most utilized marker for vascular differentitation (Mukai et al., 1980).

Lymphangiosarcomas affect all races including blacks (DiSimone et al., 1970), orientals (Tomita et al., 1978) and caucasians. The median time from mastectomy to the development of lymphangiosarcomas is 10 years (Woodward et al., 1972), ranging from 5 (Yap et al., 1981) to 27 years (Enzinger & Weiss, 1988). The average age of patients is about 62 years (Southwick & Slaughter, 1955).

In a review of 894 women who survived 5 years following radical mastectomy with or without radiation therapy, Shirger (1962) reported an incidence of Stewart–Treves syndrome of 0.45%. Fitzpatrick (1969), however, encountered only 7 patients (0.07%) who developed lymphangiosarcoma when he reviewed the records of 9000 patients treated with mastectomy. All seven had had adjuvant radiation therapy. He noted that lymphoedema was a complication more likely to follow Halsted's operation as compared with less radical surgery and he stated that the risk of lymphoedema increased if the axilla was irradiated. Since lymphangiosarcoma is a rare disease, most reports consisted of a small number of patients. By 1972, approximately 200 cases of lymphangiosarcomas complicating chronic lymphoedema had been recorded in the world literature (Woodward et al., 1972).

Clinically the tumour appears as bluish, purplish, reddish skin nodule(s) on a lymphoedematous arm. These lesions tend to grow, coalesce, ulcerate and spread proximally and distally to involve the entire arm. The disease also tends to metastasize through the blood stream to distant organs, particularly the lung.

Treatment of extremity lymphangiosarcoma includes local excision, wide excision, amputation, radiation therapy, chemotherapy, and combined methods (Southwick & Slaughter, 1955; Chu & Treves, 1963; DiSimone et al., 1970; Yap et al., 1980; Sordillo et al., 1981; Yap et al., 1981). The prognosis is poor, as most patients die within 5 years of diagnosis despite various forms of treatment. The median survival is about 2.5 years (Sordillo et al., 1981) with very few long-term survivals.

The purpose of this report is to describe two patients with Stewart–Treves syndrome who survived at least 13 and 19 years following combined modality treatment. They were among the first patients with this condition to be treated with Actinomycin D chemotherapy and radiation therapy, with curative intent.

**Case reports**

**Case 1**

The patient was a 66-year-old white female who underwent a left radical mastectomy in 1952 for infiltrating duct carcinoma...
Case reports

Figure 1. Recurrent lymphangiosarcoma involving the left anterior chest wall near the amputation stump.

with metastases to the axillary lymph nodes. She received a course of post-operative radiation therapy, details of which were unavailable. Shortly after surgery the patient developed left arm lymphoedema.

In September 1966 she noticed induration and pain throughout her left arm and, 2 weeks later, numerous bluish skin nodules appeared; biopsy showed lymphangiosarcoma. It was decided to treat the patient with a left interscapulo-thoracic amputation combined with Actinomycin D chemotherapy. The first dose of Actinomycin D, 2 mg intravenously, was administered 6 days prior to surgery and her second dose was on November 30, 1966, the day of surgery. Her surgical specimen demonstrated lymphangiosarcoma present in multiple subcutaneous nodules and diffusely in the dermis. The margins of resection were free of tumour.

2 months later two purplish skin nodules were noted on her left anterior chest wall (Fig. 1). Biopsy again demonstrated lymphangiosarcoma (Fig. 2). The patient was referred for radiation therapy which was started on February 27, 1967.

Using a single portal measuring 15 x 15 cm, the left anterior chest wall was irradiated to a total dose of 5500 cGy in 27 fractions over 31 elapsed days, using a 6 MeV electron beam. The two nodules received an additional boost dose of 500 cGy, delivered in 2 fractions in 4 days through a reduced field.

3 weeks later a small new lesion was noted at the upper border of the radiation field. This was treated with a 3.3 cm circular field to a total dose of 3200 cGy in 16 fractions delivered in 3 weeks, using 6 MeV electron beam. All three nodules disappeared following therapy.

The patient again received Actinomycin D chemotherapy, 2.0 mg intravenously monthly, from May 12, 1967 until October 30, 1970, which she tolerated well. She was last seen in 1985, 19 years after radiation treatments; she was without any evidence of recurrent lymphangiosarcoma (Fig. 3). Since the patient resides in Sweden, there has been no further follow-up.

Case 2

The patient was a 66-year-old white female who had undergone a right radical mastectomy in 1962, at the age of 37, for infiltrating duct carcinoma with negative axillary nodes. She received no adjuvant radiation therapy. Lymphoedema of the right arm was noted shortly after mastectomy. In 1977, 15 years following the radical mastectomy, a small bluish,
purplish, flat lesion was noted on the lateral aspect of her right arm. Incisional biopsy revealed lymphangiosarcoma and the patient was referred for radiation therapy. Since lymphangiosarcoma tends to have extensive local involvement, it was planned to irradiate a wide area of the arm. The lateral aspect of the arm was treated first. A total dose of 5400 cGy was delivered in 27 fractions over 44 elapsed days, using a 15 MeV electron beam. This was followed by irradiation of the medial aspect of the right arm and a total dose of 5000 cGy in 25 fractions was delivered over a period of 36 days. A narrow, longitudinal strip of the arm was spared in order to preserve adequate lymphatic drainage.

Since Case 1 had done so well with Actinomycin D, the same drug was administered to Case 2 after completion of radiation therapy. She received Actinomycin D, 2.0 mg intravenously, once weekly for 3 consecutive weeks, followed by a 2 week rest period. Maintenance therapy continued from April 1978 to November 1979. The patient remained in good health until December 1988 when she developed carcinoma of the opposite breast. She underwent a left modified radical mastectomy and the pathology was infiltrating duct carcinoma with negative axillary lymph nodes. She was last seen in September 1990. 13 years after treatment there was no evidence of recurrent lymphangiosarcoma and she was also free of breast cancer.

Discussion

Lymphangiosarcoma is an aggressive cancer with a rapidly progressing clinical course. McConnell and Haslam (1959) described three phases of development of post-mastectomy lymphangiosarcomas, i.e., prolonged lymphoedema, angiomatosis and, finally, angiosarcoma. However, overlapping among the three phases is possible. Chu and Treves (1963) suggested that longer survival might be associated with the diagnosis in early phases. The importance of early recognition of a suspicious bluish, reddish lesion on a lymphoedematous arm, immediate biopsy and prompt treatment cannot be overemphasized.

Lymphangiosarcoma occurs in a lymphoedematous extremity. Prevention of lymphoedema would logically lower the incidence of the development of lymphangiosarcoma. During the past few decades, there has been a definite trend toward performing less radical surgery in the treatment of breast cancer. Modified radical mastectomy and partial mastectomy have largely replaced the classical Halsted radical mastectomy, because the survival results are similar with reduced morbidity including the incidence and severity of lymphoedema of the arm. Although no survey regarding the frequency of lymphangiosarcoma in currently treated patients has been available, it is our clinical impression that we are seeing less numbers of patients inflicted with this disease in recent years.

Management strategies include surgery, radiation therapy and chemotherapy. Surgical treatment should consist of either wide excision or amputation. Sordillo et al (1981), in a review of 44 cases seen at the Memorial Hospital, New York City, reported a median survival of 48 months in 11 patients treated with amputation, as compared with 20 months median survival in other patients with radiation therapy or chemotherapy. However, the numbers of cases were relatively small so no firm conclusion can be made regarding the efficacies of various treatments.

Radiation therapy has been used frequently for adjunctive, curative or palliative treatments. Lymphangiosarcomatous lesions do respond to radiation therapy (Southwick & Slaughter, 1955; Chu & Treves, 1963; DiSimone et al, 1970) with an occasional cure. Since lymphangiosarcoma is usually a multifocal disease with extensive clinical or subclinical involvement, radiation therapy should play a major role in its management. We recommend wide-field technique, even when the lesion may appear to be small and limited, and high doses of radiation of the order of 5500 to 6000 cGy delivered in 27–30 fractions over a period of about 6 weeks. This would mean treating practically the entire arm, sparing only a narrow longitudinal strip of skin to preserve lymphatic drainage. An example of the wide-field technique can be seen in the treatment of Case 2 whose lesion appeared to be clinically solitary.

Chemotherapy is a relatively new method of treatment compared with surgery and radiation therapy, so our experience regarding its efficacy in lymphangiosarcoma is extremely limited. It is also difficult to determine which regimen is more effective, since lymphangiosarcoma is a rare disease and very few patients are available for study.

Yap et al (1981) reviewed 22 patients seen at M. D. Anderson Hospital for post-mastectomy lymphangiosarcoma over a 20 year period. 13 patients were treated regionally or systemically or both, with a variety of single or combination chemotherapeutic agents; the overall response rate was 42%. Five patients who failed to respond to chemotherapy had a median survival of 4 months, whereas the six chemotherapy responders had a median survival of 26.5 months.

Our two cases of long-term survival were managed with high doses of radiation therapy and Actinomycin D. Case 1 was actually an amputation failure and the two doses of Actinomycin D, given before and during surgery, were inadequate to prevent recurrence. However, the recurrent disease was salvaged by a combination of radiation therapy and Actinomycin D. The excellent results may suggest that chemotherapy and radiation therapy have additive or synergistic effects. Our first patient was treated 24 years ago and the second 13 years ago. At that time Actinomycin was the drug of choice for these two patients. More recently, the most frequently used agent for treatment of vascular sarcomas is doxorubicin (Adriamycin), either alone or in combination with other agents.

We recommend close follow-up of all patients with post-mastectomy lymphoedema of the arm. Biopsy of any suspicious lesion should be carried out without delay. Aggressive management with surgery, radiation therapy and chemotherapy may offer an optimistic outlook if the neoplasm is discovered in its early phases. Our experience with these two patients showed encouraging results with a combination of radiation therapy and Actinomycin D treatments.
Acute epidural haematoma complicating myelography in a normotensive patient with normal blood coagulability

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Acute epidural haematomas are usually idiopathic, but they may occur following spinal trauma (Coin et al, 1979), epidural anaesthesia (Helperin et al, 1971) or in association with spinal tumours (Harris et al, 1978) or vascular malformations (Muller et al, 1982). We report the computed myelographic, computed tomographic (CT) and magnetic resonance imaging (MRI) findings in an unusual case complicating lumbar myelography, requiring surgical decompression, which was not evident on the myelogram.

Case report

A 75-year-old woman with chronic low back pain and sciatica presented with increasingly severe right leg and buttock pain. This had developed after surgery for an L4/5 spondylolisthesis and spinal canal stenosis 2 years previously. In 1984 she had an

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