Radiation Treatment of Postmastectomy Lymphangiosarcoma

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Since the entity of postmastectomy lymphangiosarcoma was first reported by Stewart and Treves in 1948, postmastectomy lymphangiosarcoma has become a well recognized, uncommon malignant tumor which occurs in the upper extremity following mastectomy for mammary carcinoma. The postmastectomy lymphangiosarcoma occurred at an average age of 63.9 years and at an average of 10 years and 3 months following mastectomy. The lymphangiosarcoma raised from blood and lymphatic vessel. The histologic appearance has been observed edematous dermis and dilated lymphatics lining with malignant cells. Most authors recommend radical amputation for treatment, either shoulder disarticulation or forequarter amputation. Other modalities of treatment including radiotherapy were considered as ineffetive.

The present report provides a case of the regression of postmastectomy lymphangiosarcoma with chronic lymphedema by external irradiation. Radiation therapy was used as primary therapy. Total tumor dose of 6500 cGy in 9 wks was delivered using 6 MV x-ray and 8 MeV electron.

Key Words: Postmastectomy lymphangiosarcoma, Lymphedema, Mastectomy, Radiation therapy

INTRODUCTION

Postmastetomy lymphangiosarcoma is an uncommon malignant tumor which occurs in the upper extremity following mastectomy for mammary carcinoma. Since 1948, when Stewart and Treves first recognized the syndrome of postmastectomy lymphangiosarcoma[1], the occurrence of lymphangiosarcoma associated with chronic lymphedema has become a well recognized entity. This aggressive neoplasm is often associated with postmastectomy lymphedema of the upper extremities. The onset skin changes show the blue-red or purple, well defined macular or papular lesions in the edematous skin and subcutaneous tissues. In later stages, the lesions are multiple and coalesce to form a large ulceration and hemorrhagic appearance, and infrequent involving regional muscle layers. This neoplasm is arising from endothelium of lymphatic spaces and proliferation of lymphatic vessels lining by malignant -appearing cells[2].

The best method of treatment is not known. But most authors recommend radical amputation rather than radiation therapy or chemotherapy, because of the diffuse nature of these tumors[3]. The present report provides the regression of post-Mastectomy lymphangiosarcoma with chronic lymphedema by external Radiation.

CASE OF REPORT

A 79 year old woman had undergone mastectomy for a infiltrating ductal ca of left breast in 1977. Postoperative radiotherapy was not administered. She had developed edema at left upper extremity after 2 years following mastectomy. But she did well for 10years, after which time she developed persistent purplish discoloration at the skin of left forearm in August 1986. In October 1987, a skin biopsy confirmed lymphangiosarcoma(Fig. 1). She was referred to our department for radiation therapy.

An evaluation of metastatic lesion or recurrent disease of previous breast ca was entirely negative. The skin lesion was multiple bluish nodules within a skin area measuring 7.5×7.1 cm. The nodules were discrete, slightly elevated and indurated, the largest measuring 3 mm in diameter. Also noted were larger areas of bluish discoloration of surface in association with mild tenderness and severe pain.

The plain x-ray film of left forearm revealed no abnormality. A CT scan showed the size of the left forearm is larger than the right, and there is tissue of irregular outline and increased attenuation in the subcutaneous tissues encircling the limb on the

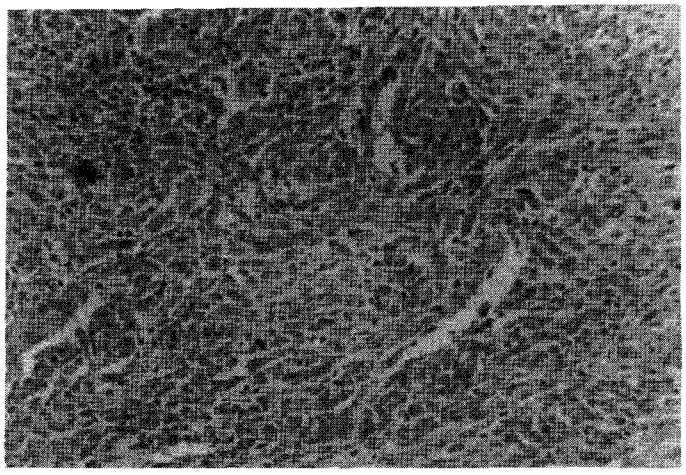

Fig. 1. Anastomosing vessels lined with endothelial cells protruding into and filling the lumina. (HE x150)

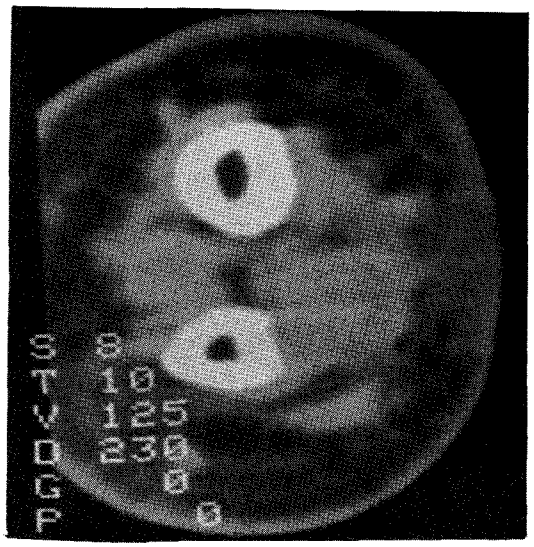

Fig. 2. The mass blended with surrounding lymphedema and its attenuation was lower than that of the muscle without definite evidence of muscle invasion.

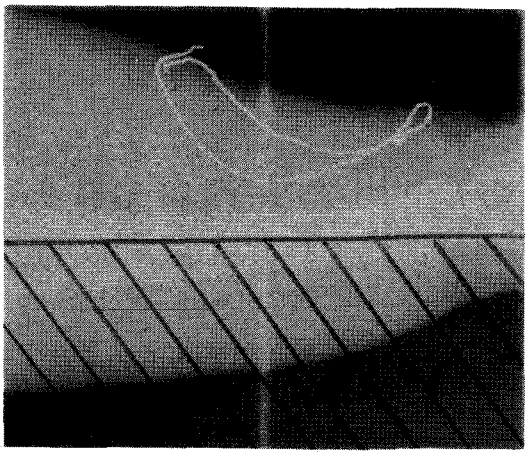

Fig. 3. Radiation field.

right, representing lymphedema and dilated lymphatic channels. The mass blended with surrounding lymphedema and its attenuation was lower than that of the muscle, without definite evidence of muscular invasion(Fig. 2). The thickness of mass measured 1.5 cm. She had been evaluated with chemical profile, urinalysis, CBC, and no abnormality founded.

Radiation therapy was used as primary therapy. Total tumor dose of 6500 cGy in 9 weeks was in daily fractionation with a fraction size of 200 to 250 cGy was delivered using 6 MV x-ray and 8 MeV electron. The portal technique were parallel opposing fields and anterior one portal. The initial portal size was 10×12 cm(Fig. 3). Complication were not observed except for mild radiation dermatitis. Clinically partial remission of lesion was noted after completion of irradiation. There were loss of nodules and blurring of purplish disocloration with improved painful sensation and tenderness. Following the RT (6 months), she showed no evidence

of disease except for small sized, faint discoloration of the skin.

DISCUSSION

Since the entity of postmastectomy lymphangiosarcoma was first delineated by Stewart and Treves, in 1948[1], the occurrence of lymphangiosarcoma associated with chronic lymphedema has become a well recognized entity. The postmastectomy lymphangiosarcoma occurred at an average age of 63.9 years and at an average of 10 years and 3 month following mastectomy. The favorite site was the inner aspect of upper arm. The lymphangiosarcoma raised from blood and lymphatic vessel. The histologic appearance have been observed edematous dermis with dilated lymphatics. In the subcutaneous tissue, there were numerous nodules composed of round of oval tumor cells, growing in loosely arranged cords and sheets. The cells contained relatively large hyperchromatic nuclei with scanty, poorly demarcated, eosinophilic cystoplasm. Vascular channels of various sizes and configuration were prominent throughout the tumor[2,4].

The best method of treatment is not known. Because of the diffuse nature of these tumors. most authors recommend radical amputation, either shoulder disarticulation or forequarter amputation. Local excision of lcoalized tumor is usually followed by recurrent disease. Woodward and colleagues reported 23 cases of postmastectomy lymphangiosarcoma and review the world literature[3]. These authors reported that six patients using irradiation as primary therapy had limited or no tumor response, and radiation therapy has resulted two long term survivors only. Some patients had transient regression of primary lesion after irradiation. But recurrence can be expected in nearly all instances. A few patients have had chest wall spread after amputation, and this have been controlled by irradiation. In our case, there are good response of primary lesion by irradiation. However we cannot decided that this response was transient regression or not, because duration of the follow-up was short term. Postoperative radiation therapy after mastectomy increased lymphedema of extremity, but no evidence of carcinogen for lymphangiosarcoma. Combined chemotherapy was not resulted in an increased number of survivors[3,5].

The prognosis of postmastectomy lymphangiosarcoma is very poor. Fifty per cent of the 129 patients were dead within 19 month after treatment. The most common of the isolated metastasis was chest wall, result in death without dissemination of disease. The important factor of prognosis may be early diagnosis in some case[3]. Early diagnosis also may be the key to survival in some case. Close physical examination to the skin the edematous extremity should lead to increased survival rate.

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═국문초록═

유방 절제술후 임파육종의 치료 1예

가톨릭의과대학 방사선치료실

최일봉 · 길학준 · 김미희 · 김춘열 · 박용휘

유방절제술후 임파육종은 유방암 환자에서 유방절제술후 상지에 생기는 흔치 않은 악성 암이다. 유방절제술후 임파육종에 평균 발병 연령은 63.9세이고 유방절제 후 발병할 때까지의 기간은 평균 10년 3개월이다. 유방절제술 후 임파육종은 혈관과 임파관에서부터 발생하는 것으로 알려져 있고 조직학적 소견은 진피의 부종과 악성세포로 둘러싸인 확장된 임파선이 특징이다.

견관절 분리술이나 전사분부 절제술 등의 광범위한 절제가 가장 효과적인 치료법으로 알려져 있고 방사선 치료를 포함한 다른 치료방법은 큰 도움이 않되는 것으로 보고되고 있다.

저자들은 유방절제술 후 만성 림프부종을 동반한 임파육종이 외부 방사선 조사에 의하여 종괴의 감소를 보여 이에 보고하는 바이다.