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전완 원위부 심 수지 굴근에 발생한 선천성 혈관 외피세포종 - 증례 보고 -

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혈관 외피 세포종은 드문 악성 혈관 종양으로 대부분 성인에서 발생한다. 영아에서 발생하는 경우는 선천성 또는 영아형 혈관 주변 세포종이라 불리우며, 아주 드물게 발생하고 성인형에 비해 양호한 예후를 보이지만 재발을 막기 위해서는 충분한 절제연을 가지고 절제 되어져야 한다. 본 증례는 출생후 1일째 내원하여, 65일째 광범위 절제술을 시행한 좌측 전완부의 혈관 외피 세포종의 증례를 보고하고자 한다.

색인 단어: 전완부, 선천성 혈관 외피 세포종

INTRODUCTION

Hemangiopericytoma is an uncommon vascular tumor originally described by Stout and Murray in 1942[10]. This neoplasm accounts for less than 1% of all vascular tumors. The majority of such tumors are acquired or adult type, but approximately 10% of all cases occur in children with the congenital form comprising from 3.3% to 7% of all hemangiopericytoma, and 0.08% of all pediatric tumors[7]. Because hemangiopericytomas are a tumor of blood-vessel origin, they may occur anywhere in the body. Approximately 30~50% of congenital or infantile hemangiopericytoma occur in the extremities[3,5]. About 50 cases of infantile or congenital hemagiopericytoma have been reported in the literature to date[2].

Congenital or infantile hemangiopericytoma is generally and clinically more benign than adult type, and the occurrence of spontaneous tumor regression has supported a conservative approach in the treatment of congenital hemangiopericytoma[4]. However, metastasis and local invasion have recently been described[6,9]. We describe a neonate who presented with a huge mass in the volar aspect of left distal forearm. The mass was confirmed on biopsy as congenital hema-

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giopericytoma.

CASE REPORT

A male infant weighing 3560 g was born in February 2002 at 40 weeks' gestation. A firm tumor was noted in the volar asepect of left distal forearm at birth. At 3 days of age, the infant was referred to our department for an evaluation of the tumor. On physical examination, a firm, smooth-surfaced with sharp margin 4×5×3 cm occupied the volar asepect of left distal forearm. The overlying skin was normal and no local heat was noted. The laboratory data showed normal blood findings, including normal serum lactic dehydrogenase. A chest x-ray was normal. X-ray films of the forearm showed a soft tissue mass. Ultrasonography revealed mixed solid and cystic heterogenous echoic mass in the flexor digitorium longus muscle (Fig. 1). A needle biopsy showed that the tumor consisted of tightly packed cells around ramifying thin walled, endothelium-lined sinusoidal vascular channels. Magnetic resonance imaging (MRI) performed at 65 days of age, T2 weighted sagittal (Fig. 2A) and T2 weighted axial (Fig. 2B) image showed a solid, well-demarcated mass with a relatively high signal intensity compared to the muscle tissue, but without calcification. Tumor containing septae on the volar asepect of left distal forearm were observed. There was irregular enhancement after intravenous contrast was injected (Fig. 3A, B). At 65 days of age, a wide resection was performed. Grossly, the cut surface showed

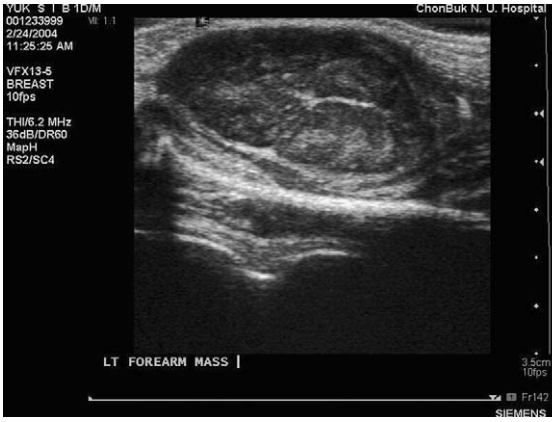

Fig. 1. Ultrasonography revealed mixed solid and cystic heterogenous echoic mass in the flexor digitorium longus muscle.

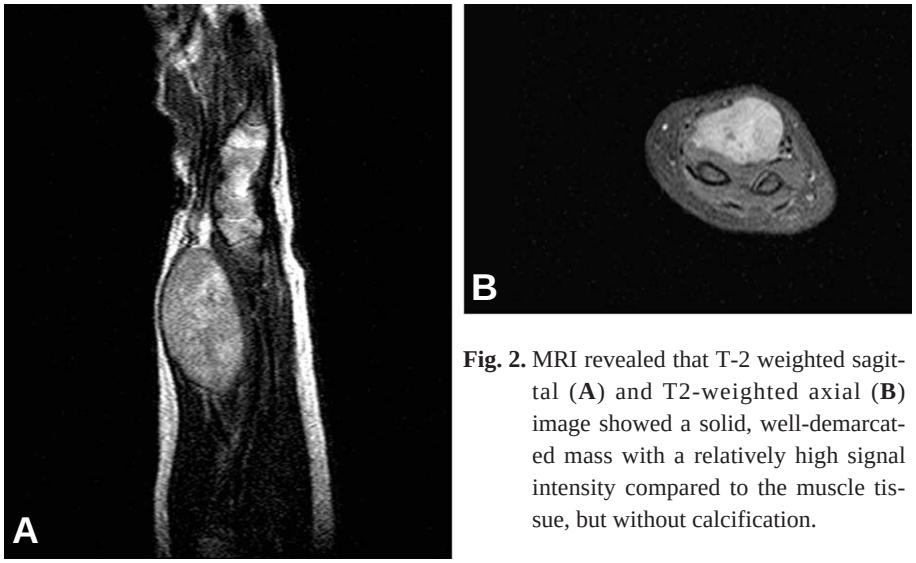

Fig. 2. MRI revealed that T-2 weighted sagittal (**A**) and T2-weighted axial (**B**) image showed a solid, well-demarcated mass with a relatively high signal intensity compared to the muscle tissue, but without calcification.

2.3×1.5 cm sized relatively well circumscribed glistering yellowish gray mass with focal hemorrhage in the skeletal muscle (Fig. 4). Histologically, the tumor showed continuous ramifying vascular pattern in association with a relatively bland mesenchymal cells. Tumor cells were tightly packed round to fusiform cells with indistinct cytoplasmic borders and arranged around the vessels (Fig. 5). The mean mitotic rate was 2 mitosis per 10 HPF. Atypical mitosis or significant cytologic atypia was not seen. Immunohistochemically tumor cells were strongly positive for CD34 and focal positive for smooth muscle actin (Fig. 6). No immunoreactivity was detected

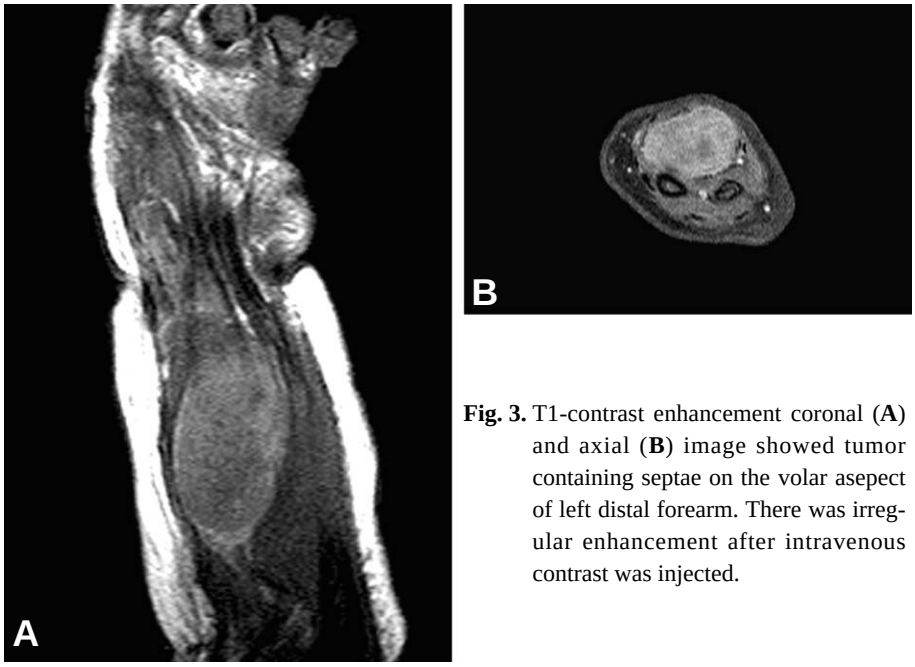

Fig. 3. T1-contrast enhancement coronal (**A**) and axial (**B**) image showed tumor containing septae on the volar asepect of left distal forearm. There was irregular enhancement after intravenous contrast was injected.

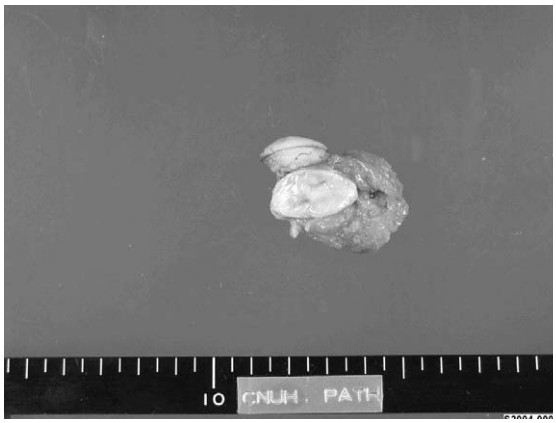

Fig. 4. The cut surface shows relatively well circumscribed glistering yellowish gray mass in the skeletal muscle.

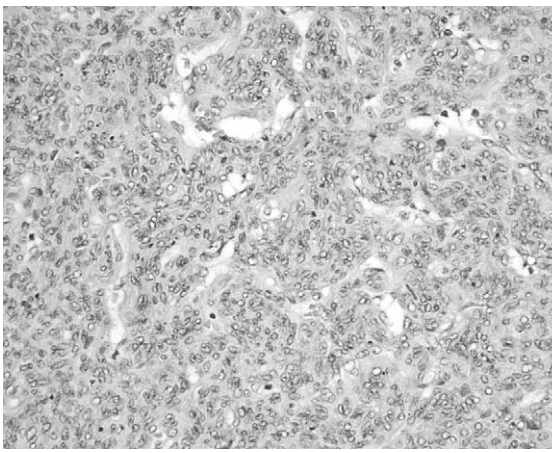

Fig. 5. Tightly packed round to fusiform tumor cells are arranged around the ramifying Vessels.

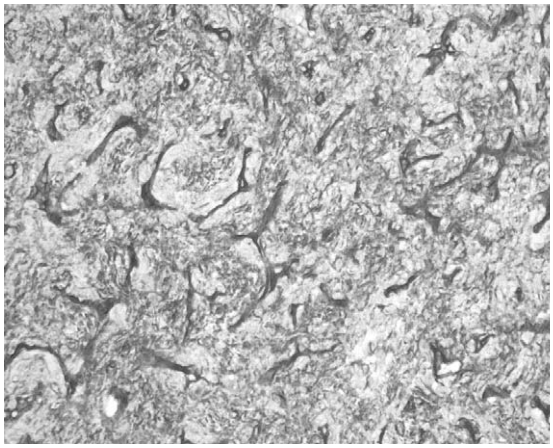

Fig. 6. Tumor cells are strongly immunoreactive for CD34.

for cytokeratin, epithelial membrane antigen, factor VIII related antigen, CD31, desmin, S-100 protein, and CD68.

The overall appearance was consistent with a diagnosis of congenital hemangiopericytima. The patient has shown normal finge function and remains well with no recurrence for 3 years postoperatively.

DISCUSSION

In 1960, Kauffman and Stout[8] made the differentiation between the adult and infantile forms of tumor as they noted the more benign outcome of tumors that arose in children younger than 1 year. Baker[5] in 1992 reviewed the literature and identified 58 cases of infantile hemangiopericytoma that had been reported. Approximately 10% of all hemagiopericytoma that occur are of the infantile form, which Kauffman and Stout defined as tumors occurring in children younger than 1 year. Unlike the adult for, which affects both sexes equally, congenital forms tend to occur more frequently boys. Approximately one third are located in the extremities. Another one third to one half occur in the head and neck region, and one third in the trunk. There have been case reports of patients with multiple region[8,9]. Whether this represents multifocal disease or metastases is unclear. Tumor in the trunk and extremities of infants are usually located in the subcutaneous tissue, differing form the adult form, which arises from deeper structures. They are painless, firm, rubbery masses. There is seldom discoloration in the extremity lesion, as compared with those that are in the oral and nasal region, which often take on a bluish hue.

Microscopically, congenital form bears a close rsemblance to adult form, but many lesions, especially superficial ones, are multilobulated, often with distinct intravascular and perivascular satellite nedules outside the main tumor mass and frequent endvascular growth. There is often increased mitotic figures that indicate a poor prognosis for adult type but generally do not with infantile form. Most of congenital or infantile forms tend to follow benign clinical course, they are curable by local excision or may regress spontaneously. In rare instance, however, there may be local infiltrative growth or recurrence and even metastasis. Deep seated lesions and those occurring in older children seem to pursue a more aggressive clinical course than superficial ones that appear during the first years of life[1]. The heangiopericytoma-like pattern found in the lobular or tufted hemangioma, infantile myofibromatosis, and infantile fibrosarcoma must be distinguished from that of infantile hemangiopericytoma. Some authors believe that many of these lesions of infantile hemagiopericutoma belong to the infantile myofibromatosis group.

The biologic behavior of congenital hemangiopericytoma is unpredictable, and there is

no clinical criteria available to predict a spontaneous regression or a sufficiently safe tumor regression rate to adopt a wait-and-see policy. So far, most accepted management of congenital or infantile hemangiopericytoma is wide resection of the lesion whenever possible. Metastasis or local recurrence may occur after several year of disease-free intervals, and therefore, close and long-term observation is necessary. In cases where recurrence or metastasis does occur, chemotherapy has been used with varying degrees of success, and radiation therapy also has been used with success when either microscopic or gross tumor tissue remains.

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Abstract

Congenital Hemagiopericytoma in the Flexor Digitorium Profundus Muscle of the Distal Forearm - A Case Report -

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Hemangiopericytoma is a rare malignant vascular tumor that usually occurs in adults. The occurrence of these tumors in infants, known as congenital or infantile hemangiopericytoma, is even rare and their behavior may be more benign than the adult type. Despite of the generally good prognosis associated with this neoplasm, a complete surgical excision has so far been recommended to avoid recurrence, because no definite criteria for determining whether or not the tumor will regress spontaneously have been established to date. We describe a 1-day-old male neonate with congenital hemangiopericytoma, presenting with a left forearm mass at birth. Wide resection was performed at 65 days of age and hemangiopericytoma was diagnosed by histology. There was no tumor recurrence during 32 months of follow-up.

Key Words: Forearm, Congenital hemagiopericytoma

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