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하지의 만성 림프부종에 발생한 혈관육종
- Stewart-Treves 증후군 -

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혈관육종은 악성 연부 조직 종양 중 매우 드문 형태의 종양으로 그 중 만성 림프부종에 발생한 것을 Stewart-Treves syndrome이라 부르기도 한다. 이는 일반적으로 유방암의 치료로 행해지는 유방제거술 및 방사선 치료 후 상지의 장기간의 림프부종의 합병증으로 주로 발생되는 것으로 알려져 있으며 예후는 좋지 않은 것으로 보고하고 있다.

저자들은 17년 전 자궁암으로 자궁적출술을 받은 환자가 수술 후 발생한 장기간의 하지의 림프부종으로 인해 1년 전 타병원에서 대퇴부의 수술적인 치료를 받았으나 이 후 대퇴부의 혈관육종이 발생하여 광범위 절제술을 시행한 예를 경험하였기에 이를 문헌 고찰과 함께 보고하는 바이다.

색인 단어: 림프부종, 혈관육종

Angiosarcoma is a vascular origin malignant tumor, and generally, it occurs in the skin, breast, liver and deep soft tissues, and it is developed in the incidence of less than 1% of the entire sarcomas[2]. Among them, cases developed during chronic lymphedema are referred to as Stewart-Treves syndrome[9]. In 1948, Stewart and described it for the first time in lymphedema patients after mastetomy[9]. Stewart-Treves syndrome usually develops more than 10 years after surgery and radiation therapy for breast cancer, and the incidence has been reported to be from 0.07 to 0.45%[3]. In general, this disease develops in cases with chronic lymphedena in the upper limbs after mastectomy, and rarely, it is developed in the lower limbs with lymphedema like our case[9]. They are treated by wide excision as well as radiation therapy, nonetheless, their prognosis has been reported to be poor[6,7,10]. We report a rare case of angiosarcoma in a chronically lymphedematous leg treated with wide excision. Our report is followed by a review of the literature.

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Case report

A 58 years old female patient was admitted to our hospital with the chief complaint of a mass in the left femoral area developed from one year ago. She had the disease history of hysterectomy for cervical cancer 17 years ago, and from 16 years ago, lymphedema was developed in the left lower limbs. The lymphadema in the lower extremities was persistent for 16 years, and she had the history of volume reduction surgery for lymphedema 1 year ago at another hospital. The past history of trauma was absent, and in physical examination at the time of admission, lymphedema in the lower limbs was severe to the level that walking is impossible, and in the medial side of the left thigh, a protruded red mass 3 x 3 cm in size was detected (Fig. 1). In simple radiograph performed at the time of admission, results of the enhancement of contrast in the left femoral soft tissues could be seen (Fig. 2), and in MRI, edematous change in

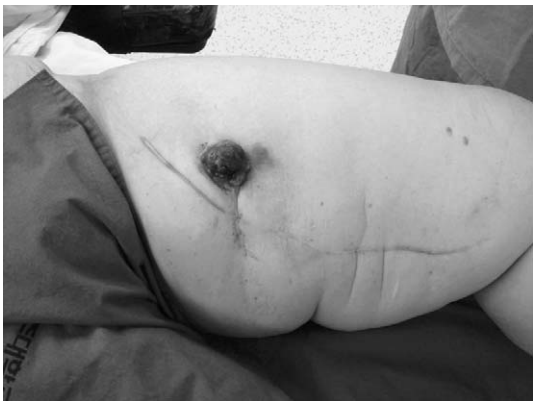

Fig. 1. Dark colored protruding mass at anteromedial side of lymphedematous thigh is shown.

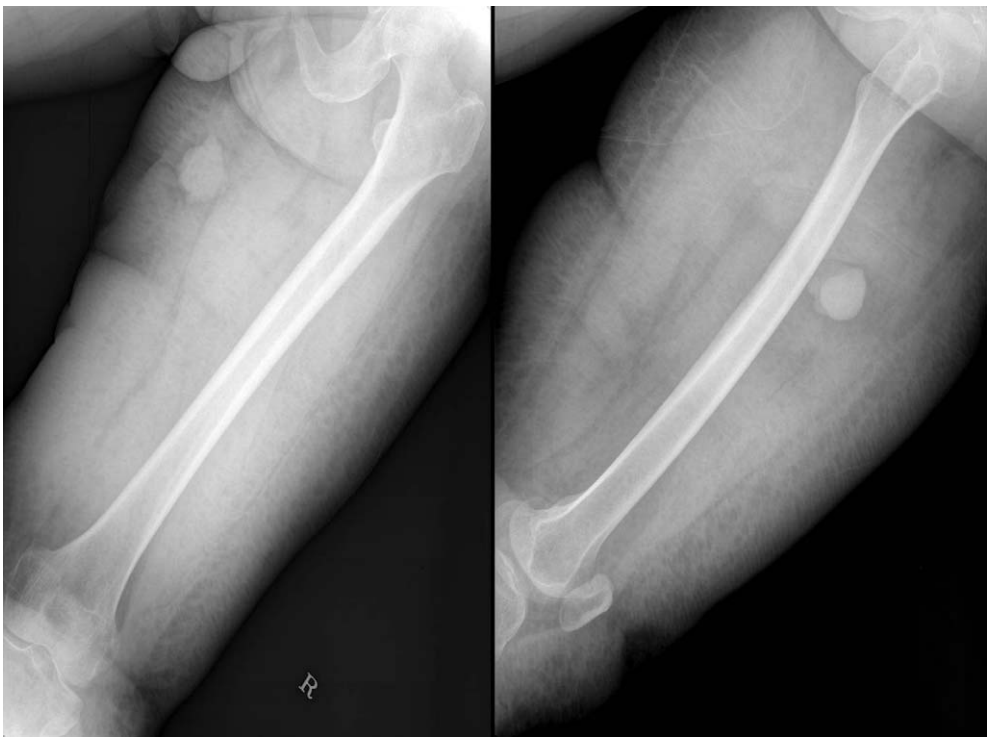

Fig. 2. Anteroposterior view of the left thigh shows diffuse high signal alterations and edema at subcutaneous fat layers.

the subcutaneous adipose tissues was observed, and in anterolateral area of the thigh, a mass 3.5×2.4×3 cm in size was observed (Fig. 3). In histological examination, it was diagnosed as high grade angiosarcoma. Pathologic findings show papillary projection pattern, vascular channels lined by multiple layer of atypical endothelial cells, pleomorphic and prominent mitotic activity. Wide excision with safety margin of 2 cm was performed (Fig. 4). After surgery, as radiation therapy, the left femoral area was radiated, 180 cGy/day 28 times, total 5040 cGy, and 15 mg adriamycin was administered 8 times. Nonetheless, from 1 year after surgery, a tumor recurred in the lateral side of thigh, and thus amputation was recommended, but the patient refused additional treatments.

Discussion

Angiosarcoma is a rare tumor with the incidence less than 1% of the soft tissue sarcomas and different from other soft tissue sarcomas, it is not located in the deep area

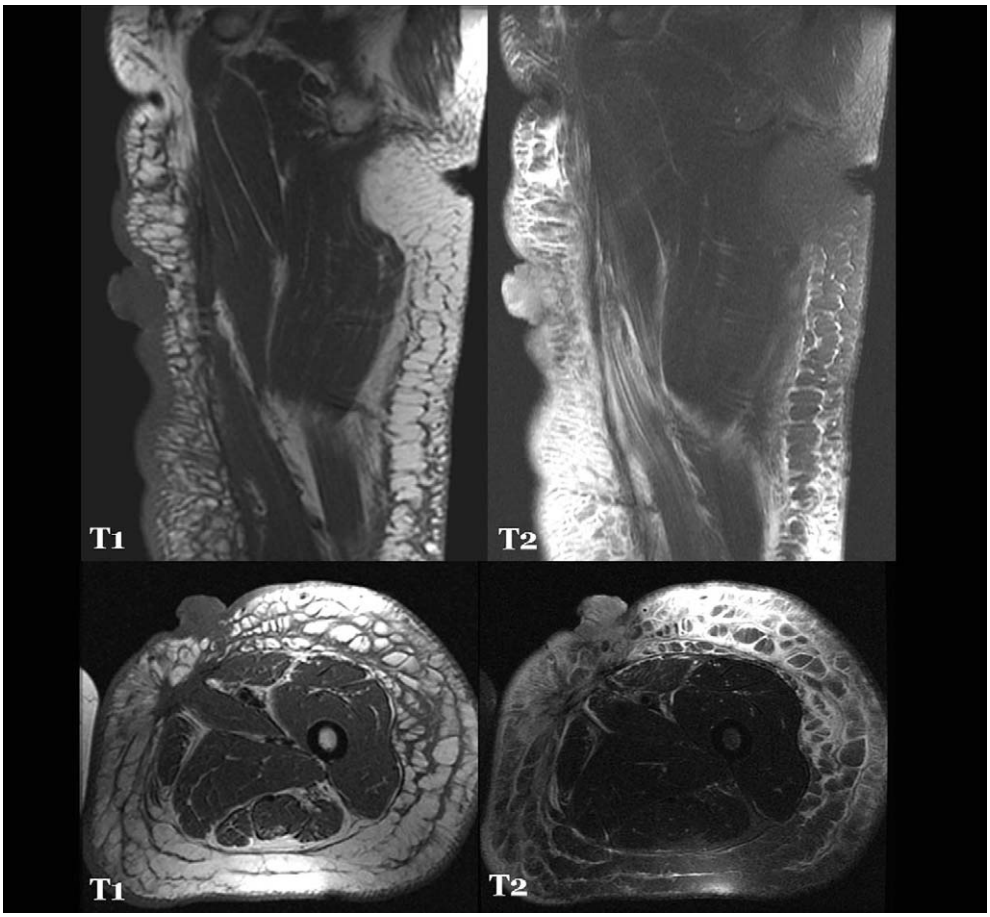

Fig. 3. Magnetic resonance imaging shows a soft tissue mass like signals which identified at left anteromedial proximal thigh (about 3.5×2.4×3 cm sized). After contrast administration heterogenous enhancement was seen at this mass. There were also diffuse thickening and enhancement along the superficial fascia over the adductor longus muscle.

and developed in the superficial layer primarily[2]. As its induction factors, radiation, lymphedema, ulcer, or chlorovinyl and other chemical substances have been reported[3]. Among them, chronic lymphedema has been known to be the most important factor, and although rare, cases developed angiosarcoma without edema have been reported. Among them, angiosarcoma developed in the area with chronic lymphedema is referred to as Stewart-Treves syndrome, and in 1948, Stewart and described it for the first time in lymphedema patients after mastetomy[9]. Stewart-Treves syndrome develops more than 10 years after surgery and radiation therapy for breast cancer, and the incidence is very low, and it has been reported to be from 0.07 to 0.45%[3] (Table 1). Cases developed Stewart-Treves syndrome have been reported by Komorowski, Schmitz-Rixen, and others, and it is not abundant[3,7,8]. Stewart-Treves syndrome has to be differentiated from Kaposi's sarcoma, breast cancer metastasis, malignant lymphoma, etc., and the development of angiosarcoma in the lower extremities is not frequent, nonetheless, angiosacroma may be induced by previous radiation therapy, after surgery for cervical cancer, filariasis, frequently recurring erysipelas, lower extremity fracture, etc.

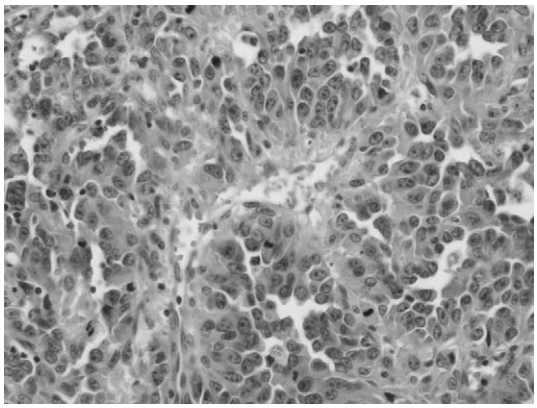

Fig. 4. Pathologic findings show papillary projection pattern. vascular channels lined by multiple layer of atypical endothelial cells (HE, ×40) the tumor cell is larger than normal endothelial cell. Pleomorphic and prominent mitotic activity. CD31(+), CD34(+), Factor VIII(+), CK7(-), CK20(-), Pan CK(-), HMB 45(-)

Table 1. Clinical data of reported cases

author	Age/Sex	Site	Periods with edematous condition	Primary cause	Previous condition
Stewart[9]	52/F	arm	30 years	Breast cancer	Mastectomy/RT
	60/F	arm	9 years	Breast cancer	Mastectomy/RT
	59/F	arm	10 years	Breast cancer	Mastectomy/RT
	37/F	arm	6 years	Breast cancer	RT
	65/F	arm	25 years	Breast cancer	Mastectomy
Komorowski[3]	50/F	arm	14 years	Breast cancer	Mastectomy
	60/M	leg	25 years	Primary chronic lymphedema	
Rodriguez-Bujaldon[7]	83/F	arm		Breast cancer	Mastectomy/RT
	42/M	leg		Primary chronic lymphedema	
Lee[4]	74/F	arm	10 years	Breast cancer	Mastectomy/RT
Chen[1]	71/M	arm	8 years	Malignant melanoma	Wide excision/CTx
Malhaire[5]	73/F	arm	10 years	Breast cancer	Mastectomy/CTx/RT

* RT : Radiotherapy, CTx : Chemotherapy

[1,7,8]. In most patients, the diameter is larger than 10 cm at the time of admission, and grossly, it appears as a blue or purple nevus or parpule and thus it may be considered to be a trace of contusion, and it may form a mass. In addition, because of lymphedema for a long time as well as hyperkeratic nodule, tubercle and crack, sarcoma may be ignored and thus misdiagnosed in some cases. Stewart-Treves syndrome occured usually in upper extremity after mastectomy and radiotherapy, and there have been In our case, the patient was underwent hysterectomy for cervical cancer 17 years ago, developed lymphedema in the lower extremities 1 year after surgery that was persistent for 16 years, and thus for the treatment of lymphedema, volume reduction surgery was performed, nonetheless, angiosarcoma was developed in the area with lymphedema.

As treatments, wide or radical excision has been performed most commonly, and amputation is performed in some cases, Additionally, radiation therapy may be administered, it may be from 4500 cGy to 5000 cGy, and for severe cases, it may be administered at the dose from 7000 cGy to 7500 cGy, and cases completely cured by radiation therapy have been reported, nonetheless, radiation therapy, chemotherapy, etc. did not show satisfactory results overall, and thus it is still controversial[5,6]. There are few reports about the cases for lower extremity to date, almost reports are about upper extremities after mastectomy and radiation therapy (Table 1). Our case is unusual in lower extremity. In our case, after wide excision, 5040 cGy radiation therapy and chemotherapy using adriamycin administered, nonetheless, from 1 year after surgery, a tumor recurred in the lateral side of thigh, and thus amputation was recommended, nevertheless, the patient refused additional treatments. It has been reported that the prognosis of angiosarcoma is very poor and local recurrence is frequent, and metastasized in the lung, liver, etc. within 2 years, and the survival is between average 19 months and 24 months, and 5-year survival rate has been reported to be 10%[3,4,6]. We experienced a rare case of angiosarcoma in a chronically lymphedematous leg and treated with wide excision.

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Abstract

Angiosarcoma in a Chronically Lymphedematous Leg - Stewart-Treves Syndrome -

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Angiosarcoma is a very rare disease of soft tissue sarcoma, and angiosarcoma arising in a region of chronic lymphedema is referred to as Stewart-Treves syndrome. it typically occurs in postmastectomy lymphedema of the arm and sporadically in a lymphedematous leg. The prognosis, even with wide surgical excision and subsequent radiotherapy, is poor.

The authors experienced a case of angiosarcoma in a chronically lymphedematous leg which had undergone hysterectomy due to cervical cancer 17 years ago and lymphedema of leg persists for about 16 years. We report a case of angiosarcoma with a brief review of the literature.

Key Words: Lymphedema, Angiosarcoma

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