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Pedunculated Cutaneous Hemangiopericytoma

— A report of an unusual case —

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INTRODUCTION

Hemangiopericytoma is a rather uncommon neoplasm, first described and named by Stout and Murray[1] in 1942, however it has been well described over the past forty seven years with many case reports and review articles published on this subject. Nevertheless,there continues to be difficulties in the diagnosis and in the determining the prognosis of this tumor. We have observed a case of a large, benign hemangiopericytoma of long duration, in the skin of the lower abdomen.

CASE PRESENTATION

A 65-year-old woman was presented to our hospital, with a huge pedunculated slowly growing mass on the lower abdomen for about 30 years. On physical examination, this tumor was attached to the abdominal skin and subcutis with a short broad stalk(Fig. 1). Chest X-ray, E.K.G., and laboratory tests including C.B.C., urinalysis, liver function tests and serum protein electrophoresis, were unremarkable. After a complete excision of this mass, she has been well and without evidence of a recurrence of the tumor for 2 years.

Grossly, the tumor mass was a huge well circumscribed and partly lobulated firm mass, measuring 23×20×10 cm. It was covered by a thin pseudocapsule and showed a tan-brown, multinodular external surface. On section, it had a gray to tan brown,

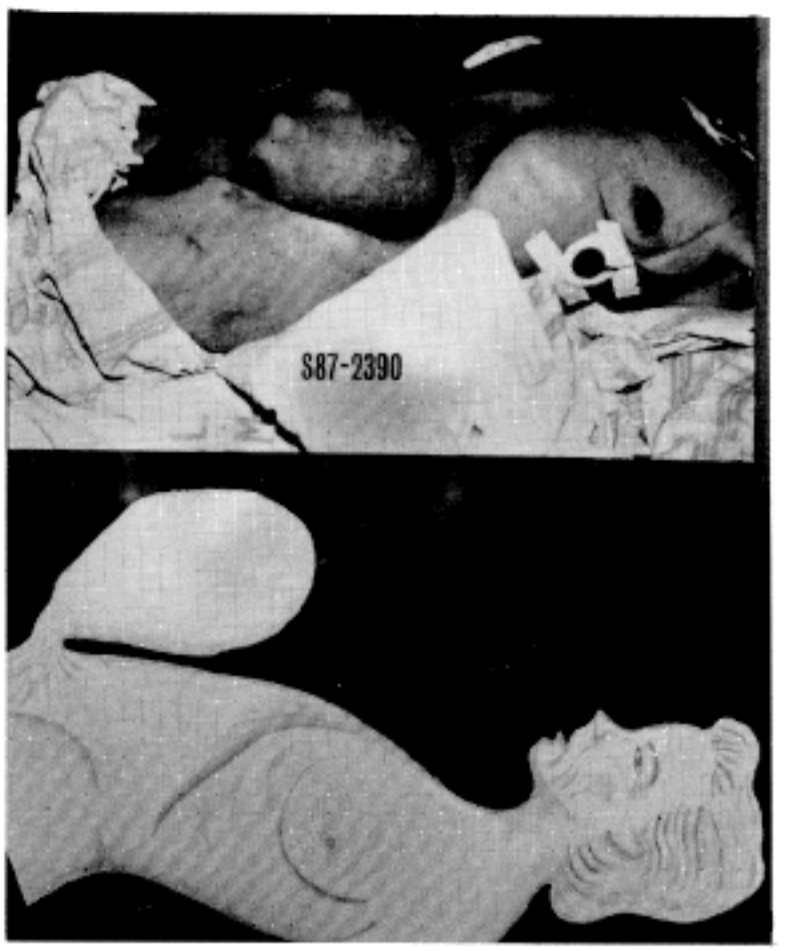

Fig. 1. A huge pedunculated tumor mass with a short broad stalk.

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rubbery, solid and multilobulated cut surface with some gelatinous and mucoid areas between lobules (Fig. 2).

Histologically, the tumor was richly vascular, and it was composed of tightly packed tumor cells around thin walled vascular channels, ranging from capillary-sized vessels to ramifying sinusoidal spaces (Fig. 3). The vascular spaces were frequently compressed and obscured by the surrounding tumor cells. The tumor cells were spindle-shaped with elongated or oval nuclei and indistinct cytoplasmic borders. In some areas, the cells were arranged in a storiform pattern (Fig. 4). There were some areas of myxoid change and fibrosis. Perivascular hyalinization was also seen. Mitoses were rare and cellular anaplasia was not apparent. A reticulin stained preparation showed dense reticulin fibers surrounding collapsed or dilated vascular channels, and individual tumor cells (Fig. 5). Based on the above pathologic findings, we diagnosed this tumor as a hemangiopericytoma.

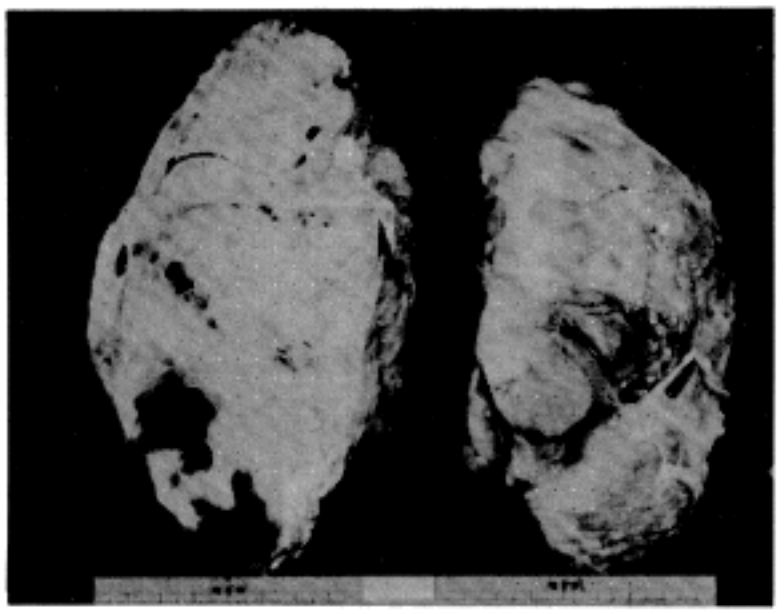

Fig. 2. A well circumscribed and partly lobulated firm mass.

DISCUSSION

Hemangiopericytoma is a rather uncommon mesenchymal neoplasm, first described by Stout and Murray[1] in 1942. The hemangiopericytoma is considered to be a tumor of pericytes[2-7]. Since all capillaries are enveloped by pericytes, hemangiopericytomas can arise anywhere in the body[3,4,8,10]. The majority of these tumors occur in the lower extremities, pelvis, head and neck regions, and trunk, in that order of frequency[4,10,11]. This tumor affect both

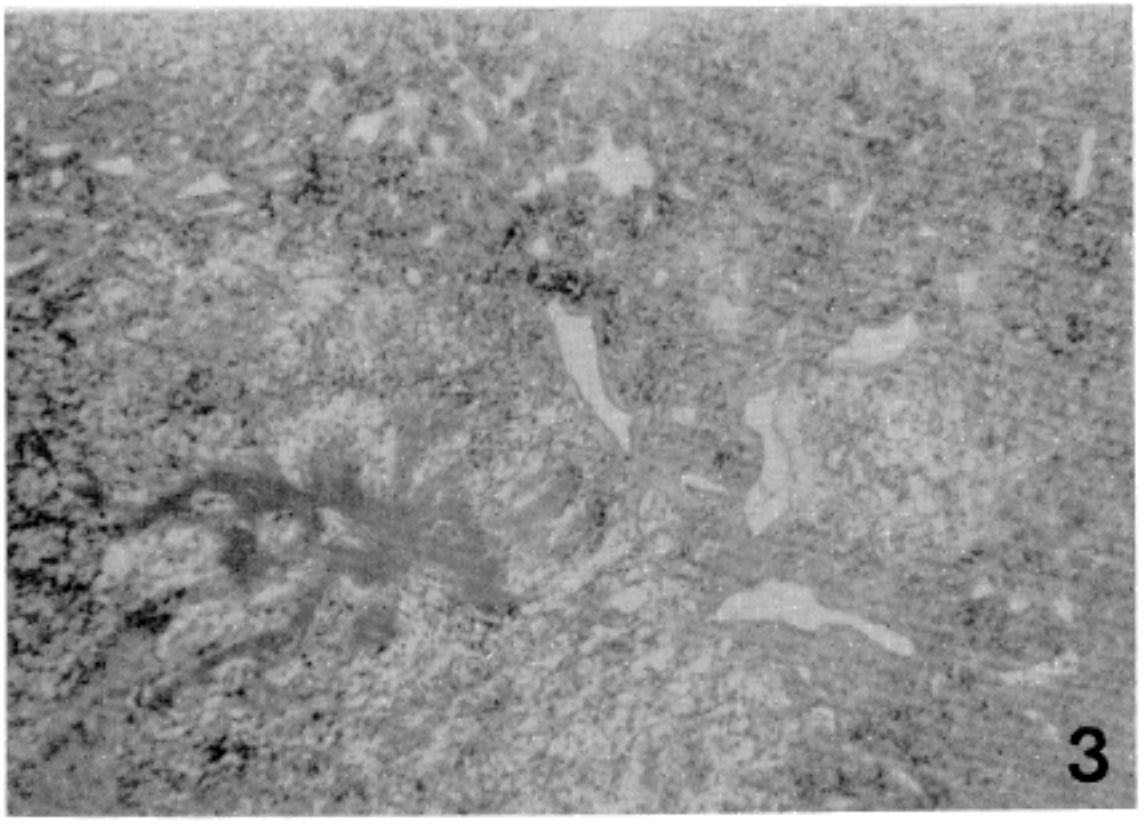

Fig. 3. Richly vascular pattern consisting of anastomosing vessels of varying calibers (H&E, × 40).

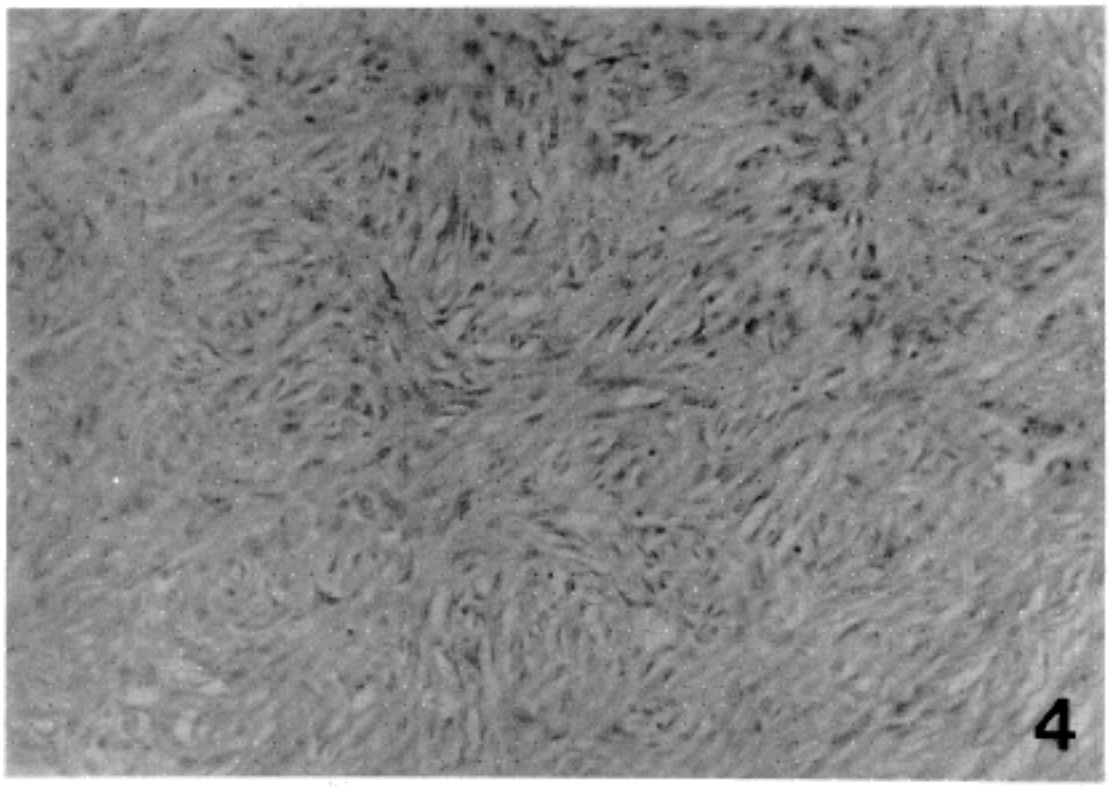

Fig. 4. Area showing a distinct storiform pattern (H&E, ×100).

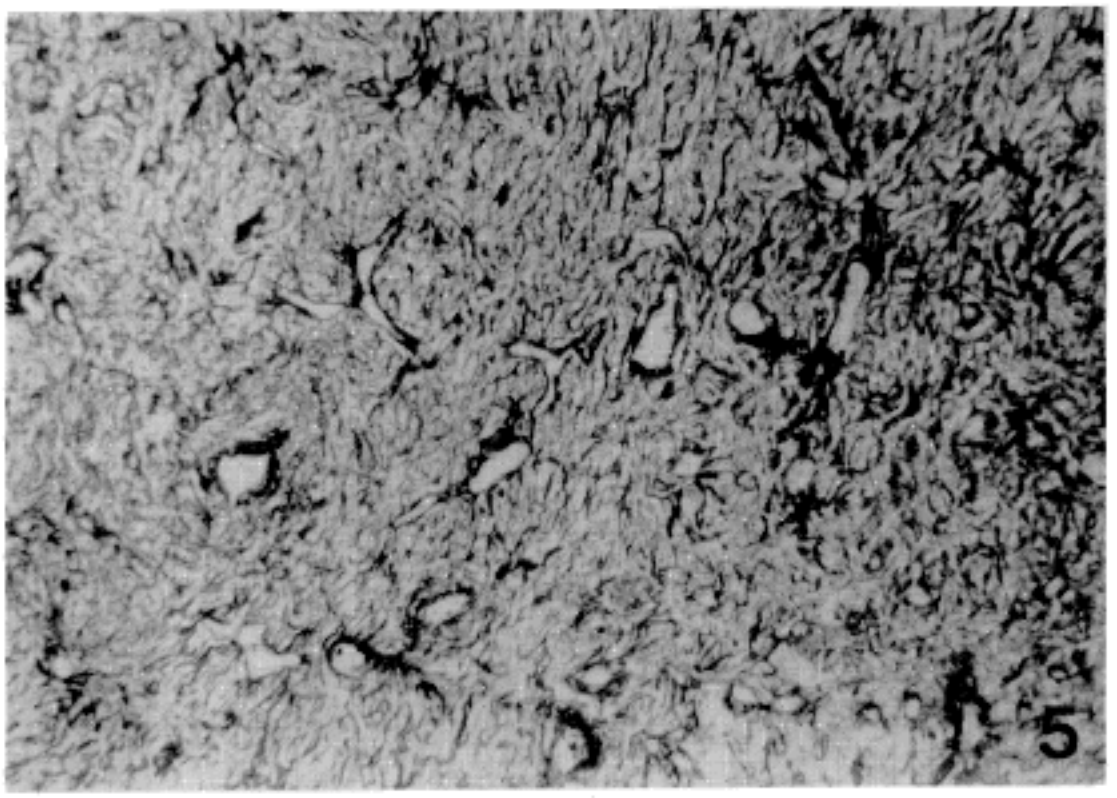

Fig. 5. Dense reticulin fibers surrounding vessels and tumor cells (Reticulin stain, ×40).

sexes in equal incidence and occurs chiefly in adults[3,10,11]. Hemangiopericytoma often occurs as a slowly enlarging painless mass, which shows variable symptoms according to its location[3,8,10,11].

Grossly, this tumor usually presents as a solitary fairly well circumscribed mass. The size of the tumor varies from 0.8 cm to 21 cm in greatest dimension with a median of 6.5 cm[11]. On section, they show a solid homogeneous appearence with soft but often hard, or rubbery consistency[8,10,11]. The color of cut surfaces are gray-white, pink-white, tan, or red-brown[8,10,11].

On histologic examination, this tumor shows a characteristic vascular pattern. The vascular spaces are lined by a single layer of flattened endothelium. These spaces are variable in size, ranging from

capillaries to large sinusoidal spaces. These vascular spaces are interconnecting in a ramifying configuration, compressed, or even obliterated[10,11]. The tumor cells show oval to spindle-shaped nucleus and ill-defined cytoplasmic borders. Mitoses are rare or absent. The tumor cells are arranged in compact sheets around the thin-walled vascular channels. The tumor cells are spindle-shaped and show a storiform pattern in some areas. Occasionally, focal degenerated areas of a tumor will show myxoid change resembling myxoid liposarcoma, but lipoblasts and a plexiform capillary pattern are never seen in these myxoid areas[10,11].

The differential diagnosis includes richly vascular mesenchymal neoplasm such as infantile hemangiopericytoma, glomus tumor, fibrohistiocytoma, and other sarcomas with hemangiopericytoma-like patterns (extraskeletal mesenchymal chondrosarcoma, infantile fibrosarcoma, synovial sarcoma, etc.)[10~12].

Most hemangiopericytomas have a benign course. However, histologic findings, such as a high mitotic rate, increased cellularity, and the presence of necrosis and hemorrhage, indicate a neoplasm that may have a more aggressive biologic potential[10,11].

Wide surgical excision is the treatment of choice for hemangiopericytoma[3,4,9,10,11,13]. Because of local aggressiveness and the infiltrative growth pattern of this neoplasm, the recurrence rate is relatively high in all series (25 to 50 per cent). Therefore, postoperative irradiation has been suggested by some workers[13,14]. Indeed combined surgical and radiation theraphy yields better local tumor control than surgery alone[13,15].

In our case, a 65-year-old woman had a huge, slowly growing, painless, pedunculated mass, located in the subcutis on the lower abdomen for about 30 years. The size of this tumor was 23 cm in the greatest dimension.

Through our review of the literature, our case of hemangiopericytoma was thought to be unusual due to its large size and long duration. In addition, in light of the above, it is noteworthy that our case is apparently benign as our patient has been well and without evidence of recurrence for two years.

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— 국문초록 —

혈관외피종

— 1증례 보고 —

가톨릭대학 의학부 임상병리학교실

강유미 · 지미경 · 양기화
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정 수 일

혈관외피종은 1942년 Stout와 Murray에 의해 처음 기술되었으며 혈관주의 세포에서 기원한 간엽성 종양으로 생각되어진다. 조직학적으로 특징적인 혈관양상을 보이며 이와 유사한 혈관양상을 보이는 다른 종양들과의 감별이 필요하다. 이 종양은 국소재발율이 높으며 방사선 치료가 도움이 된다고 한다. 이 증례는 65세 여자의 하복부 피부에서 발생하여 30년 동안 서서히 자란 장경 23cm 크기의 종괴로 가느다란 유경부에 의하여 하복부 피부 및 피하조직과 연결되어 있었다. 절제된 종괴는 육안적으로 피막에 의해 둘러싸여 있었고 외면은 다갈색의 다소 돌출된 크고 작은 결절들이 관찰되었다. 단면상 회갈색의 고무양 경도를 보이는 고형성 부위가 다소 다엽성 구조를 이루고 있었고 이러한 다엽성 구조 사이로 젤라틴양 점액성 부위가 관찰되었다. 현미경적으로 종양은 모세혈관 크기에서 커다란 sinusoid까지 다양한 크기의 혈관들이 풍부하게 관찰되었고 종양세포들은 혈관주위에 특별한 양상없이 밀집되어 있었다. 종양은 미만적 또는 국소적으로 섬유화된 부분이 관찰되었고 혈관주변에 초자양 변화도 보였다. 종양의 변성된 부위에서는 점액성 지방육종과 유사한 점액성 변화가 관찰되었으나 지방아세포 또는 총상(plexiform) 모세혈관 양상은 보이지 않았다.

본 증례는 종양절제후 약 2년간 재발을 보이지 않았다.

Key Words: Hemangiopericytoma, Pedunculated skin tumor