Original Paper

Pediatric Neurosurgery

Pediatr Neurosurg 2000;33:26–30

Received: February 1, 2000
Accepted: June 26, 2000

Endovascular Treatment of Huge Cervicofacial Hemangioma Complicated by Kasabach-Merritt Syndrome

Masaki Komiyama[a] Hideki Nakajima[a] Shouhei Kitano[b] Hiroaki Sakamoto[b]
Hiroko Kurimasa[c] Hajime Ozaki[c]

Departments of [a]Neurosurgery, [b]Pediatric Neurosurgery and [c]Pediatrics, Osaka City General Hospital, Osaka, Japan

Key Words
Embolization · Hemangioma · Kasabach-Merritt syndrome · Thrombocytopenia

Abstract
A 2-month-old girl with a huge cervicofacial hemangioma complicated by Kasabach-Merritt syndrome was presented. Two weeks corticosteroid treatment was ineffective, and deterioration of coagulation parameters and compromise of the airway and left vision due to huge hemangioma prompted use of endovascular treatment. Transfemoral arterial embolization using polyvinyl alcohol particles resulted in rapid clinical improvement. Endovascular treatment should be considered as an important therapeutic option for hemangioma complicated by Kasabach-Merritt syndrome.

Introduction

Most hemangiomas in infancy are clinically benign and involute spontaneously during childhood, but some occasionally cause thrombocytopenia, a microangiopathic hemolytic anemia, and a consumption coagulopathy, which was first reported by H.H. Kasabach, a radiologist, and K.K. Merritt, a pediatrician, in 1940 [1]. There have been many therapeutic methods for hemangioma complicated by Kasabach-Merritt syndrome with unpredictable success rates. Among them, there have been few reports on endovascular treatment for hemangioma with Kasabach-Merritt syndrome [2–8]. We present an infant with cervicofacial hemangioma complicated by Kasabach-Merritt syndrome successfully treated by transarterial particulate embolization.

Case Presentation

A 2-month-old girl without remarkable birth and family history presented with slight swelling in the left preauricular region. The patient was born at a gestation period of 41 weeks with a birth weight of 3,664 g. Since the swelling continued to increase and became harder in the following several days, the patient was referred to our hospital. On admission, the patient was alert without infectious signs. Body weight was 5.5 kg and height was 59.5 cm. A hard swelling of the left temporal and buccal regions with petechiae was observed. Laboratory data on admission were white blood cells 9,210/mm^3, hemoglobin 10.2 g/dl, platelet count 44,000/mm^3, fibrinogen 100 mg/dl (normal 150–350 mg/dl), fibrin degradation products value 17 μg/ml (normal <5 μg/ml) and C-reactive protein 0.0 mg/dl. Clini-

KARGER
Fax + 41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

1016–2291/00/0331–0026$17.50/0

Accessible online at:
www.karger.com/journals/pne

Dr. Masaki Komiyama, MD
Department of Neurosurgery, Osaka City General Hospital
2-13-22, Miyakojima-Hondouri
Miyakojima, Osaka 534-0021 (Japan)
Tel. +81 6 6929 1221, Fax +81 6 6929 1091, E-Mail komiyama@japan-mail.com

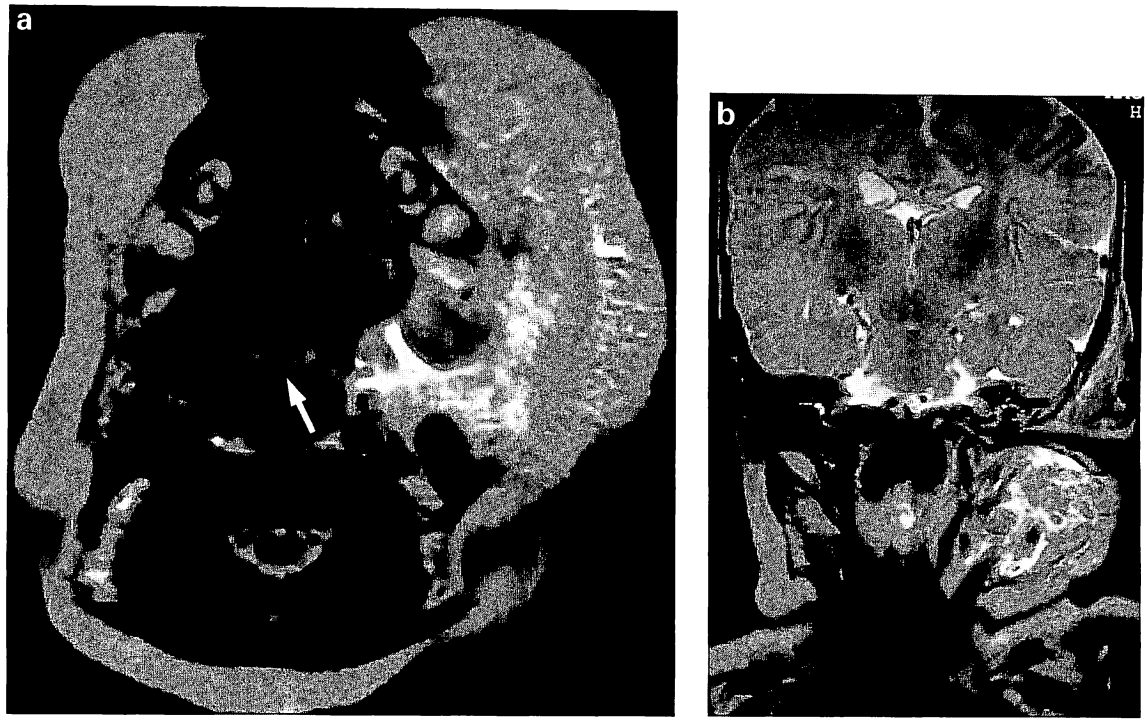

Fig. 1. T_2-weighted magnetic resonance images showing a large mass in the left temporal, buccal and cervical regions. Tracheal deviation (arrow) is observed. **a** Axial image. **b** Coronal image.

cal picture and coagulation parameters were consistent with a diagnosis of Kasabach-Merritt syndrome. Computed tomography and magnetic resonance imaging showed huge hemangioma in the left cervicofacial region (fig. 1). There was no bony involvement.

Systemic prednisolone therapy started at the dose of 2.0 mg/kg/day since admission. In the following 2 weeks, the swelling with ecchymoses increased to such a degree that the left eye could not open, and dyspnea/tachypnea occurred due to the tracheal compression, which required endotracheal intubation. The skin color over the tumor changed to blue and/or brown due to subcutaneous hemorrhage (fig. 2). The coagulation system further deteriorated. Laboratory data on day 14 (1 day before embolization) were as follows: hemoglobin 11.1 g/dl, platelet count 23,000/mm^3, fibrinogen <40 mg/dl, fibrinogen degradation products value 25.4 μg/ml, antithrombin III 29.0 mg/dl (normal 23.6–33.5 mg/dl), prothrombin time 14.4 s (normal 10.0–13.0 s) and activated partial thromboplastin time 40.6 s (normal 25.0–37.0 s). These were consistent with a diagnosis of disseminated intravascular coagulation.

Under general anesthesia, the diagnostic and therapeutic angiography was carried out through the femoral route on day 15. A 4-french introducing vascular sheath was inserted into the right femoral artery and a 4-french catheter with a hockey-stick tip configuration was used for diagnostic angiography. Left carotid angiogram showed no contribution from the internal carotid system to the tumor, but the left facial artery, superficial temporal artery and posterior auricular artery contributed to the hypervascular tumor (fig. 3a, b). There were no arteriovenous shunts in the tumor, but fine

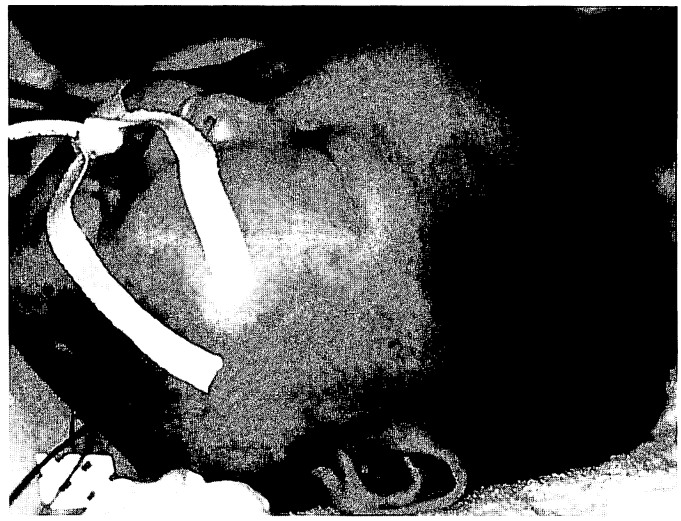

Fig. 2. Photograph immediately before embolization showing the marked swelling of the left cervicofacial region. Ecchymoses are also prominent.

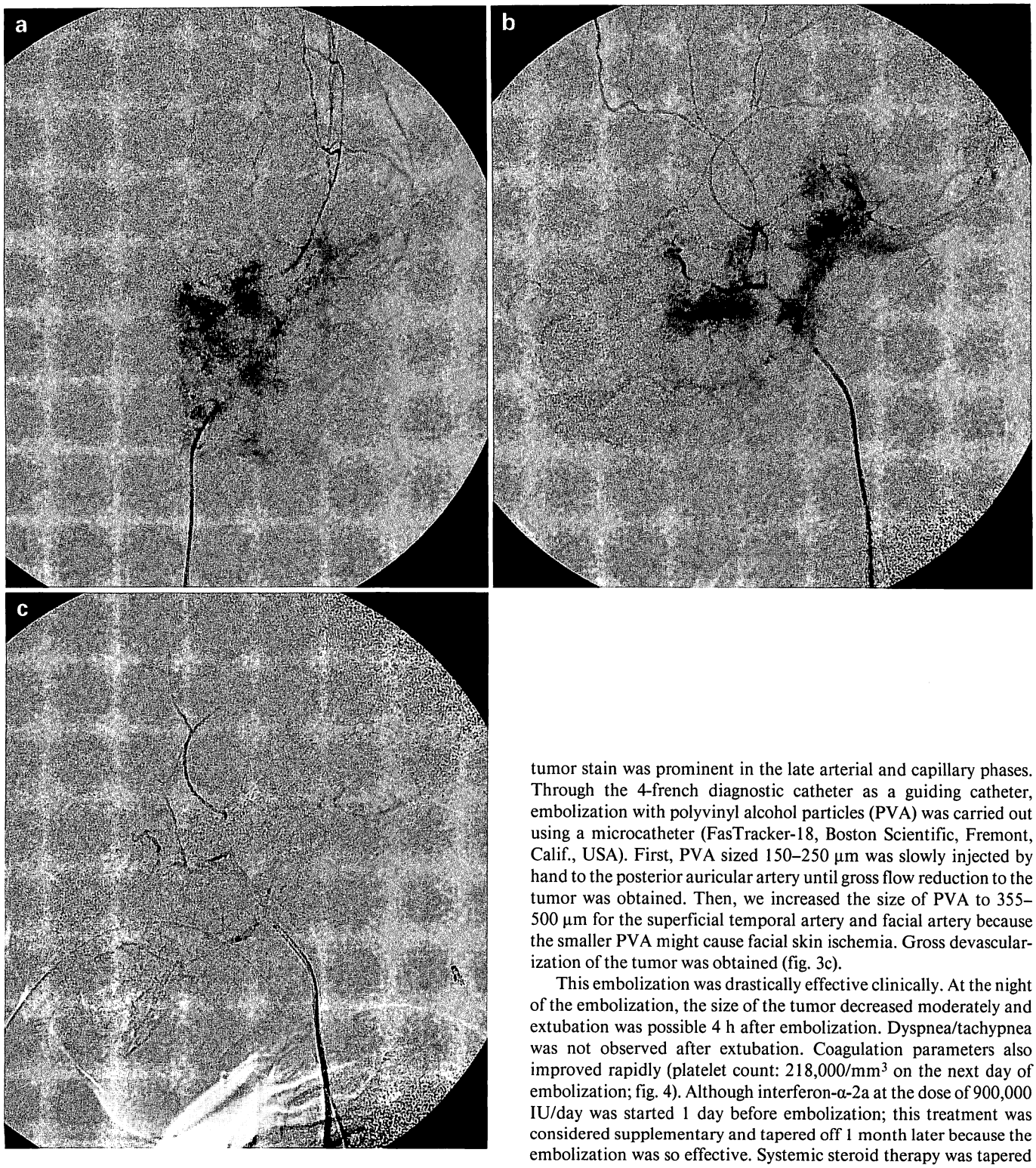

Fig. 3. Left external carotid injection (before embolization) showing fine tumor stain without arteriovenous shunts. **a** Frontal view. **b** Lateral view, late arterial phase. **c** Postembolization angiogram (lateral view, late arterial phase) showing gross devascularization of the tumor.

tumor stain was prominent in the late arterial and capillary phases. Through the 4-french diagnostic catheter as a guiding catheter, embolization with polyvinyl alcohol particles (PVA) was carried out using a microcatheter (FasTracker-18, Boston Scientific, Fremont, Calif., USA). First, PVA sized 150–250 μm was slowly injected by hand to the posterior auricular artery until gross flow reduction to the tumor was obtained. Then, we increased the size of PVA to 355–500 μm for the superficial temporal artery and facial artery because the smaller PVA might cause facial skin ischemia. Gross devascularization of the tumor was obtained (fig. 3c).

This embolization was drastically effective clinically. At the night of the embolization, the size of the tumor decreased moderately and extubation was possible 4 h after embolization. Dyspnea/tachypnea was not observed after extubation. Coagulation parameters also improved rapidly (platelet count: 218,000/mm^3 on the next day of embolization; fig. 4). Although interferon-α-2a at the dose of 900,000 IU/day was started 1 day before embolization; this treatment was considered supplementary and tapered off 1 month later because the embolization was so effective. Systemic steroid therapy was tapered off 1 week after embolization. The patient's cosmetic appearance improved drastically following the embolization. The skin color over the tumor became normal, and no apparent swelling was observed 2 weeks after embolization. The patient was discharged on day 54. She was stable hematologically and asymptomatic for 2 years and 10 months up to the last follow-up.

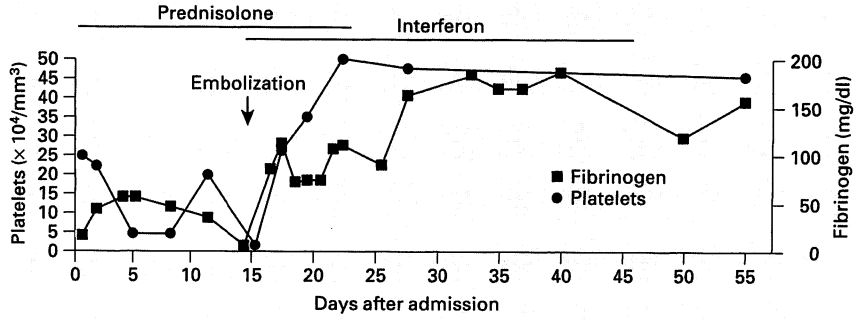

Fig. 4. Chronological changes of the coagulation parameters.

Discussion

Hemangioma is usually benign because involution begins before 1 year, reaches its peak at 2 years and gradually diminishes thereafter [9]. However, some hemangiomas become life-threatening and are called 'alarming hemangioma' because of airway obstruction, high output heart failure and consumption coagulopathy, i.e. Kasabach-Merritt syndrome. In general, the mortality of hemangioma complicated by Kasabach-Merritt syndrome is reported to be 12–24% due to hemorrhage, infection, invasion of vital structures and multiple organ failure [10–12].

It is reported that hemangioma in infancy causing Kasabach-Merritt syndrome is either kaposiform hemangioendothelioma or a neonatal form of tufted angioma, which should be discriminated from classic capillary hemangioma of infancy [11, 12]. Several investigators think that tufted angioma is part of a spectrum of kaposiform hemangioendothelioma [11, 12]. The classic hemangioma is differentiated by clinical presentation, magnetic resonance imaging and level of growth factors. Magnetic resonance imaging demonstrates kaposiform hemangioendothelioma showing diffuse enhancement with ill-defined margins, cutaneous thickening, stranding of subcutaneous fat, hemosiderin deposits and small feeding/draining vessels, while common hemangioma shows a well-defined, homogeneous, enhancing, soft-tissue mass, with fast-flow vessels within and around the tumor [12]. Kaposiform hemangioendothelioma shows low urinary levels of the basic fibroblast growth factor and vascular endothelial growth factor, which is contrasted to the common hemangioma [9].

Pathogenesis of hematological disorder in the hemangioma complicated by Kasabach-Merritt syndrome is generally considered due to intralesional trapping of platelets and fibrinogen, and activation of coagulation pathways within the tumor, resulting in localized intravascular coagulopathy [13, 14]. Furthermore, localized intravascular coagulopathy may extend to disseminated intravascular coagulation. Laboratory data show reduced platelets and fibrinogen, and reduced factors II, V and VII [10]. Prothrombin time and activated partial thromboplastin time are either prolonged or within normal limits in Kasabach-Merritt syndrome.

There are several therapeutic options in the treatment of hemangioma with Kasabach-Merritt syndrome, which include platelet transfusion, corticosteroids, cryoprecipitate, heparin, ε-aminocaproic acid, dipyridamole, aspirin, cyclophosphamide, interferon, irradiation, local compression, surgical resection, laser therapy, cryosurgery and embolization [1–8, 10–17]. The purposes of the treatment are to control coagulopathy and to eradicate or reduce the size of the lesions. Interestingly, one therapeutic method does not always provide the same effects on the hemangioma even in the same individual [5, 7, 10, 11]. Thus, the treatment should be individualized, and a plan of sequential treatment progressing from simple measures to more complex therapy is followed [3, 16].

Angiography in Kasabach-Merritt syndrome is occasionally problematic due to hemorrhage from the arterial puncture site [15], but the case of Stanley et al. [3], the cases of Weber et al. [16] and our case had no hemostatic problem after angiography. It is uncertain that all hemangiomas complicated by Kasabach-Merritt syndrome have tumor stain and/or feeding arteries that can be embolized. Although the number of the reported cases is small, they seemed to have tumor stain and feeding arteries. Thus, we believe that the majority of hemangiomas complicated by Kasabach-Merritt syndrome are candidates for transarterial embolization.

Transarterial embolization has been applied to several patients with a variety of success rates. Multiple sessions

were required in some cases, but in the patient of Stanley et al. [3] and in our patient, only one procedure provided dramatic improvement in the clinical symptoms and hematological parameters. Embolic materials included microfibrillar collagen hemostat [5], a water-insoluble plant amino acid [5], gelatin pellets [2], absolute alcohol [8] and PVA particles [3, 4, 6]. In Kasabach-Merritt syndrome, localized intravascular coagulation occurs in the vascular space of the hemangioma, and no increased coagulopathy is seen in the other parts of the body. In this sense, it is reasonable that reduction of the inflow to the hemangioma by selective transarterial embolization reduces the localized coagulopathy effectively. In fact, size reduction of the hemangioma and dramatic improvement of hematological profiles followed embolization in our case. Some cases required embolization in several sessions [2] while others did not. This seems to depend on the aggressiveness of embolization (degree of devascularization) and penetration of the embolic materials into the tumor. We believe that embolization should be aggressive when performed in the severely ill patients who require immediate clinical improvement. Transarterial embolization could be a preoperative procedure to reduce intraoperative bleeding, but in the thrombocytopenic state, surgical extirpation of the hemangioma carries a potentially high risk.

The reasons for the dramatic clinical improvement in our case are not clear, but there were several factors such as (1) timing of embolization, (2) degree of devascularization, (3) vascular anatomy and (4) embolic materials. After the failure of steroid treatment, we proceeded to embolization. At the time of embolization, systemic coagulopathy might still be reversible if proper treatment is given. As discussed above, local coagulopathy in the tumor was markedly increased. Embolization should minimize the blood flow to the tumor if locally increased coagulopathy in the lesion is isolated from the rest of the body. We embolized the arterial pedicles of the posterior auricular artery, superficial temporal artery and facial artery. These arteries were selectively catheterized with a microcatheter, which enabled selective injection of the PVA with calibrated size. For fear of skin necrosis, we used mainly the PVA sized 355–500 μm. Smaller sized PVA could be used, but we believe that control of the blood flow to the tumor is more important than penetration of the PVA deep into the tumor for the control of the localized intravascular coagulopathy.

In conclusion, we believe that transarterial embolization of the hemangioma complicated by Kasabach-Merritt syndrome should be considered as one of the therapeutic options, especially when the patient requires urgent clinical improvement.

References

1 Kasabach HH, Merritt KK: Capillary hemangioma with extensive purpura: Report of a case. Am J Dis Child 1940;59:1063–1070.

2 Argenta LC, Bishop E, Cho KJ, Andrews AF, Coran AG: Complete resolution of life-threatening hemangioma by embolization and corticosteroids. Plast Reconstr Surg 1982;70:739–742.

3 Stanley P, Gomperts E, Woolley MM: Kasabach-Merritt syndrome treated by therapeutic embolization with polyvinyl alcohol. Am J Pediatr Hematol Oncol 1986;8:308–311.

4 Eurvilaichit C, Kraiphibul P, Nontasut S: Kasabach-Merritt syndrome treated by transarterial embolization and radiotherapy. J Med Assoc Thai 1987;70:431–435.

5 Larsen EC, Zinkham WH, Eggleston JC, Zitelli BJ: Kasabach-Merritt syndrome: Therapeutic considerations. Pediatrics 1987;79:971–980.

6 Sato Y, Frey EE, Wicklund B, Kisker CT, Smith WL: Embolization therapy in the management of infantile hemangioma with Kasabach-Merritt syndrome. Pediatr Radiol 1987; 17:503–504.

7 Teillac-Hamel D, Prost YD, Bodemer C, Andry P, Enjolras O, Sebag G, Brunelle F, Hubert P, Nihoul-Fekete C: Serious childhood angiomas: unsuccessful alpha-2b interferon treatment. A report of four cases. Br J Dermatol 1993;129:473–476.

8 Yang YH, Lee PI, Lin KH, Tsang YM: Absolute ethanol embolotherapy for hemangioma with Kasabach-Merritt syndrome. Acta Paed Sin 1998;39:51–54.

9 Mueller BU, Mulliken JB: The infant with a vascular tumor. Semin Perinatol 1999;23:332–340.

10 Shim WKT: Hemangiomas of infancy complicated by thrombocytopenia. Am J Surg 1968; 116:896–906.

11 Enjolras O, Wassef M, Mazoyer E, Frieden IJ, Rieu P, Drouet L, Taïeb A, Stalder JF, Escande JP: Infants with Kasabach-Merritt syndrome do not have 'true' hemangiomas. J Pediatr 1997;130:631–640.

12 Sarkar M, Multiken JB, Kozakewich HPW, Robertson RL, Burrows PE: Throbocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg 1997;100:1377–1386.

13 Kontras SB, Green OC, King L, Duran RJ: Giant hemangioma with thrombocytopenia. Am J Dis Child 1963;105:188–195.

14 Propp RP, Scharfuran WB: Hemangioma-thrombocytopenia syndrome associated with microangiopathic hemolytic anemia. Blood 1966;28:623–633.

15 Rodriguez-Erdmann F, Button L, Murray JE, Moloney WC: Kasabach-Merritt syndrome: Coagulo-analytical observations. Am J Med Sci 1971;261:9–15.

16 Weber TR, Connors RH, Tracy TF, Bailey PV: Complex hemangiomas of infants and children. Individualized management in 22 cases. Arch Surg 1990;125:1017–1021.

17 Hatley RM, Sabio H, Howell CG, Flickfinger F, Parrish RA: Successful management of an infant with a giant hemangioma of the retroperitoneum and Kasabach-Merritt syndrome with alpha-interferon. J Pediatr Surg 1993;28: 1356–1359.