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Leiomyosarcoma arising in the Great Saphenous Vein

— A Case Report —

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Though leiomyosarcoma usually occurs in the gastrointestinal tract and uterus, it rarely occurs in the wall of large veins and arteries. We present a case of primary leiomyosarcoma arising in the great saphenous vein of the left inner thigh and spreading for some extent along the vein in a 54 year old female. Diagnosis was comfirmed by desmin stain and electron microscopy. Postoperative course was fine. This is the first report of this in Korean literature.

Key Words: *Leiomyosarcoma, Saphenous vein, Vascular leiomyosarcoma, Smooth muscle*

INTRODUCTION

The majority of leiomyosarcomas occur in the muscular layers of the gastrointestinal tract and uterus. Less commonly these tumors may be found in the skin and soft tissue and are rarely found in the large veins and arteries of the extremities (Berlin et al, 1984; Dahl et al, 1981, Dorfman & Fishel 1963; Enzinger & Weiss, 1990; Fields & Helwig, 1981; Haug & Losli, 1954; Jernstrom & Gowdy, 1975; Wile et al, 1981).
Recently, the authors have had experience of a primary leiomyosarcoma arising in the great saphenous vein with intraluminal growing and have reported this because no case of primary leiomyosarcoma within the great saphenous vein had been reported to date in Korean literatures.

CASE REPORT

The patient was a 54-year-old female who was admitted to thee Lee Rha General Hospital complaining of a slow growing, painless, palpable mass at the proximal one third of the left inner thigh. She had had the condition for three years. otherwise she was healthy and no other metastatic lesion was noted. Simple tumor excision was carried out under the impression of benign soft tissue tumor.

At operation, there was a round to ovoid, well encapsulated mass in the subcutaneous tissue attached to the cord-like fibrous tissue. No bleeding from the cord-like fibrous tissue was noted when the fibrous cord was cut. It was easily separated from surrounding soft tissue Simple mass excision was done.

The mass measured 5×3×3cm and had a rubbery and firm consistency. On sectioning, the tumor was pseudoencapsulated by glistening dense fibrous tissue, proved to be the original venous wall later. It was composed of grayish white soft tissue with lobular appearance (Fig. 1). The attached a 5cm long fibrous cord revealed no definite lumen.

Microscopically, the tumor consisted of interlacing and parallel bundles of spindle cells with a variable degree of anaplasia. In areas, tumor giant cells as well as normal and abnormal mitotic figures (over 5/10 HPF) were frequently observed (Fig. 2). The tumor grew into the vascular lumen (Fig. 3). Sections from the fibrous cord-like tissue revealed proliferation of basically the same spindle cells with hyalinization as the those of main tumor, suggesting a spread for some extent of the tumor along the lumen of the vein (Fig. 4). Masson's trichrome stain revealed reddish staining of the cytoplasm and reticulin stain revealed bundles of tumor cells surrounded by reticulum fibers seen as in usual smooth muscle tumors. Elastic stain (Victoria blue stain) revealed well preserved outer venous wall. Desmin stain was positive in the cytoplasm of spindle cells and a few tumor giant cells (Fig. 5). Ultrastructurally, tumor cells were poorly preserved due to previous

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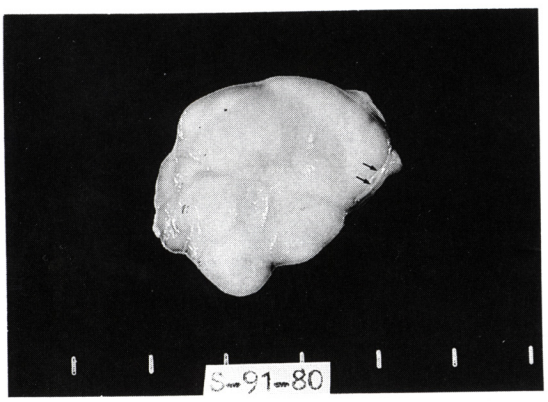

Fig. 1. Well circumscribed and pseudoencapsulated tumor within the veinous wall (arrow) with lobulation

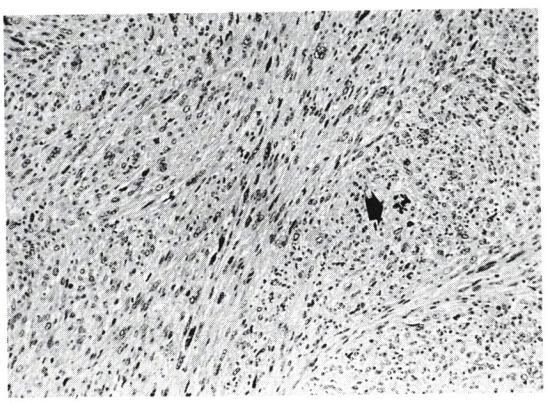

Fig. 2. Leiomyosarcoma cells showing cigar shaped nuclei with an atypical mitosis (arrow) (H-E, X100)

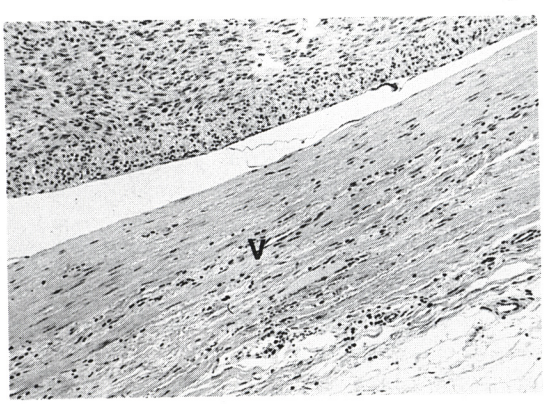

Fig. 3. Intraluminal growing of leiomyosarcoma, attached to the wall of the saphenous vein (V) (H-E, X100)

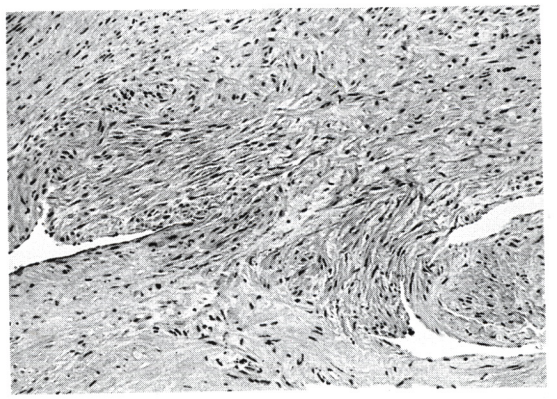

Fig. 4. Intraluminal obliteration by the tumor extended from the main tumor (H-E, X100)

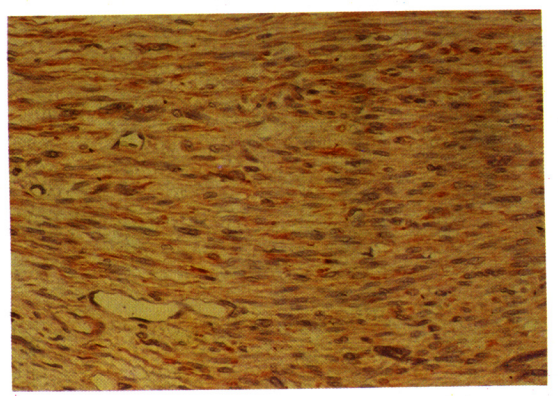

Fig. 5. Strong positive reaction of immunohistochemical staining for desmin in the tumor cells (Desmin stain, ×200)

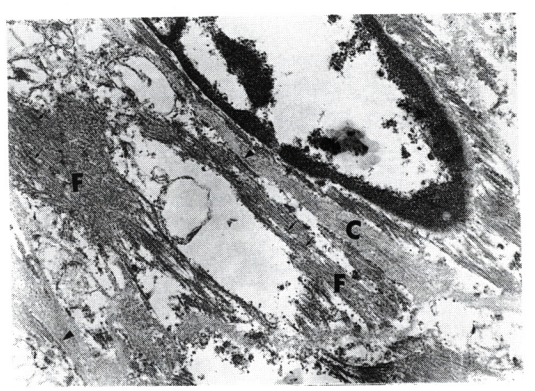

Fig. 6. Abundant myofilaments (F), dense bodies (thin arrows), basal lamina and vesicles in the cytoplasm of the tumor cells with intercellular collagen (C) (EM, ×20,000)

formalin fixation. However, evidence of smooth muscle cells such as abundant intracytoplasmic thin filaments with dense bodies, a few pinocytic vesicles and focal basal lamina were present. The interstitium contains collagen fibrils (Fig. 6).

DISCUSSION

Leiomyosarcoma of vascular origin comprises a rare group of tumors with fewer than 200 cases reported in the literatures since the initial report of a leiomyosarcoma of the midportion of the inferior vena cava by Perl in 1871 (Enzinger & Weiss, 1990). Leiomyosarcomas are most common in large veins such as the inferior vena cava, and are rare in arteries. In the extensive literature review about the 86 cases of primary leiomyosarcomas of vascular origin was done by Kevorkian and Cento (1973). They reported thirty three cases arose in the inferior vena cava, thirty five cases in other medium sized or large veins such as the femoral, saphenous and iliac veins. Ten cases in the pulmonary artery alone and eight cases in the systemic arteries.

The clinical manifestations related to these tumors are diverse and are determined by the location, rate of growth, and degree of collateral blood flow or drainage in the affected part (Brewster et al, 1976). Leiomyosarcomas arising in the inferior vena cava occur in mid-or late adult life with an average age in 6th decades with female predominancy (80%-90%). The location of the tumor within the vessel is significant because it determines the type of symptoms and surgical resectability. Leiomyosarcomas of other veins most often arise in the veins of the lower extremities, including the saphenous, iliac and femoral and affect both sexes equally. They usually present as mass lesions of variable duration, which occasionally produce lower leg edema. Leiomyosarcomas of the pulmonary artery are the most common form of arterial leiomyosarcoma, occurring in adults. Their symptoms are referable to decreased pulmonary outflow (Berlin et al, 1984; Enzinger & Weiss, 1990; Kevorkian & Cento, 1973; Shmookler et al, 1977).

The electron microscopic identification of myofilaments (60-80Å) can be helpful in confirming the diagnosis of leiomyosarcoma occurring in an uncommon location. Other smooth muscle characteristics such as pinocytotic vesicles, peripheral densities and plaque-like junctions with an intermediate dense line are inconsistent findings although their presence is suggestive of a smooth muscle neoplasm. A patchy distribution of cells containing myofilaments was a common finding in smooth muscle tumors (Mackay et al, 1987). Immunohistochemical staining for desmin is also useful for the differentiation of leiomyosarcoma from other sarcomas (Osborn, 1984).

Leiomyosarcomas of venous origin have been described as polypoid or nodular masses that are firmly attached to the vessel at some point and have spread for a variable extent along its surface, the histologic features of the tumors are basically similar to those in the retroperitoneum, although they have sometimes show marked vascular proliferation, which is applicable for the term "vascular leiomyosarcoma" in a narrow sense (Varela-Duran et al, 1979). Leiomyosarcoma arising in the vessel usually does not show prominent hemorrhage or necrosis. Mitoses are rather easy to identify in these tumors and the histological criteria of malignancy of soft tissue leiomyosarcoma are equally applicable to these lesions. Tumors having 5 mitoses/10HPF should be considered malignant. Tumors having between 1 and 4 mitoses/10HPF are best considered potentially malignant, especially if they are large and have areas of necrosis and significant nuclear atypia (Berlin et al, 1984; Enzinger & Weiss, 1990).

The morbidity and mortality in association with these tumors is primarily a result of direct extension of these tumors alongthe vessels with circulatory disturbances especially in the large vessels. Therefore, prognosis and results of therapy are variable depending on the site of origin and its resectability. Complete removal of the tumor is treatment of choice so far. In addition, its mestastatic or recurring potential is not predicatable. Berlin et al (1984) reported 6 cases of leiomyosarcoma of venous origin, which actually had metastasized at the time of surgery or from 1 to 22 months after surgery. Metastases occured mainly in the liver, lung, and less often in regional lymph nodes or the intraabdominal organs.

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