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Case report

Retrospective study on the outcomes of infantile tufted angioma complicated by Kasabach-Merritt Phenomenon

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ABSTRACT

Objective: To analyze the clinical characteristics and treatment of pediatric tufted angiomas(TA)complicated by Kasabach-Merritt Phenomenon (KMP).

Method: A retrospective analysis was conducted on the clinical data and follow-up data of 13 patients diagnosed with TA complicated by KMP. Five male and 8 female patients with an average age of 5.7 months (range, 29 days to 1 year) were treated with surgery between January 2009 and June 2012. According to the size and location of lesions and the degree of thrombocytopenia, complete or subtotal resection was performed. The median follow-up period was 3.4 years (range, 1.7 years to 5.2 years). Therapeutic outcomes were evaluated by platelet count and lesion size.

Results: Curative treatment of KMP is defined as restoration of normal hemostasis and elimination of tumor cells. Twelve patients achieved curative treatment and one died of multiple organ failure after operation. Ten patients received complete resection and three patients received incomplete resection. Thrombocyte count, hemoglobin and blood coagulation were respectively restored to normal levels within 1–3 days and 1–2 weeks post complete resection operation. One of the three patients who received subtotal resection operation died. In the other two patients, the platelet count fluctuated over time but remained above 60×10^9 /L, a significantly higher level than the preoperational level. Residual lesions slowly disappeared after continuous medication 3–6 months post operation.

Conclusion: Early surgical treatment of patients with TA complicated with KMP resulted in significantly higher curative rate and reduced side-effects of drugs.

1. Introduction

Tufted angioma (TA) is a rare vascular tumor that usually presents as brown, blue, or dusky red to violaceous infiltrated macules, plaques, or nodules of various sizes. In a few patients, local tender hyperhidrosis or hypertrichosis were observed. TA was named due to its typical distribution pattern as small tufts of capillaries in histology [1–3]. It is also called Nakagawa angioblastoma since it was first reported by Nakagawa in 1949 [4]. MacMillan and Champion described it as a progressive capillary hemangioma in 1971 [5]. Wilson Jones reported 10 similar cases in 1976 and formally designated these vascular tumors as TA based on the characteristic histological findings [6, 7].

TA is sporadic and most lesions develop slowly. Some cases can be complicated by Kasabach-Merritt Phenomenon (KMP), an aggressive and life-threatening condition characterized by profound thrombocytopenia and consumptive coagulopathy with a high mortality rate of 37.5% [8]. However, no guidelines for the treatment of TA complicated with KMP has been established, and only a limited number of TA cases

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Table 1
Clinical data of 13 TA patients with KMP.

Case	Gender	Age at admission	Age at onset of TA	Age at onset of KMP	Lesion site	The least PLT($\times 10^9$/L)	Lesion size(cm^3)	Associated symptoms and signs	Previous treatments	Outcome
1	F	1 year	At birth	1 month	Right thigh	9	16 × 10 × 1.5	None	Methylprednisone	Heal
2	F	6 months	1 month	5 months	Right thigh	6	15 × 10 × 2	None	Pulse laser	Heal
3	F	1 year	1 month	4 months	Right abdomen	23	15 × 20 × 3	None	Prednisone	Heal
4	M	2 months	At birth	2 months	Abdomen	45	6 × 7 × 1	Hyperhidrosis	None	Heal
5	F	6 months	At birth	4 months	Right neck, scapula, back	69	12 × 7 × 1.5	Tender, hyperhidrosis, hypertrichosis	Methylprednisone,prednisone	Heal
6	M	2 months	At birth	1 month	Left occipital	25	7 × 7 × 3	Tender, hyperhidrosis	Symptomatic treatment	Heal
7	F	29 days	At birth	At birth	Left face, submandibular, neck	35	6 × 5 × 3	None	Dexamethasone	Heal
8	M	9 months	At birth	At birth	Left abdomen	75	10.5 × 6.5 × 2.5	Hyperhidrosis	Vincristine	Heal
9	F	4 months	At birth	3 months	Chin, submental	15	10 × 8 × 2	Hyperhidrosis, hypertrichosis	Prednisone,IFN, propranolol	Heal
10	F	7 months	At birth	At birth	Right lower leg, knee	12	14 × 6 × 4	Tender	Antibiotics, symptomatic treatment	Heal
11	F	54 days	At birth	21 days	Right waist, gluteal,perineum	15	25 × 25 × 5	Tender	Incision and drainage	Heal
12	M	7 months	4 days	1 month	Right thigh, hip, groin	4	40 × 21 × 4	None	Interventional embolization two times	Heal
13	M	5 months	20 days	20 days	Right cervicofacial, mastoid region submental, occipital, neck	25	15 × 10 × 3	Hyperhidrosis	Steroid,repeated platelet and RBC transfusion	Dead

associated with KMP have been reported [9–11]. In the present study, we retrospectively analyzed the clinical manifestations and outcomes of surgical treatment of patients diagnosed with TA complicated by KMP. The analysis provides systematic clinical information on the management of TA patients complicated by KMP.

2. Materials and methods

2.1. Clinical materials

13 patients with TA complicated by KMP admitted into the Henan Provincial People's Hospital between January 2009 and June 2012 were enrolled in this study. Data collected included gender, admission age, onset age of TA and KMP, location of lesion, results of laboratory tests, imaging tests (when available), associated symptoms and signs, treatment and outcomes.

Diagnosis of TA was based on clinical manifestations, Doppler ultrasound and MRI examination [2, 3, 11]. The pathological diagnosis after surgery was consistent with the initial diagnosis. The histology showed tightly packed capillaries in a cannonball pattern located in the mid-to-deep dermis and surrounded by a crescent-shaped channel [1, 3, 10]. Laboratory evaluation of severe thrombocytopenia and consumptive coagulopathy is essential for the diagnosis of KMP [8, 12]. Effective treatment of KMP was defined as a recovery of hemostatic parameters and reduction of tumor size. Curative treatment of KMP was defined as restoration of hemostatic parameters and elimination of the tumor [12, 13].

This study was approved by the Henan Provincial People's Hospital Clinic Ethics Board, and conformed to the provisions of the Declaration of Helsinki. Written informed consents were obtained from all study subjects.

2.2. Therapeutic Methods

All patients received sequential combination therapy with medicine and surgeries.

2.2.1. Pre-operational stage

Patients received intravenous methylprednisolone (3.0–5.0 mg/kg/day) or gamma immunoglobulin (400 mg/kg/day) depending on the level of platelet count and previous treatment before admission into this hospital. If the platelet count continued to drop, infusion of gamma immunoglobulin (400 mg/day) was prolonged for 7 days. If the platelet count increased to the normal level or above 5×10^9/L, carbonyldiamide at a dosage of 400 g/L was injected into the local lesion daily or every other day. Prior to surgery, supportive care including nutritional support and symptomatic treatment for anemia was provided. When necessary, transfusion of platelet, cryoprecipitate, or plasma was prescribed to improve coagulation function.

2.2.2. Operational stage

Surgical tourniquets were routinely used at the excising lesions on the extremities. Infusion of saline or 1:500,000 epinephrine diluted in saline solution into the lesions was also given when the surgery involved more than one anatomical regions on the trunk. Since the lesions often invaded skin, subcutaneous tissue, musculoskeletal and bones, the tumor must be differentiated from the proximate normal tissues, and nerves as well as blood vessels must be protected. The excision of the muscle depended on the depth of infiltration and if the muscle is still functional. Optimal incision and suture techniques of the lesions on the abdominal wall could reduce the incidence of wound failure and postoperative hernia. Complete or subtotal resection was performed based on the size and location of the lesion, invasion, and proximity with surrounding critical structures. Lastly, the redundant skin was trimmed, full thickness flap was reserved, drain tube was placed, the wound was closed without tension, and the wound was covered and wrapped with proper compression to reduce postoperative bleeding.

2.2.3. Post-operational stage

Efforts were paid to keep airway clear and prevent airway obstruction after surgery, especially during the head and neck procedures. The position of limb and trunk were lifted for comfort and the local blood flow was frequently checked. The drain tube was ensured to